Childs Nerv Syst (2006) 22: 95–98
DOI 10.1007/s00381-004-1092-2

CASE REPORT

Jacopo Lenzi
Maurizio Salvati
Alessandro Frati
Antonino Raco
Angelo Pichierri
Felice Giangaspero
Roberto Delfini

Intraventricular neurocytoma with massive brain stem involvement in a 5-year-old child

Received: 20 May 2004
Published online: 31 March 2005

J. Lenzi · A. Raco · A. Pichierri · R. Delfini
Department of Neurosurgery,
University La Sapienza,
Rome, Italy

J. Lenzi (✉)
Via dei Fienili 58a,
00186 Rome, Italy
e-mail: Jacopole@yahoo.it
Tel.: +39-06-49979119
Fax: +39-06-49979118

M. Salvati · A. Frati · F. Giangaspero
Department of Neurosurgery,
INM Neuromed IRCCs,
Pozzilli (IS), Italy

Abstract *Introduction:* Central neurocytoma is a tumor of neuronal origin that should be taken into consideration in the differential diagnosis of intraventricular neoplasms. Reports of neurocytomas with an extraventricular localization are rare: to our knowledge, the case described here is the first in which a neurocytoma developed within the ventricles but also invaded the brain stem. *Case report:* The authors describe the unusual case of a 5-year-old boy with an intraventricular neurocytoma presenting with massive involvement of the basal nuclei and the brain stem. The patient underwent first biopsy and then surgery for ventricular-peritoneal shunting and partial removal of the tumor. Histology showed the tumor to be a typical neurocytoma with Mib-1 <2%. The postoperative course was uneventful. At 3 years' follow-up, the patient's clinical condition is stable and there are no signs of disease progression. *Discussion:* The literature is reviewed and the characteristics of this unusual tumor are discussed.

Keywords Neurocytoma · Pediatric · Extraventricular involvement

Introduction

Central neurocytoma is a tumor of neuronal origin that should be included in the differential diagnosis of intraventricular neoplasms, together with choroid plexus papillomas, ependymomas, oligodendrogliomas, subependymal giant cell astrocytomas, intraventricular meningiomas, and astrocytomas [1–6].

Central neurocytomas were described for the first time by Hassoun et al. [8] and by the early 1990s had become a well-defined clinical–pathological entity.

In the literature, sites other than ventricular are described [7, 8] as well as involvement of paraventricular structures such as the thalamus.

The peculiarity of the case described here lies in the massive involvement of the extraventricular structures by the lesion and the very young age of the patient.

Case report

A 5-year-old boy was referred to us after his parents had been informed by his teachers that they had noticed differences in the functioning and size between the two sides of his body: he showed difficulty coordinating bimanually, predominantly using his right hand, and had an unstable gait. In his anamnesis, pyscho-motor development appeared normal, but his parents mentioned that he had begun to experience alterations of his sleep/wake rhythm at about 3.5 years of age. At the same time he stopped talking and his social behavior displayed modifications. Physical examination showed a mild left hemiparesis, an underdeveloped left upper limb, hyposthenia, and hypotonia of the left arm; when running he appeared clumsy and this spastic aspect was accentuated. The child was lively but uncooperative, and seemed to have problems

understanding our instructions. He also stuttered and had dyslalia, with varying substitutions.

An MRI showed a neoformation with polylobulated morphology (5×8×6.5 cm) situated in a diencephalic and mesencephalic site, in a right median–paramedian location. The mass involved the brain stem, which appeared swollen, the gray nuclei on the right and the striate body on the left, with initial involvement of the left thalamus. It englobed the optic chiasm the vessels of the polygon, and the middle cerebral arteries, prevalently on the right side. It also involved the third ventricle, compressed and lifted up the right frontal horn and infiltrated the anterior ventricular carrefour producing bi-ventricular hypertensive hydrocephalus (Fig. 1).

The mass was hypointense on T1-weighted sequences, hyperintense on T2-weighted images, and did not show enhancement after contrast medium had been administered. Electroencephalography was normal, whereas visual evoked potentials showed significant latency alterations.

The child was subjected to a surgical biopsy under general anesthesia via a burr-hole with the aid of the Neuronavigator to identify the intraventricular target (Mach 3.0 StealthStation 1999, Medtronic, Minneapolis, MN, USA).

Histological diagnosis confirmed a neurocytoma with Mib-1 <1%. Owing to his stationary clinical status, we decided not to subject the patient to further treatment for the time being.

However, 2 weeks after being discharged, the child was re-admitted: he was drowsy, difficult to wake, and presented signs of intracranial hypertension. An emergency brain CT scan was performed, which documented bi-ventricular hydrocephalus. Surgery was performed to position a V-P shunt. This was followed a few days later by another operation to partially remove the lesion via a frontal transcortical approach with the aid of the Neuronavigator. The lesion was soft and, in places, easy to aspirate; it did not bleed a lot but infiltrated and appeared to originate from the basal ganglia. Once the edges of the ventricular cavity had been identified by the navigator, the lack of a clear tumoral plane prompted us to interrupt surgical removal.

