


Epidemiology for primary brain tumors: a nationwide population-based study

Amélie Darlix^{1,2}  · Sonia Zouaoui^{3,4} · Valérie Rigau⁵ · Faiza Bessaoud³ · Dominique Figarella-Branger⁶ · Hélène Mathieu-Daudé³ · Brigitte Trétarre³ · Fabienne Bauchet³ · Hugues Duffau^{2,4} · Luc Taillandier⁷ · Luc Bauchet^{2,4}

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Abstract Primary central nervous system tumors (PCNST) are rare tumors responsible for high mortality and morbidity. Their epidemiology is poorly known, and clinical data are scarcely analyzed at a national level. In this study, we aimed at providing descriptive epidemiological data and incidence rates for all histological subtypes of PCNST according to the WHO classification. We conducted a nationwide population-based study of all newly diagnosed and histologically confirmed PCNST in France, between 2006 and 2011. A total of 57,816 patients were included: male 46.4%, median age at diagnosis 56 years old (range 0–99). For all newly diagnosed

PCNST with histological confirmation the crude incidence rate was 15.5/10⁵ per 100,000 person-years. To enable international comparisons, standardized rates were calculated: 14.1/10⁵ (population of reference: USA), 14.5/10⁵ (population of reference: Europe), and 12.0/10⁵ (population of reference: world). 23.4% of samples were cryopreserved. Resection was performed in 79.1% of cases. Results are detailed (incidence rate, sex ratio, median age at diagnosis, number of cryopreserved samples, and type of surgery) for each of the 143 histological subtypes of PCNST, including all rare tumors. For example, incidence rates (population of reference: USA) were 0.018/10⁵ for anaplastic gangliogliomas, 0.054/10⁵ for malignant meningiomas, and 0.036/10⁵ for hemangiopericytomas. Our study is the first to describe incidence rates and epidemiological data for all histological subtypes of PCNST, including rare tumors, at a national level. Its methodology ensures the exhaustiveness of the data collection for histologically-proven cases. Histological population-based studies have many perspectives in the field of clinical epidemiology and research.

The original version of this article was revised: In the first line of Table 1 (line “Tumors of neuroepithelial tissue”), column “Male, n”, the number was corrected to read 14,621.

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✉ Amélie Darlix
amelie.darlix@icm.unicancer.fr

¹ Department of Medical Oncology, Institut Régional du Cancer de Montpellier (ICM)—Val d’Aurelle, 208 rue des Apothicaires, 34298 Montpellier, France

² INSERM U1051, 80 avenue Augustin Fliche, 34091 Montpellier, France

³ Department of Epidemiology, French brain tumor database, GNOLR, Registre des Tumeurs de l’Hérault, Institut Régional du Cancer de Montpellier (ICM)—Val d’Aurelle, 208 rue des Apothicaires, 34298 Montpellier, France

⁴ Department of Neurosurgery, Gui de Chauliac Hospital, 80 avenue Augustin Fliche, 34090 Montpellier, France

⁵ Department of Pathology, Gui de Chauliac Hospital, 80 avenue Augustin Fliche, 34090 Montpellier, France

⁶ Department of Neuropathology and INSERM U911, Timone Hospital, Université de la Méditerranée, 40 rue Ste Baume, 13010 Marseille, France

⁷ Neuro-oncology Unit, University Hospital, 29 avenue du Maréchal de Lattre de Tassigny, Nancy, France

Introduction

Primary central nervous system tumors (PCNST) are defined as all primary tumors located within the central nervous system (CNS) including the envelopes of the CNS and the origin of the nerves [1–3]. They show a malignant, benign or uncertain evolution. PCNST represent a heterogeneous group of tumors, with 143 histological subtypes according to the World Health organization (WHO) classification [1–3]. They have different causes, prognostic factors and treatments. The incidence rate (IR) of all PCNST ranges from $17.6/10^5$ to $22.0/10^5$ in American [4] and European studies [5–7]. However, considering the high number of histological subtypes, nearly each subtype may be considered as a rare tumor. These tumors are a major public health issue as they are responsible for high morbidity [8] and mortality [4, 8].

Epidemiological data on PCNST are poor, as the declaration is not mandatory in many countries. Moreover, brain tumor registration at the worldwide level is often limited to malignant tumors, with the exclusion of non-malignant or borderline tumors [9]. The difficulties for tumor registries are numerous. An exhaustive recording of all cases is very time-consuming as it requires collecting data from a high number of sources: death certificates, data from pathologists, radiologists, neurologists and neurosurgeons [6]. For these reasons, except in countries where declaration is mandatory, tumors with no histological diagnosis are particularly challenging to collect.

There are few national registries. Among them, the Central Brain Tumor Registry of the United States (CBTRUS) provides extensive data on PCNST, classified according to a histology-grouping scheme (without detail of all the WHO subtypes) [4]. The Austrian Brain Tumor Registry [5] and the registries from Scandinavia [10–12] and England [13] also report descriptive epidemiological data at a national level. Some specific registries [14–16] and regional registries (in Spain, Italy, France...) [6, 17, 18] also provide information. However, the registries that collect all histological subtypes described by the WHO 2007 classification do not report results on all subtypes as they use various histology-grouping schemes [4]. Moreover, clinical and biological data, despite their usefulness (in particular for rare tumors), are rarely investigated.

Our study primary objective was to provide descriptive epidemiological data and the IRs of all histological subtypes of PCNST at the national level, including all rare subtypes of PCNST for which a histological registry is crucial. Even though our data is based on the 2007 WHO classification [1, 2], and not on the recently published 2016 classification [3], it will serve as a reference for future epidemiological studies, as the incidence rates

of all subtypes of PCNST described by the 2007 classification have not yet been reported.

Materials and methods

Settings

The French Brain Tumor Database (FBTDB) is a national histological database of all PCNST in France. It is based on a network of all neurosurgeons, pathologists and neuro-oncologists involved in PCNST, in collaboration with all the societies focused on PCNST. Its methodology has previously been published [19–21]. The FBTDB's main objective is to prospectively record all histologically-proven cases of PCNST diagnosed in France. Data are collected from two sources. First, a data sheet, that is available in all operating rooms where neurosurgery is performed, is filled by both the neurosurgeon and the pathologist. Second, listings of all cases analyzed by each pathology department are collected annually, to ensure the exhaustiveness of the collection [22].

Design

We conducted a nationwide population-based study of newly diagnosed and histologically confirmed PCNST in Metropolitan France between 2006 and 2011.

Patients

All patients (no age limit) with a histologically-proven PCNST diagnosed between 2006, January 1st and 2011, December 31st in metropolitan France were included (metropolitan France includes mainland France and nearby islands in the Atlantic Ocean and Mediterranean Sea, and excludes overseas territories). Tumors were described using the 2000 WHO classification until 2006 [23], then the 2007 WHO classification [1, 2]. Note: pituitary tumors do not belong to the PCNST according to the WHO classification [1], but are included in a few PCNST registries such as the CBTRUS, and were included in this study.

Exclusion criteria were as follows: secondary tumors of the SNC (metastases), and duplicate records for recurrent disease (in that case, data from the first surgery was recorded if within the inclusion period).

Main measures

The IR, sex ratio, median age at diagnosis, number of cryopreserved samples, and type of surgery (resection or biopsy) were provided for each histological subtype

according to the ICD-O-3 (International Classification of Diseases for Oncology) PCNST classification [1–3, 23] and to the Systematized Nomenclature of Medicine (SNOMED) codes from Louis et al. (the list of the included codes is provided in Table 1) [2].

In order to reduce the number of cases with an imprecise histology, in particular gliomas labeled as “not otherwise specified”, an analysis of the pathological report by an expert pathologist (V.R.) was performed for cases of diffuse gliomas with unclear grading according to the WHO classification.

Statistical analysis

Crude and age-standardized IRs per 100,000 person-years were computed for each histological subtype of PCNST. Crude IR is the total number of new cases divided by the corresponding population and multiplied by 100,000. To enable international comparisons, standardized IRs based on a direct standardization model were also calculated with Europe, USA and worldwide populations as references. Crude rates, worldwide-, Europe- and USA-population standardized rates will be referred to, from now on, as CR, WSR, ESR and USR, respectively. The statistical analyses were performed using the SAS software.

Ethical approval

This study was approved by the French legislation, and by all the French Societies involved in neuro-oncology: Société Française de Neuropathologie (SFNP), the Société Française de Neurochirurgie (SFNC) and the Club Neuro-Oncologie of the Société Française de Neurochirurgie (CNO-SFNC), and the Association des Neuro-Oncologues d'Expression Française (ANOCEF).

Results

Characteristics of the population

Between 2006 and 2011, 57,816 incident cases of PCNST were recorded. The case distribution according to the year of diagnosis is presented in Table 2 and shows no major fluctuation in time. Among the patients, 46.4% were male, corresponding to a ratio male/female of 0.86. The mean age was 52.6 years (51.9 for males and 53.3 for females). The median ages at diagnosis for all cases, male cases and female cases were 56 (range 0–99), 56 (range 0–96) and 56 (range 0–99) years, respectively. At diagnosis, 4068 patients (7.0%) were aged under 20, and 2948 patients (5.1%) were under 15. The tumor distribution according to the histology group is shown in Fig. 1. Tumors of the

neuroepithelial tissue accounted for about 44% (including gliomas 39%) while tumors of the meninges accounted for about one-third and tumors of the cranial and paraspinal nerves for about 10% of cases. The characteristics of all the cases [number of cases, median ages by gender, number of cryopreserved samples, and type of surgery (biopsy or resection)] according to the histological subtype are presented in Table 1. Resection was performed in 79.1% of patients for whom surgical data was available, i.e. 73.2% of all patients (and 64.9% of tumors of the neuroepithelial tissue, 62.3% of glioblastomas, 93.1% of ependymal tumors, 90.4% of embryonal tumors, 98.0% of meningiomas...) (Table 1). 13 508 samples (23.4%) were cryopreserved, among which 7048 gliomas, 3481 meningiomas, and a number of rare subtypes of PCNST (Table 1).

Incidence data

The CR for all newly diagnosed PCNST with histological confirmation was $15.5/10^5$ per person-years (Table 3). To allow international comparisons, age-standardized IRs were calculated: USR $14.1/10^5$, ESR $14.5/10^5$, and WSR $12.0/10^5$. For patients aged <15, the CR for all PCNST was $4.34/10^5$. Table 3 shows the standardized IR for all the histological subtypes of PCNST. Concerning rare tumors, examples of CR are the following: CR_{anaplastic gangliogliomas}: $0.018/10^5$, among rare subtypes of meningiomas: CR_{chordoid meningiomas}: $0.021/10^5$ and CR_{anaplastic meningiomas}: $0.056/10^5$, among medulloblastomas: CR_{all medulloblastomas}: $0.179/10^5$ ($0.56/10^5$ for patients aged <15 years), and CR_{desmoplastic medulloblastomas}: $0.025/10^5$, CR_{all hemangiopericytomas}: $0.041/10^5$, CR_{choroid plexus carcinoma}: $0.006/10^5$, CR_{central neurocytoma}: $0.042/10^5$.

Discussion

Our study reports, for the first time, data on all histological subtypes of PCNST according to the WHO classification, together with their IRs, at a national level. It provides the very first data as regards to IRs for many rare subtypes of PCNST. This work was made possible by the collaboration of neurosurgeons and neuropathologists, with the methodological support of epidemiologists, and emphasizes the relevance of creating multidisciplinary networks and databases involving clinicians, pathologists and epidemiologists.

Differences and similarities between histological population-based studies and registries for PCNST

The IRs for all PCNST in our study are: CR $15.5/10^5$ (compared with $17.6/10^5$ in the Gironde Registry [6]), USR $14.1/10^5$ (compared with $22.0/10^5$ in the CBTRUS [4]),

Table 1 Case distribution by histological subtype, with number of cases, sex-ratio, median ages by gender, type of surgery, and number of cryopreserved samples (period of inclusion: 2006–2011)

Histology	ICD-O SNOMED	n	Male, n	Female, n	Sex ratio	Median age male	Median age female	Median age, female	Cryopre- servation, n	Resection, n	Resection ^a , %	Biopsy, n	Biopsy ^a , %
Tumors of neuroepithelial tissue		25,183	14,621	10,562	1.38	56	56	57	7740	12,621	64.9	6835	35.1
Gliomas		22,642	13,206	9436	1.40	58	58	59	7048	11,064	62.5	6650	37.5
Glioma, NOS	9380/3	481	279	202	1.38	55.0	54.0	56.0	66	144	36.8	263	63.2
Astrocitic tumors		15,331	8975	6356	1.41	61.0	61.0	62.0	4897	7188	62.3	4344	37.7
Astrocytoma, NOS	9400/3	279	157	122	1.29	48.0	47.0	48.0	58	101	41.4	143	58.6
Piloicytic astrocytoma	9421/1	1074	551	523	1.05	12.0	12.0	13.0	327	687	87.9	95	12.1
Piloxyoid astrocytoma	9425/3	10	4	6	0.67	4.5	2.0	7.5	4	5	62.5	3	37.5
Subependymal giant cell astrocytoma	9384/1	89	48	41	1.17	15.0	14.5	17.0	28	65	98.5	1	1.5
Pleomorphic xantho-astrocytoma	9424/3	87	44	43	1.02	28.0	29.5	28.0	28	42	85.7	7	14.3
Fibrillary astrocytoma	9420/3	183	111	72	1.54	42.0	46.0	38.0	56	55	37.2	93	62.8
Gemistocytic astrocytoma	9411/3	93	55	38	1.45	46.0	46.0	47.0	31	44	56.4	34	43.6
Protoplasmic astrocytoma	9410/3	16	7	9	0.78	42.5	42.0	52.0	3	7	46.7	8	53.3
Anaplastic astrocytoma	9401/3	635	366	269	1.36	58.0	57.5	59.0	186	178	32.6	368	67.4
Glioblastoma	9440/3	12,410	7355	5055	1.45	63.0	64.0	64.0	4026	5780	62.3	3496	37.7
Giant cell glioblastoma	9441/3	196	115	81	1.42	57.0	56.0	60.0	58	112	75.7	36	24.3
Gliosarcoma	9442/3	146	92	54	1.70	61.0	59.5	64.0	62	104	92.9	8	7.1
Gliomatosis cerebri ^b	9381/3	113	70	43	1.63	52.0	50.5	59.0	30	19	25.7	55	74.3
Oligodendroglial tumors		3428	1950	1478	1.32	49.0	49.0	49.0	1091	1872	60.9	1201	39.1
Oligodendroglioma	9450/3	1878	1078	800	1.35	44.0	44.0	44.0	573	982	58.3	702	41.7
Anaplastic oligodendrogloma	9451/3	1550	872	678	1.29	55.0	55.0	55.0	518	890	64.1	499	35.9
Oligoastrocytic tumors		2045	1184	861	1.38	50.0	50.5	50.0	613	967	55.5	774	44.5
Oligoastrocytic tumors, NOS	9382/3	147	89	58	1.53	45.0	45.0	44.0	35	49	62.8	29	37.2
Oligoastrocytoma	9382/3	713	407	306	1.33	40.0	42.0	40.0	219	326	54.5	272	45.5
Anaplastic Oligoastrocytoma	9382/3	1185	688	497	1.38	56.0	55.0	57.0	359	592	55.6	473	44.4
Ependymal tumors		1357	818	539	1.52	42.0	41.0	43.0	381	882	93.1	65	6.9
Subependymoma	9383/1	135	99	36	2.75	53.0	53.0	45.5	45	93	91.2	9	8.8
Myxopapillary ependymoma	9394/1	214	132	82	1.61	38.0	37.5	44.0	58	146	97.3	4	2.7
Ependymoma, NOS	9391/3	706	401	305	1.31	45.0	45.0	45.0	176	448	92.8	35	7.2

Table 1 (continued)