Histological diagnosis confirmed that the lesion was a neurocytoma and altered the Mib-1 to 2%. Postoperatively, there were no complications and the only clinical sign was the pre-existing left hemiparesis. Control MRI performed 1 month after operation demonstrated the remains of the partial removal and a considerable reduction in the amount of pathological tissue inside the ventricle on the right side, especially the posterior and superior portions. There was signal enhancement after gadolinium from the walls of the surgical cavity, at the level of the anterior portion of the mass next to the frontal horn of the right lateral ventricle. Infiltration of the pons on the right was unchanged.

At present, the child is being monitored by serial clinical and radiological controls and physiotherapy has achieved a slight reduction in the left hemiparesis. Gadolinium-enhanced MRI performed 3 years after the first one has shown that there is no progression of the disease and the clinical picture is stationary.

Discussion

Central neurocytomas were described for the first time in 1982 by Hassoun et al. [8] and by the early 1990s had become a well-defined clinical–pathological entity. This tumor localizes in the median region, generally within the cerebral ventricles [18]. Ultrastructural examination confirms a clear neuronal origin. Until a few years ago, this tumor was considered to have a favorable prognosis. It accounts for 0.25–0.5% of all brain tumors [2, 3] and its peak incidence is between 20 and 40 years of age, with no gender preference.

Hassoun et al. [8], who were the first to describe this tumor, suggested that it originated from the cells of the septum pellucidum. Later, the cells of the subependymal

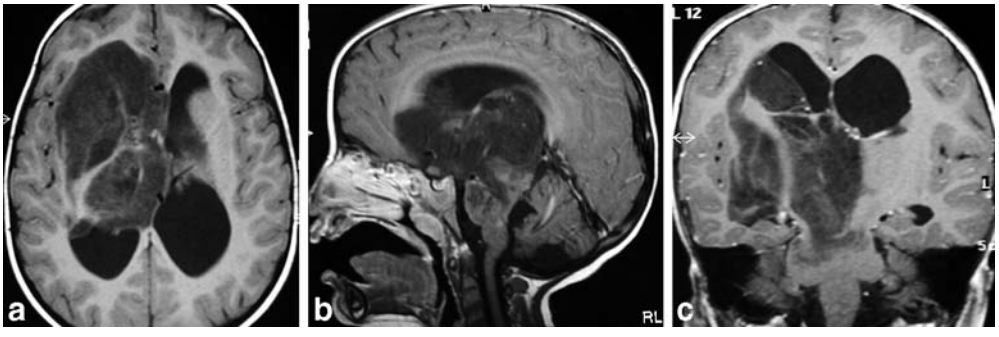

Fig. 1 An **a** axial, **b** sagittal, and **c** coronal T1-weighted MRI scan showing a hypointense neoformation with polylobulated morphology (5×8×6.5 cm) situated in a diencephalic and mesencephalic site, in a right median–paramedian location. The mass involves the brain stem, which appears swollen, the gray nuclei on the *right* and the striate body on the *left*, with initial involvement of the left thalamus. It also involves the third ventricle, compresses and lifts up the right frontal horn, and infiltrates the anterior ventricular carrefour producing bi-ventricular hypertensive hydrocephalus

zone were also considered to be possible cells of origin in clinical and experimental studies [4]. Yasargil and coworkers [17] suggested that this tumor could originate from a totipotent parent cell present in the periventricular matrix that persists in the brain parenchyma of mammals throughout their adult life. Kim and coworkers [9] took into consideration the angiographic features of 17 cases of central neurocytoma and suggested that they originate from a neuronal cell mass of the subependymal zone lying in the floor of the lateral ventricle near the foramen of Monro. In fact, in the literature, a case of intraventricular neurocytoma with a thalamic origin has been described [15]. In the case described here it was not possible to evaluate the true origin of the tumor, which presented clear signs of paraventricular infiltration.

Extraventricular neurocytomas (EVN) are mentioned, but not formally listed, in the 2000 WHO Classification of Tumors of the Nervous System. Nonetheless, reports of such lesions are on the increase and recent studies have better defined their clinical–pathological features [9, 11].

Our case demonstrated the histological features common to central neurocytomas, but with some aspects of differentiation peculiar to extraventricular lesions. Neurocytes demonstrated finely granular, slightly eosinophilic cytoplasm and cytoplasmic clearing, which, combined with round nuclei, suggest a diagnosis of oligodendroglioma. They displayed astrocytic, typically pilocytic features and/or ganglionic differentiation. Focal glio-fibrillary acid protein (GFAP) reactivity is present in tumor cells with neurocytic features [11].