Histology	ICD-O	SNOMED	n	Male, n	Female, n	Sex ratio	Median age, male	Median age, female	Median age, cryopreservation, n	Resection, n	Resection, %	Biopsy, n	Biopsy, %
Cellular ependymoma	9391/3		41	23	18	1.28	33.0	37.0	10	19	90.5	2	9.5
Papillary ependymoma	9393/3		19	14	5	2.80	32.0	35.0	4	13	100.0		0.0
Clear cell ependymoma	9391/3		40	26	14	1.86	22.0	34.0	10	28	90.3	3	9.7
Tanicytic ependymoma	9391/3		18	11	7	1.57	45.0	45.0	6	10	83.3	2	16.7
Anaplastic ependymoma	9392/3		184	112	72	1.56	14.0	17.0	72	125	92.6	10	7.4
Choroid plexus tumors			179	81	98	0.83	25.0	30.5	48	119	90.8	12	9.2
Choroid plexus papilloma	9390/0		151	67	84	0.80	27.0	30.5	44.00	105	92.1	9	7.9
Atypical choroid plexus papilloma	9390/1		4	2	2	1.00	64.5	64.5	1.00	3	100.0		0.0
Choroid plexus carcinoma	9390/3		24	12	12	1.00	23.5	23.5	3.00	11	78.6	3	21.4
Other neuroepithelial tumors			57	31	26	1.19	47.0	44.0	12	30	85.7	5	14.3
Astroblastoma	9430/3		11	3	8	0.38	37.0	34.0	4	7	100.0		0.0
Chordoid glioma of the third ventricle	9444/1		5	3	2	1.50	34.0	51.5	2	1	25.0	3	75.0
Angiocentric glioma	9431/1		3	1	2	0.50	22.0	18.0	2	2	66.7	1	33.3
Esthesioneuroblastoma ^e	9522/3		38	24	14	1.71	49.0	54.0	4	20	95.2	1	4.8
Neuronal and mixed neuronal-glioma tumors			1220	650	570	1.14	24.0	26.0	300	745	92.0	65	8.0
Dysplastic gangliocytoma of cerebellum (Lhermitte-Duclos)	9493/0		8	4	4	1.00	36.0	35.5		3	75.0	1	25.0
Desmoplastic infantile astrocytoma/ganglioglioma	9412/1		28	18	10	1.80	4.0	10.5	4	19	100.0		0.0
Dysembryoplastic neuroepithelial tumor	9413/0		279	163	116	1.41	17.0	19.0	78	188	94.5	11	5.5
Gangliocytoma	9492/0		57	25	32	0.78	40.0	48.5	9	32	86.5	5	13.5
Ganglioglioma	9505/1		536	285	251	1.14	19.0	18.0	147	341	92.2	29	7.8
Anaplastic ganglioglioma	9505/3		66	38	28	1.36	39.5	37.5	17	35	77.8	10	22.2
Central neurocytoma	9506/1		158	76	82	0.93	31.0	30.5	29	75	92.6	6	7.4
Extraventricular neurocytoma	9506/1		0										
Cerebellar liponeurocytoma	9506/1		0										
Papillary glioneuronal tumor	9509/1		4	3	1	3.00	13.5	16.0	3	3	100.0		0.0

Table 1 (continued)

Histology	ICD-O SNOMED	n	Male, n	Female, n	Sex ratio	Median age, male	Median age, female	Median age, n	Cryopreservation, n	Resection, n	Resection, %	Biopsy, n	Biopsy, %
Rosette-forming glioneuronal tumor of the fourth ventricle	9509/1	6	3	3	1.00	22.0	18.0	26.0	2	5	100.0		0.0
Paraganglioma	8680/1	78	35	43	0.81	53.5	50.0	55.0	11	44	93.6	3	6.4
Tumors of the pineal region		152	78	74	1.05	39.0	37.5	41.0	26	73	64.6	40	35.4
Pinealoma, NOS	9360/1	34	22	12	1.83	41.5	45.0	29.0	6	17	70.8	7	29.2
Pineocytoma	9361/1	60	24	36	0.67	49.0	41.5	51.0	9	30	63.8	17	36.2
Pineal parenchymal tumor of intermediate differentiation	9362/3	14	7	7	1.00	27.0	37.0	26.0	3	7	63.6	4	36.4
Pineoblastoma	9362/3	35	20	15	1.33	19.0	20.5	17.0	6	17	68.0	8	32.0
Papillary tumor of the pineal region	9395/3	9	5	4	1.25	36.0	41.0	26.5	2	2	33.3	4	66.7
Embryonal tumors		933	575	358	1.61	10.0	10.0	10.0	306	590	90.4	63	9.6
Medulloblastoma, NOS	9470/3	552	348	204	1.71	12.0	12.0	12.0	176	360	93.3	26	6.7
Desmoplastic medulloblastoma	9471/3	94	69	25	2.76	13.4	8.0	7.0	48	65	94.2	4	5.8
Medulloblastoma with extensive nodularity	9471/3	4	1	3	0.33	6.5	6.0	7.0	2	2	100.0		0.0
Anaplastic medulloblastoma	9474/3	15	11	4	2.75	5.0	6.0	5.0	7	11	91.7	1	8.3
Large cell medulloblastoma	9474/3	3	2	1	2.00	6.0	7.0	6.0	2	3	100.0		0.0
CNS primitive neuroectodermal tumor	9473/3	124	75	49	1.53	12.5	14.0	10.0	28	59	77.6	17	22.4
CNS Neuroblastoma	9500/3	36	18	18	1.00	2.0	1.0	2.5	7	19	70.4	8	29.6
CNS Ganglioneuroblastoma	9490/3	11	6	5	1.20	19.0	27.5	19.0	3	3	60.0	2	40.0
Medulloepithelioma	9501/3	4		4	0.00	4.0		4.0	1	2	100.0		0.0
Ependymoblastoma	9392/3	1		1	0.00	25.0		25.0					
Atypical teratoid/rhabdoid tumor	9508/3	89	45	44	1.02	1.0	1.0	2.0	32	66	93.0	5	7.0
Tumors of the cranial and paraspinal nerves		5487	2564	2923	0.88	52.0	51.0	53.0	913	3620	95.7	163	4.3
Schwannoma (neurinoma, neurinoma)	9560/0	4781	2241	2540	0.88	53.0	52.0	53.0	802	3224	96.8	107	3.2
Schwannoma ^d (Neurofibromatosis)	9560/0	219	100	119	0.84	55.0	52.5	56.0	25	115	70.6	48	29.4
Cellular schwannoma	9560/0	54	19	35	0.54	52.5	45.0	55.0	9	32	100.0		0.0

Table 1 (continued)

History	ICD-O	SNOMED	n	Male, n	Female, n	Sex ratio	Median age, male	Median age, female	Cryopreservation, n	Resection, n	Resection, %	Biopsy, n	Biopsy, %
Plexiform schwannoma	9560/0		23	6	17	0.35	29.0	27.0	1	13	100.0		0.0
Melanotic schwannoma	9560/0		7	3	4	0.75	46.0	42.5	3	6	100.0		0.0
Neurofibroma, NOS	9540/0		213	103	110	0.94	38.0	39.5	33	132	98.5	2	1.5
Plexiform neurofibroma	9550/0		26	14	12	1.17	29.0	27.0	5	14	100.0		0.0
Neurofibroma ^d (Neurofibromatosis)	9540/0		79	41	38	1.08	32.0	33.5	14	35	94.6	2	5.4
Perineurioma, NOS	9571/0		10	2	8	0.25	41.0	42.0	2	8	88.9	1	11.1
Perineurioma, malignant	9571/3		0										
Malignant peripheral nerve sheath tumor (MPNST)	9540/3		55	23	32	0.72	45.0	42.0	11	31	91.2	3	8.8
Epithelioid MPNST	9540/3		2	1	1	1.00	63.5	50.0	1	2	100.0		0.0
MPNST with mesenchymal differentiation	9540/3		16	9	7	1.29	38.5	43.0	7	6	100.0		0.0
Melanotic MPNST	9540/3		1	1			49.0	49.0		1	100.0		0.0
MPNST with glandular differentiation	9540/3		1	1			28.0	28.0		1	100.0		0.0
Tumors of the meninges			18,806	5390	13,416	0.40	56.0	56.0	3810	13,371	97.8	298	2.2
Tumors of meningeothelial cells			16,018	3983	12,035	0.33	58.0	57.0	3481	11,637	98.0	238	2.0
Meningioma, NOS	9530/0		4156	970	3186	0.30	57.0	56.0	473	2909	98.4	48	1.6
Meningothelial meningioma	9531/0		4897	1178	3719	0.32	57.0	56.0	1288	3650	97.8	81	2.2
Fibrous (fibroblastic) meningioma	9532/0		1332	229	1103	0.21	58.0	58.0	320	996	98.8	12	1.2
Transitional (mixed) meningioma	9537/0		2239	492	1747	0.28	58.0	57.0	620	1674	97.6	42	2.4
Psammomatous meningioma	9533/0		636	86	550	0.16	63.0	63.0	118	471	97.3	13	2.7
Angiomatous meningioma	9534/0		256	102	154	0.66	59.5	59.0	54	183	96.8	6	3.2
Rare variety meningioma ^e , NOS	9530/0		349	78	271	0.29	57.0	56.0	54	238	97.9	5	2.1
Microcystic meningioma	9530/0		118	29	89	0.33	55.5	54.0	39	104	100.0		0.0
Secretory meningioma	9530/0		118	13	105	0.12	59.5	59.0	38	84	97.7	2	2.3
Lymphoplasmacyte-rich meningioma	9530/0		3		3	0.00	56.0	56.0	1	3	100.0		0.0
Clear cell meningioma	9538/1		49	14	35	0.40	52.0	50.0	20	43	97.7	1	2.3
Chordoid meningioma	9538/1		79	20	59	0.34	52.0	52.0	21	58	98.3	1	1.7

Table 1 (continued)

Histology	ICD-O SNOMED n	Male, n	Female, n	Sex ratio	Median age, male	Median age, female	Median age, cryopreservation, n	Resection, n	Resection, %	Biopsy, n	Biopsy, %	
Rhabdoid meningioma	9538/3	7	4	3	1.33	61.0	39.0	62.0	3	5	100.0	0.0
Metaplastic meningioma	9530/0	79	17	62	0.27	61.0	54.0	61.0	16	54	96.4	2
Atypical meningioma	9539/1	1479	644	835	0.77	62.0	64.0	60.0	353	1038	98.1	20
Papillary meningioma	9538/3	13	6	7	0.86	44.0	39.0	49.0	3	7	100.0	0.0
Anaplastic meningioma	9530/3	208	101	107	0.94	62.0	64.0	61.0	60	120	96.0	5
Mesenchymal tumors		1949	953	996	0.96	41.0	40.0	43.0	178	1169	97.0	36
Benign mesenchymal tumor ^e , NOS		2		2		31.5		31.5		2	100.0	0.0
Lipoma	8850/0	165	102	63	1.62	16.0	26.5	9.0	10	127	96.2	5
Angiolipoma	8861/0	23	12	11	1.09	47.0	49.0	47.0	2	17	100.0	0.0
Hibernoma	8880/0	0										
Liposarcoma	8850/3	2	2			64.0	64.0			1	100.0	0.0
Solitary fibrous tumor	8815/0	76	34	42	0.81	57.5	58.5	57.0	24	50	100.0	0.0
Fibrosarcoma	8810/3	3	1	2	0.50	56.0	42.0	68.5		1	50.0	1
Malignant fibrous histiocytoma	8830/3	0										50.0
Leiomyoma	8890/0	6	2	4	0.50	36.5	44.5	23.5	1	3	100.0	0.0
Leiomyosarcoma	8890/3	12	4	8	0.50	65.5	57.0	72.0	1	5	100.0	0.0
Rhabdomyoma	8900/0	0										
Rhabdomyosarcoma	8900/3	13	7	6	1.17	9	5	15.5		10	90.9	1
Chondroma	9220/0	9	5	4	1.25	51.0	52.0	38.0		6	100.0	0.0
Chondrosarcoma	9220/3	59	20	39	0.51	45.0	47.5	44.0	10	28	93.3	2
Osteoma	9180/0	61	16	45	0.36	44.0	31.5	45.0	5	46	100.0	0.0
Osteosarcoma	9180/3	12	8	4	2.00	29.0	33.5	28.0	2	7	100.0	0.0
Osteochondroma	9210/0	9	3	6	0.50	30.0	15.0	40.0	1	3	100.0	0.0
Haemangioma	9120/0	1316	648	668	0.97	40.0	38.0	41.0	87	752	96.8	25
Epithelioid hemangioma-thelioma	9133/1	4	3	1	3.00	62.0	60.0	64.0		1	100.0	0.0
Hemangiopericytoma	9150/1	103	41	62	0.66	54.5	56.0	53.5	19	63	100.0	0.0
Anaplastic haemangiopericytoma	9150/3	47	26	21	1.24	56.0	55.5	56.0	12	31	93.9	2
Angiosarcoma	9120/3	6	5	1	5.00	60.0	59.0	73.0		3	100.0	0.0
Kaposi sarcoma	9140/3	0										
Ewing's sarcoma—PNET	9364/3	21	14	7	2.00	27.0	22.0	44.0	4	13	100.0	0.0
Primary melanocytic lesions		90	53	37	1.43	58.0	57.0	60.0	8	49	84.5	9

Table 1 (continued)

History	ICD-O SNOMED	n	Male, n	Female, n	Sex ratio	Median age, male	Median age, female	Cryopreservation, n	Resection, n	Resection, %	Biopsy, n	Biopsy, %
Diffuse melanocytosis	8728/0	0										
Melanocytoma	8728/1	3	1	2	0.50	42.0	63.5		2	66.7	1	33.3
Malignant melanoma	8720/3	87	52	35	1.49	57.0	58.0	8	47	85.5	8	14.5
Meningeal melanomatosis	8728/3	0										
Other neoplasms related to the meninges		749	401	348	1.15	46.0	45.5	143	516	97.2	15	2.8
Haemangioblastoma	9161/1	749	401	348	1.15	46.0	45.5	143	516	97.2	15	2.8
Lymphomas and haematopoietic neoplasms		2053	1080	973	1.11	66	68	443	320	21.9	1140	78.1
Malignant lymphoma	9590/3, 9680/3	2010	1057	953	1.11	66.0	68.0	439	299	20.8	1139	79.2
Plasmacytoma	9731/3	43	23	20	1.15	59.0	59.0	4	21	95.5	1	4.5
Granulocytic sarcoma	9930/3	0										
Germ cell tumors		255	195	60	3.25	19.0	18.0	49	114	65.5	60	34.5
Germinoma	9064/3	117	99	18	5.50	19.0	17.5	25	43	46.2	50	53.8
Embryonal carcinoma	9070/3	5	3	2	1.50	27.0	29.0		1	50.0	1	50.0
Yolk sac tumor	9071/3	5	4	1	4.00	34.5	17.0	1	3	100.0		0.0
Choriocarcinoma	9100/3	3	3			30.0		1	1	50.0	1	50.0
Teratoma, NOS	9080/1	8	6	2	3.00	30.5	15.5	1	1	100.0		0.0
Teratoma, mature	9080/0	76	53	23	2.30	20.0	37.0	11	41	93.2	3	6.8
Immature germ cell tumors ^e , NOS	9080/3	22	17	5	3.40	15.0	14.0	2	10	66.7	5	33.3
Teratoma, immature	9080/3	16	8	8	1.00	13.5	1.0	7	11	100.0		0.0
Teratoma with malignant transformation	9084/3	0										
Mixed germ cell tumor	9085/3	3	2	1	2.00	18.0	18.0	1	3	100.0		0.0
Tumors of the sellar region		681	348	333	1.05	41.0	44.0	89	443	91.9	39	8.1
Craniopharyngioma	9350/1	597	310	287	1.08	37.0	46.0	78	396	91.5	37	8.5
Adamantinous craniopharyngioma	9351/1	66	30	36	0.83	36.5	37.0	9	37	97.4	1	2.6
Papillary craniopharyngioma	9352/1	14	6	8	0.75	44.5	47.0	2	9	90.0	1	10.0
Granular cell tumor	9582/0	0										
Pituitaryoma	9432/1	4	2	2	1.00	57.5	39.5		1	100.0		0.0
Spindle cell oncocytoma of the adenohypophysis	8291/0	0										
Pituitary tumors ^f		4786	2316	2470	0.94	52.0	49.0	390	2769	93.3	200	6.7
Pituitary adenoma, typical	8272/0	2163	1124	1039	1.08	54.0	52.0	184	1411	92.2	120	7.8

Table 1 (continued)