As far as their biological behavior is concerned, similar to central neurocytomas, most extraventricular neurocytomas (EVNs) are well differentiated and do not recur, especially after total resection. However, one third of EVNs do recur within a relatively short follow-up period. Subtotal resection, high proliferation rates, atypical histological features, and older patient age appear to be associated with an increased likelihood of recurrence [11].

The peculiar aspect of our case consisted of the intraventricular development of the lesion accompanied by massive infiltration of the extraventricular structures such as the thalamus and, above all, the basal nuclei and the brain stem, a unique finding in a neurocytoma.

The primary aim of treatment for intraventricular lesions, whether or not there is extraventricular development, must be achieving a precise diagnosis and obtaining as radical a removal as possible.

Histological examination is very important, not only for identifying any notes of atypia associated with a poorer prognosis in the literature, but also for calculating the Mib-1 index, which indicates the degree of tumor aggressiveness.

Radiotherapy for treatment of neurocytomas has been described in the literature, particularly when there is a residue or a progression of the disease [9, 10]. In our case, owing to the relatively low Mib-1 and the young age of the patient, we decided not to perform adjunctive radiotherapy, leaving this option open in the eventuality of a future progression of the disease.

In our opinion, chemotherapy does not play a significant therapeutic role, with the exception of those rare cases with Mib-1 >4% or when the tumor has spread.

In conclusion, the case described here confirms that the behavior of neurocytomas without notes of atypia and a relatively low Mib-1 is predominantly benign, even when there is neuroradiological evidence of marked infiltration.

References

1. Ashkan K, Casey AT, D'Arrigo C, Harkness WF, Thomas DG (2000) Benign central neurocytoma. Cancer 89:1111–1120
2. Brandes AA, Amist P, Gardiman M, Volpin L, Danieli D, Guglielmi B, Carollo C, Pinna G, Turazzi S, Monfardini S (2000) Chemotherapy in patients with recurrent and progressive central neurocytoma. Cancer 88:169–174
3. Brat DJ, Scheithauer BW, Eberhart CG, Burger PC (2001) Extraventricular neurocytomas. Pathological features and clinical outcome. Am J Surg Pathol 25:1252–1260
4. Cobery ST, Noren G, Friehs GM, Chougule P, Zheng Z, Epstein MH, Taylor W (2001) Gamma knife surgery for treatment of central neurocytomas. Report of four cases. J Neurosurg 94:327–330
5. Dodds D, Nonis J, Mehta M, Rampling R (1997) Central neurocytoma: a clinical study of response to chemotherapy. J Neurooncol 34:279–283
6. Eng DY, DeMonte F, Ginsberg L, Fuller GN, Jaeckle K (1997) Craniospinal dissemination of central neurocytoma. Report of two cases. J Neurosurg 86:547–552
7. Giangaspero F, Cenacchi G, Losi L et al (1997) Extraventricular neoplasm with neurocytoma features. A clinicopathological study of 11 cases. Am J Surg Pathol 21:206–212
8. Hassoun J, Gambrelli D, Grisoli F, Pellet W, Salamon G, Pellissier JF, Toga M (1982) Central neurocytoma. An electron-microscopic study of two cases. Acta Neuropathol (Berl) 56:151–156
9. Kim CY, Paek SH, Kim DG (2003) Linear accelerator radiosurgery for central neurocytoma: a case report. J Neurooncol 61:249–254
10. Rades D, Fehlauer F (2002) Treatment options for central neurocytoma. Neurology 59:1268–1270
11. Rades D, Fehlauer F, Schild S, Lamszus K, Alberti W (2003) [Treatment for central neurocytoma: a meta-analysis based on the data of 358 patients] (in German). Strahlenther Onkol 179:213–218

12. Rades D, Schild SE, Ikezaki K, Fehlauer F (2003) Defining the optimal dose of radiation after incomplete resection of central neurocytomas. Int J Radiat Oncol Biol Phys 55:373–377
13. Salvati M, Cervoni L, Caruso R, Gagliardi FM (1997) Central neurocytoma: clinical features of 8 cases. Neurosurg Rev 20:39–43
14. Sgouros S, Jackowski A, Carey MP (1994) Central neurocytoma without intraventricular extension. Surg Neurol 42:335–339
15. Sgouros S, Walsh AR, Barber P (1994) Central neurocytoma of thalamic origin. Br J Neurosurg 8:373–376
16. Soontornniyomkij V, Schelper RI (1996) Pontine neurocytoma. J Clin Pathol 49:764–765
17. Yasargil MG, Von Ammon K, Von Deimling A, Valavanis A, Wichmann W, Wiestler OD (1992) Central neurocytoma: histopathological variants and therapeutic approaches. J Neurosurg 76:32–37
18. Zentner J, Peiffer J, Roggendorf W, Grote E, Hassler W (1992) Periventricular neurocytoma: a pathological entity. Surg Neurol 38:38–42