Histology	ICD-O SNOMED	n	Male, n	Female, n	Sex ratio	Median age, male	Median age, female	Cryopreservation, n	Resection, n	Resection ^a , %	Biopsy, n	Biopsy ^a , %
Non-secreting pituitary adenoma	8272/0	348	188	160	1.18	59.5	59.0	39	217	94.3	13	5.7
Pituitary adenoma with functional activity—Secreting	8272/0	178	78	100	0.78	53.0	50.5	5	53	93.0	4	7.0
Adenoma lactant	8204/0	1	1	0		37.0	37.0		1	100.0		0.0
FSH-secreting pituitary adenoma	8272/0	734	444	290	1.53	61.0	61.0	81	440	96.7	15	3.3
Prolactinoma	8271/0	407	115	292	0.39	30.0	29.0	23	220	93.2	16	6.8
Pituitary adenoma with multiple hormonal secretions	8272/0	143	59	84	0.70	52.0	53.0	13	105	92.1	9	7.9
GH-secreting pituitary adenoma	8272/0	325	164	161	1.02	46.0	50.0	16	122	97.6	3	2.4
TSH-secreting pituitary adenoma	8272/0	10	4	6	0.67	49.5	45.5		5	100.0		0.0
ACTH-secreting pituitary adenoma	8272/0	463	131	332	0.39	43.0	42.0	24	190	91.3	18	8.7
Malignant tumor of the pituitary gland (neuroendocrine carcinoma)	8272/3	14	8	6	1.33	56.5	53.0	5	5	71.4	2	28.6
Miscellaneous		565	304	261	1.16	50.5	48.0	74	191	60.4	125	39.6
Chordoma	9370/3	123	64	59	1.08	56.0	47.0	26	81	91.0	8	9.0
Malignant NOS / Suspicion	8000/1–3	140	79	61	1.29	48.0	50.0	22	45	48.4	48	51.6
Other		302	161	141	1.14	49.5	48.0	26	65	48.5	69	51.5
Total		57,816	26,818	30,998	0.87	56	56.0	13,508	33,449	79.1	8860	20.9

ICD-O International Classification of Diseases for Oncology, NOS not otherwise specified

^aThe proportion of biopsies and resection is provided for patients for which the information is available (for all tumors, 42,309 patients out of 57,816 included)

^bThe diagnosis of Gliomatosis cerebri is probably much underestimated because the histological diagnosis is most often based on a single histological specimen

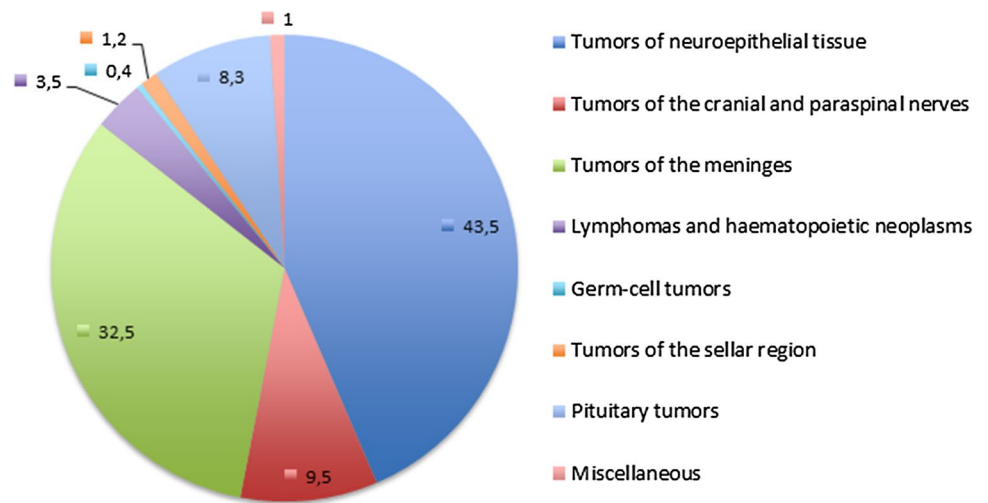
^cEsthesioneuroblastoma is included in the head and neck tumors in the WHO classification (pathology and genetics head and neck tumors Leon Barnes I ARC Press Lyon 2005), but it is included in the central brain tumor registry of the United States (CBTRUS), brain and central nervous system tumor histology groupings Ostrom et al. [4]

^dThe French classification allows specifying schwannoma or neurofibroma associated with neurofibromatosis (http://medphar.univ-poitiers.fr/register-cancers-poitou-charentes/documents_registro/adicap_version5_4_1_2009.pdf)

^eSpecific to the French classification (http://medphar.univ-poitiers.fr/register-cancers-poitou-charentes/documents_registro/adicap_version5_4_1_2009.pdf)

^fPathology and genetics tumours of endocrine organs. Ronald A. DeLellis IARC press Lyon 2004 p 10–47

Fig. 1 Distribution of the primary central nervous system tumors (PCNST) by histological type (%) (period of inclusion: 2006–2011)



ESR $14.5/10^5$ (compared with $18.1/10^5$ in the Austrian Brain Tumor Registry [5]), and WSR $12.0/10^5$ (Table 4). IRs in our study are lower than reported by the PCNST registries in Western countries [4–6]. However, the difference can mainly be explained by the fact that these registries include cases without histological confirmation. Indeed, non-histological cases represent 28.7% of cases in the data from the CBTRUS, 19.1% in the Austrian Brain Tumor Registry and 20.7% in the Gironde Registry. It is to be noted that the difference in incidence between a histological collection and a registry (which includes non-histologically proven cases) may provide interesting data as regards to the proportion of patients with a histological validation, as only a very small proportion of cases receives a tumor treatment without histological confirmation.

The absence of histological diagnosis is more frequent in the following situations: non-malignant tumors and/or tumors with a slow growth such as meningiomas or pituitary tumors [24]; elderly and/or frail patients for whom no oncological treatment is proposed; tumors located in poorly accessible CNS regions (for example, some cases of brainstem tumors); and rare cases for whom the diagnosis can be made on cerebrospinal fluid markers (for example, some cases of CNS germ-cell tumors). On the contrary, our

results are similar with the literature concerning major histology groups with uncertain or malignant evolution (for example, USR for neuroepithelial tissue tumors is $6.16/10^5$ in our study compared with $6.62/10^5$ in the CBTRUS [4], USR for lymphomas and hematopoietic neoplasms is $0.45/10^5$ compared with $0.46/10^5$ in the CBTRUS [4] and $0.50/10^5$ in the Gironde registry [6]).

Regarding the sex ratio for all PCNST, 46.4% of cases are male in our study, compared to 42.1% in the data from the CBTRUS [4]. The difference can be explained by the higher IR for meningiomas in the CBTRUS, as these tumors preferentially affect women. Median age for all PCNST was 56 in our study compared to 59 in the data from the CBTRUS [4], probably due to the non-inclusion of non-histological cases in our study. Among pediatric patients (aged < 15), our IR (CR = ESR = USR = $4.3/10^5$) was comparable to that of other studies: $3.9/10^5$ in the French National Registry of Childhood Solid Tumors [16], $4.1/10^5$ in the German registry ([14], http://www.kinderkrebsregister.de/fileadmin/kliniken/dkkr/pdf/jb/jb2013_2014/jb2014_s.pdf), and $5.26/10^5$ in the CBTRUS [25].

Strengths

Our study provides interesting data as regards IRs of rare tumors (Table 3). We will discuss a few examples. Among mixed glial-neuronal tumors, anaplastic gangliogliomas are rare tumors of unknown incidence, with only one study from the “Surveillance, Epidemiology and End-Results” (SEER) database reporting data on 58 patients between 1973 and 2007 [26]. In that study, the USR for anaplastic gangliogliomas was $0.002/10^5$, compared with a USR of $0.018/10^5$ in our study. The sex-ratio was comparable, with a predominance of males. However, we found a significant difference in the median age (39.5 years old in our study vs.

Table 2 Case distribution according to the year of diagnosis

Year	Cases recorded n (%)
2006	9263 (16.02)
2007	9480 (16.39)
2008	9783 (16.92)
2009	9941 (17.19)
2010	9709 (16.79)
2011	9640 (16.67)
Total	57,816 (100)

Table 3 Crude and standardized incidence rates per 100,000 (World, Europe and USA) by histological subtype (2006–2011)

Histology	ICD-O SNOMED	n	Incidence rate adjusted on the French population	Incidence rate adjusted on the worldwide population	Incidence rate adjusted on the Europe population	Incidence rate adjusted on the USA population
Tumors of neuroepithelial tissue		25,183	6.771	5.507	6.343	6.160
Gliomas		22,642	6.088	4.623	5.587	5.418
Glioma, NOS	9380/3	481	0.129	0.111	0.122	0.120
Astrocytic tumors		15,331	4.122	2.954	3.672	3.534
Astrocytoma, NOS	9400/3	279	0.075	0.067	0.073	0.072
Pilocytic astrocytoma	9421/1	1074	0.289	0.424	0.332	0.326
Pilomyxoid astrocytoma	9425/3	10	0.003	0.005	0.003	0.003
Subependymal giant cell astrocytoma	9384/1	89	0.024	0.034	0.027	0.027
Pleomorphic xantho-astrocytoma	9424/3	87	0.023	0.026	0.025	0.025
Fibrillary astrocytoma	9420/3	183	0.049	0.050	0.051	0.048
Gemistocytic astrocytoma	9411/3	93	0.025	0.021	0.025	0.024
Protoplasmic astrocytoma	9410/3	16	0.004	0.004	0.004	0.004
Anaplastic astrocytoma	9401/3	635	0.171	0.131	0.156	0.153
Glioblastoma	9440/3	12,410	3.337	2.102	2.864	2.742
Giant cell glioblastoma	9441/3	196	0.053	0.040	0.049	0.047
Gliosarcoma	9442/3	146	0.039	0.027	0.036	0.033
Gliomatosis cerebri ^a	9381/3	113	0.030	0.025	0.029	0.028
Oligodendroglial tumors		3428	0.922	0.753	0.893	0.880
Oligodendroglioma	9450/3	1878	0.505	0.437	0.502	0.501
Anaplastic oligodendroglioma	9451/3	1550	0.417	0.316	0.390	0.379
Oligoastrocytic tumors		2045	0.550	0.446	0.528	0.519
Oligoastrocytic tumors, NOS	9382/3	147	0.040	0.035	0.039	0.039
Oligoastrocytoma	9382/3	713	0.192	0.171	0.192	0.191
Anaplastic oligoastrocytoma	9382/3	1185	0.319	0.240	0.297	0.289
Ependymal tumors		1357	0.365	0.358	0.372	0.365
Subependymoma	9383/1	135	0.036	0.028	0.035	0.034
Myxopapillary ependymoma	9394/1	214	0.058	0.055	0.059	0.059
Ependymoma, NOS	9391/3	706	0.190	0.175	0.190	0.186
Cellular ependymoma	9391/3	41	0.011	0.012	0.011	0.011

Table 3 (continued)

Histology	ICD-O SNOMED	n	Incidence rate adjusted on the French population	Incidence rate adjusted on the worldwide population	Incidence rate adjusted on the Europe population	Incidence rate adjusted on the USA population
Papillary ependymoma	9393/3	19	0.005	0.005	0.005	0.005
Clear cell ependymoma	9391/3	40	0.011	0.013	0.012	0.011
Tanicytic ependymoma	9391/3	18	0.005	0.004	0.005	0.005
Anaplastic ependymoma	9392/3	184	0.049	0.067	0.055	0.053
Choroid plexus tumors		179	0.048	0.059	0.052	0.049
Choroid plexus papilloma	9390/0	151	0.041	0.051	0.044	0.042
Atypical choroid plexus papilloma	9390/1	4	0.001	0.001	0.001	0.001
Choroid plexus carcinoma	9390/3	24	0.006	0.008	0.007	0.006
Other neuroepithelial tumors		57	0.015	0.014	0.015	0.015
Astroblastoma	9430/3	11	0.003	0.003	0.003	0.003
Chordoid glioma of the third ventricle	9444/1	5	0.001	0.002	0.001	0.001
Angiocentric glioma	9431/1	3	0.001	0.001	0.001	0.001
Esthesioneuroblastoma ^b	9522/3	38	0.010	0.008	0.010	0.010
Neuronal and mixed neuronal-glia tumors		1220	0.328	0.397	0.356	0.355
Dysplastic gangliocytoma of cerebellum (Lhermitte–Duclos)	9493/0	8	0.002	0.002	0.002	0.002
Desmoplastic infantile astrocytoma/ganglioglioma	9412/1	28	0.008	0.012	0.009	0.008
Dysembryoplastic neuroepithelial tumor	9413/0	279	0.075	0.101	0.084	0.085
Gangliocytoma	9492/0	57	0.015	0.016	0.016	0.015
Ganglioglioma	9505/1	536	0.144	0.186	0.159	0.160
Anaplastic ganglioglioma	9505/3	66	0.018	0.017	0.018	0.018
Central neurocytoma	9506/1	158	0.042	0.044	0.045	0.044
Extraventricular neurocytoma	9506/1	0				
Cerebellar liponeurocytoma	9506/1	0				
Papillary glioneuronal tumor	9509/1	4	0.001	0.002	0.001	0.001

Table 3 (continued)

Histology	ICD-O SNOMED	n	Incidence rate adjusted on the French population	Incidence rate adjusted on the worldwide population	Incidence rate adjusted on the Europe population	Incidence rate adjusted on the USA population
Rosette-forming glioneuronal tumor of the fourth ventricle	9509/1	6	0.002	0.002	0.002	0.002
Paranglioma	8680/1	78	0.021	0.016	0.020	0.019
Tumors of the pineal region		152	0.041	0.042	0.042	0.042
Pinealoma, NOS	9360/1	34	0.009	0.009	0.009	0.010
Pineocytoma	9361/1	60	0.016	0.013	0.016	0.016
Pineal parenchymal tumor of intermediate differentiation	9362/3	14	0.004	0.004	0.004	0.004
Pineoblastoma	9362/3	35	0.009	0.012	0.010	0.010
Papillary tumor of the pineal region	9395/3	9	0.002	0.002	0.002	0.003
Embryonal tumors		933	0.251	0.372	0.290	0.281
Medulloblastoma, NOS	9470/3	552	0.148	0.212	0.169	0.167
Desmoplastic medulloblastoma	9471/3	94	0.025	0.039	0.030	0.028
Medulloblastoma with extensive nodularity	9471/3	4	0.001	0.002	0.001	0.001
Anaplastic medulloblastoma	9474/3	15	0.004	0.006	0.005	0.005
Large cell medulloblastoma	9474/3	3	0.001	0.001	0.001	0.001
CNS primitive neuroectodermal tumor	9473/3	124	0.033	0.048	0.038	0.036
CNS Neuroblastoma	9500/3	36	0.010	0.016	0.012	0.010
CNS Ganglioneuroblastoma	9490/3	11	0.003	0.004	0.003	0.003
Medulloepithelioma	9501/3	4	0.001	0.002	0.001	0.001
Ependymoblastoma	9392/3	1	0.000	0.000	0.000	0.000
Atypical teratoid/rhabdoid tumor	9508/3	89	0.024	0.043	0.030	0.027
Tumors of the cranial and paraspinal nerves		5487	1.475	1.161	1.416	1.373
Schwannoma (neurilemoma, neurinoma)	9560/0	4781	1.285	0.993	1.227	1.188
Schwannoma ^c (Neurofibromatosis)	9560/0	219	0.059	0.044	0.056	0.052
Cellular schwannoma	9560/0	54	0.015	0.011	0.014	0.013
Plexiform schwannoma	9560/0	23	0.006	0.006	0.006	0.006

Table 3 (continued)

Histology	ICD-O SNOMED	n	Incidence rate adjusted on the French population	Incidence rate adjusted on the worldwide population	Incidence rate adjusted on the Europe population	Incidence rate adjusted on the USA population
Melanotic schwannoma	9560/0	7	0.002	0.001	0.002	0.002
Neurofibroma, NOS	9540/0	213	0.057	0.054	0.058	0.058
Plexiform neurofibroma	9550/0	26	0.007	0.008	0.007	0.007
Neurofibroma ^c (Neurofibromatosis)	9540/0	79	0.021	0.023	0.023	0.023
Perineurioma, NOS	9571/0	10	0.003	0.002	0.003	0.003
Perineurioma, malignant	9571/3	0				
Malignant peripheral nerve sheath tumor (MPNST)	9540/3	55	0.015	0.013	0.015	0.014
Epithelioid MPNST	9540/3	2	0.001	0.000	0.000	0.000
MPNST with mesenchymal differentiation	9540/3	16	0.004	0.004	0.004	0.004
Melanotic MPNST	9540/3	1	0.000	0.000	0.000	0.000
MPNST with glandular differentiation	9540/3	1	0.000	0.000	0.000	0.000
Tumors of the meninges		18,806	5.056	3.634	4.624	4.500
Tumors of meningotheial cells		16,018	4.307	2.939	3.872	3.756
Meningioma, NOS	9530/0	4156	1.117	0.775	1.016	0.983
Meningothelial meningioma	9531/0	4897	1.317	0.912	1.197	1.155
Fibrous (fibroblastic) meningioma	9532/0	1332	0.358	0.240	0.320	0.309
Transitional (mixed meningioma)	9537/0	2239	0.602	0.410	0.542	0.526
Psammomatous meningioma	9533/0	636	0.171	0.105	0.143	0.141
Angiomatous meningioma	9534/0	256	0.069	0.046	0.061	0.059
Rare variety meningioma ^d , NOS	9530/0	349	0.094	0.066	0.085	0.083
Microcystic meningioma	9530/0	118	0.032	0.022	0.029	0.029
Secretory meningioma	9530/0	118	0.032	0.021	0.028	0.027
Lymphoplasmacyte-rich meningioma	9530/0	3	0.001	0.001	0.001	0.001
Clear cell meningioma	9538/1	49	0.013	0.010	0.012	0.012
Chordoid meningioma	9538/1	79	0.021	0.016	0.020	0.020

Table 3 (continued)

Histology	ICD-O SNOMED	n	Incidence rate adjusted on the French population	Incidence rate adjusted on the worldwide population	Incidence rate adjusted on the Europe population	Incidence rate adjusted on the USA population
Rhabdoid meningioma	9538/3	7	0.002	0.002	0.002	0.002
Metaplastic meningioma	9530/0	79	0.021	0.014	0.019	0.018
Atypical meningioma	9539/1	1479	0.398	0.261	0.345	0.340
Papillary meningioma	9538/3	13	0.003	0.003	0.003	0.004
Anaplastic meningioma	9530/3	208	0.056	0.036	0.048	0.048
Mesenchymal tumors		1949	0.524	0.509	0.535	0.527
Benign mesenchymal tumor ^d , NOS		2	0.001	0.001	0.001	0.001
Lipoma	8850/0	165	0.044	0.060	0.050	0.047
Angiolipoma	8861/0	23	0.006	0.005	0.006	0.006
Hibernoma	8880/0	0				
Liposarcoma	8850/3	2	0.001	0.000	0.000	0.000
Solitary fibrous tumor	8815/0	76	0.020	0.014	0.019	0.018
Fibrosarcoma	8810/3	3	0.001	0.000	0.001	0.001
Malignant fibrous histiocytoma	8830/3	0				
Leiomyoma	8890/0	6	0.002	0.002	0.002	0.002
Leiomyosarcoma	8890/3	12	0.003	0.002	0.003	0.003
Rhabdomyoma	8900/0	0				
Rhabdomyosarcoma	8900/3	13	0.003	0.005	0.004	0.004
Chondroma	9220/0	9	0.002	0.002	0.002	0.002
Chondrosarcoma	9220/3	59	0.016	0.014	0.016	0.016
Osteoma	9180/0	61	0.016	0.014	0.016	0.016
Osteosarcoma	9180/3	12	0.003	0.003	0.003	0.003
Osteochondroma	9210/0	9	0.002	0.003	0.003	0.002
Haemangioma	9120/0	1316	0.650	0.635	0.669	0.666
Epithelioid hemangioendothelioma	9133/1	4	0.001	0.001	0.001	0.001
Hemangiopericytoma	9150/1	103	0.028	0.021	0.026	0.025
Anaplastic haemangiopericytoma	9150/3	47	0.013	0.009	0.012	0.011
Angiosarcoma	9120/3	6	0.002	0.001	0.001	0.001
Kaposi sarcoma	9140/3	0				
Ewing's sarcoma—PNET	9364/3	21	0.006	0.006	0.006	0.006
Primary melanocytic lesions		90	0.024	0.017	0.021	0.022
Diffuse melanocytosis	8728/0	0				
Melanocytoma	8728/1	3	0.001	0.001	0.001	0.001

Table 3 (continued)

Histology	ICD-O SNOMED	n	Incidence rate adjusted on the French population	Incidence rate adjusted on the worldwide population	Incidence rate adjusted on the Europe population	Incidence rate adjusted on the USA population
Malignant melanoma	8720/3	87	0.023	0.017	0.021	0.021
Meningeal meningioma	8728/3	0				
Other neoplasms related to the meninges		749	0.201	0.169	0.196	0.196
Haemangioblastoma	9161/1	749	0.201	0.169	0.196	0.196
Lymphomas and haematopoietic neoplasms		2053	0.552	0.330	0.456	0.454
Malignant lymphoma	9590/3. 9680/3	2010	0.540	0.322	0.446	0.444
Plasmacytoma	9731/3	43	0.012	0.007	0.010	0.010
Granulocytic sarcoma	9930/3	0				
Germ cell tumors		255	0.069	0.090	0.077	0.076
Germinoma	9064/3	117	0.031	0.041	0.035	0.035
Embryonal carcinoma	9070/3	5	0.001	0.002	0.001	0.002
Yolk sac tumor	9071/3	5	0.001	0.002	0.001	0.001
Choriocarcinoma	9100/3	3	0.001	0.001	0.001	0.001
Teratoma, NOS	9080/1	8	0.002	0.003	0.002	0.002
Teratoma, mature	9080/0	76	0.020	0.026	0.023	0.022
Immature germ cell tumors ^d , NOS	9080/3	22	0.006	0.009	0.007	0.007
Teratoma, immature	9080/3	16	0.004	0.007	0.005	0.005
Teratoma with malignant transformation	9084/3	0				
Mixed germ cell tumor	9085/3	3	0.001	0.001	0.001	0.001
Tumors of the sellar region		681	0.183	0.184	0.186	0.183
Craniopharyngioma	9350/1	597	0.161	0.161	0.164	0.161
Adamantinous craniopharyngioma	9351/1	66	0.018	0.019	0.018	0.017
Papillary craniopharyngioma	9352/1	14	0.004	0.003	0.004	0.004
Granular cell tumor	9582/0	0				
Pituicytoma	9432/1	4	0.001	0.001	0.001	0.001
Spindle cell oncocy-toma of the adeno-hypophysis	8291/0	0				
Pituitary tumors ^e		4786	1.287	1.003	1.219	1.192
Pituitary adenoma, typical	8272/0	2163	0.582	0.434	0.542	0.530
Non-secreting pituitary adenoma	8272/0	348	0.094	0.065	0.084	0.083

Table 3 (continued)

Histology	ICD-O SNOMED	n	Incidence rate adjusted on the French population	Incidence rate adjusted on the worldwide population	Incidence rate adjusted on the Europe population	Incidence rate adjusted on the USA population
Pituitary adenoma with functional activity—Secreting	8272/0	178	0.048	0.038	0.046	0.044
Adenoma lactant	8204/0	1	0.000	0.000	0.000	0.000
FSH-secreting pituitary adenoma	8272/0	734	0.197	0.132	0.175	0.169
Prolactinoma	8271/0	407	0.109	0.114	0.115	0.114
Pituitary adenoma with multiple hormonal secretions	8272/0	143	0.038	0.030	0.037	0.036
GH-secreting pituitary adenoma	8272/0	325	0.087	0.074	0.087	0.085
TSH-secreting pituitary adenoma	8272/0	10	0.003	0.002	0.003	0.003
ACTH-secreting pituitary adenoma	8272/0	463	0.124	0.110	0.125	0.125
Malignant tumor of the pituitary gland (neuroendocrine carcinoma)	8272/3	14	0.004	0.003	0.004	0.003
Miscellaneous		565	0.152	0.132	0.146	0.144
Chordoma	9370/3	123	0.033	0.026	0.031	0.030
Malignant NOS / suspicion	8000/1–3	140	0.038	0.036	0.037	0.036
Other		302	0.081	0.069	0.078	0.077
Total		57,816	15.545	12.041	14.466	14.082

ICD-O International Classification of Diseases for Oncology, NOS not otherwise specified

^aThe diagnosis of Gliomatosis cerebri is probably much underestimated because the histological diagnosis is most often based on a single histological specimen

^bEsthesioneuroblastoma is included in the head and neck tumors in the WHO classification (pathology and genetics head and neck tumors Leon Barnes IARC Press Lyon 2005), but it is included in the Central Brain Tumor Registry of the United States (CBTRUS), brain and central nervous system tumor histology groupings Ostrom et al. [4]

^cThe French classification allows specifying schwannoma or neurofibroma associated with neurofibromatosis (http://medphar.univ-poitiers.fr/registre-cancers-poitou-charentes/documents_registre/adicap_version5_4_1_2009.pdf)

^dSpecific to the French classification (http://medphar.univ-poitiers.fr/registre-cancers-poitou-charentes/documents_registre/adicap_version5_4_1_2009.pdf)

^ePathology and genetics tumours of endocrine organs. Ronald A. DeLellis IARC press Lyon 2004 p 10–47

25.5 in the SEER study). Meningiomas are among the most common PCNST [4, 22] and comprise histological subtypes that are considered as rare tumors. For example, chordoid meningiomas are a rare subtype of WHO grade II meningiomas, with an unknown incidence. Their incidence was not reported in studies from the CBTRUS [4], and only small clinical and/or pathological series have been published so far [27, 28]. We found a CR of 0.02/10⁵, with a median age of 52 years old and a sex-ratio of 0.34. With a total of 79 patients with a chordoid meningioma registered by the FBTDB, the French series could bring interesting data

regarding the prognostic factors and treatment strategies for these patients. We also described CR for atypical meningiomas (0.398/10⁵), clear cell meningiomas (0.013/10⁵), anaplastic meningioma (0.056/10⁵), rhabdoid meningioma (0.002/10⁵), and papillary meningioma (0.003/10⁵). Among mesenchymal tumors, CNS hemangiopericytomas are rare, accounting for about 0.4% of all PCNST [2]. As the diagnosis of hemangiopericytoma requires a histological confirmation (and now a STAT6 immunohistochemistry), the IR found in our study is comparable to that of a study from the SEER database (USR 0.03/10⁵ in the SEER study

Table 4 Comparison of the incidence rates (per 100,000) between data issued from the FBTDB (2006–2011) and from other registries world-wide

Histology	FBTDB ^a				CBTRUS ^b , USR (2000 USA population) [4]	Austrian Brain Tumor Registry ^b , USR (2000 USA population) [5]	Gironde Registry ^b , CR (French population) [6]
	CR	WSR	ESR	USR			
Tumors of neuroepithelial tissue	6.77	5.50	6.34	6.15	6.61	7.26	7.94
Tumors of cranial and spinal nerves	1.47	1.16	1.41	1.37	1.70	1.36	2.16
Tumors of the meninges	5.05	3.63	4.62	4.49	8.18	5.31	6.68
Lymphomas and hematopoietic neoplasms	0.55	0.32	0.45	0.45	0.46	0.57	0.58
Germ cell tumors—malignant	0.06	0.09	0.07	0.07	0.10	0.09	0.09
	0.05	0.07	0.06	0.06			
Tumors of the sellar region	0.18	0.18	0.18	0.18	0.18	1.81	
Pituitary tumors	1.28	1.00	1.21	1.19	3.29		
Miscellaneous	0.16	0.13	0.14	0.14	0.88	1.78	0.61
Total	15.5	12.0	14.5	14.1	21.42	18.1	17.60

The grouping scheme is provided in Table 1 for data from the FBTDB and in each corresponding publication for data from the CBTRUS [4], the Austrian Brain Tumor Registry [5] and the Gironde Registry [6]

^aHistological population-based study

^bRegistry including cases with and without histological validation

compared with 0.036/10⁵ in our study), which further validates our results [29]. Likewise, the IR found for medulloblastomas in our study is comparable for children to that of the Canadian pediatric registry (CR 0.48/10⁵ compared with WSR 0.57/10⁵ in our study, data not shown), and for all patients to that of the SEER database in the USA (USR 0.15/10⁵ compared with 0.20/10⁵ in our study) [30, 31]. For all other rare tumors (excepted germ-cells tumors as these tumors can sometimes be diagnosed without a biopsy), for which the epidemiological and incidence data are rare, our study brings essential data on descriptive epidemiology, and provides a basis for clinical population-based studies.

Furthermore, the methodology of the FBTDB allows for an exhaustive recording of histological cases. Indeed, all the neurosurgery departments and pathologic labs involved in PCNST in metropolitan France participated in the data recording. Moreover, the collection of annual listing of cases from all the pathology departments ensured the exhaustiveness of the data collection. Such a collection is of high value, especially for rare tumors or tumors for which a clinical and/or radiological diagnosis is difficult. For these tumors, reaching a histological diagnosis is essential for clinical data analyses, oncological care management and outcomes at the population level. The number of cryopreserved tumors for each histological subtype of PCNST is described in Table 1. This information, even though it cannot be extrapolated to other countries where the practice may vary, is important as it gives an idea on the number of biological samples that could be used for

research purposes. The percentage of resection versus biopsy is also provided for more than 42,000 tumors and for each histological subtype (Table 1). This data and the analysis of surgical practice at the population level are useful for the public health organizations.

In metropolitan France, the FBTDB not only allows a comprehensive description of the epidemiology of PCNST with histological validation, but it also provides a base for clinical and population studies. These works are useful in many areas, such as the validation of prognostic factors or therapeutic strategies [32–34], or the search for risk factors [35]. This is particularly true for rare tumors for which collaborations at the national level are necessary to be able to provide data on a significant number of patients.

Moreover, histological population-based studies have a great value in countries where the declaration of PCNST is not mandatory, in particular for rare subtypes of tumors. They are easier and cheaper than general registries, as the collection of non-histological cases is very time-consuming. As few patients receive active anti-tumor treatments without a histological confirmation, a good knowledge of the number of cases to be expected is of great importance for the public health system.

Limitations

One of the limits of our study is that, due to its methodology, the FBTDB does not collect cases without histological confirmation, which causes an obvious

bias. For a proportion of histological subtypes, our IRs are lower than that of other registries. We have previously explained that this difference mainly concerned patients who do not have antitumor treatment. Therefore, histological population-based studies are essential to validate prognostic factors or to study oncological care management on tumors that need histological confirmation before treatment, but are not exhaustive enough to determine the exact incidence of all PCNST. This bias is more pronounced for non-malignant tumors.

Another weakness of the FBTDB is the absence of a systematic pathological review for all the PCNST. In France, a pathological review is performed for patients included in clinical trials or in a national biological database [36, 37], or when the diagnostic is considered as difficult by the pathologist. Of course, in these cases, the FBTDB includes the pathological review in the final diagnosis. Recently, the French pathologists and clinicians, in accordance with the French National Cancer Institute, have decided to perform a pathologic review for all rare PCNST, as part of a national project entitled “TUCERA”. But it will take time and money before it is done in a systematic manner for all PCNST in France.

The analysis of molecular markers is increasingly performed in France for the PCNST, and is now part of the diagnosis for some histological subtypes (i.e., diffuse astrocytomas, oligodendrogliomas) in the recently published new WHO classification. With the development of these markers and personalized oncology, we can hypothesize that both the pathological and molecular analysis of the tumor will soon become mandatory before any treatment for most PCNST. Therefore, the FBTDB will now imperatively need to collect these molecular markers as well as the new integrated diagnosis [3]. However, the current data based on the 2007 WHO classification will serve as a reference for future epidemiological studies using the new classification.

Perspectives

In the field of personalized oncology, population-based studies will need to collect not only the histological diagnosis data, but also molecular, clinical, imaging, quality of life, therapeutic and outcome data to evaluate the best medical strategies at the population level. This is especially crucial for rare tumors, as for these, clinical trials are very difficult to conduct. We believe that this challenging objective will be made possible in the near future thanks to the rapid expansion of health care information systems and the improvement of computer interface systems and health information technologies.

Conclusion

This study is the first to detail all histological subtypes of PCNST, including all rare tumors, and their IRs, at a national level. Above all, this work shows the importance of multidisciplinary networks and databases involving clinicians, pathologists and epidemiologists. Histological population-based studies have many perspectives in the field of clinical epidemiology and are complementary to brain tumor registries.

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