Childs Nerv Syst (2009) 25:247–251
DOI 10.1007/s00381-008-0690-9

CASE REPORT

Pediatric primary pilocytic astrocytoma of the cerebellopontine angle: a case report

G. Mirone · L. Schiabello · S. Chibbaro · S. Bouazza · B. George

Received: 5 May 2008 / Revised: 30 June 2008 / Published online: 9 August 2008

Abstract
Introduction We describe a rare case of pediatric pilocytic astrocytoma presented as a right cerebellopontine angle (CPA) mass, completely separated from the brain stem and arising from the proximal VIII nerve portion.
Case report A 12-year-old boy, with no evidence of neurofibromatosis type 2, presented with progressive hearing loss at the right ear and headache. An initial enhanced magnetic resonance examination suggested the diagnosis of schwannoma. The tumor was resected by a suboccipital retrosigmoid approach.
Discussion Our case seems to be the first report of a primary pediatric CPA pylocitic astrocytoma arising from the VIII nerve complex and presenting internal auditory canal enlargement. It represents the third reported case of a primary CPA pilocytic astrocytoma (the second pediatric case with the first arising from V nerve) and the eighth report of primary CPA glioma, overall. We discuss the clinical, neuroradiological, and intraoperative findings, and we review the different hypothesis about the origin of these rare tumors.

Keywords Pylocitic astrocytoma · Schwannoma · Facial nerve · Cerebellopontine angle

G. Mirone · L. Schiabello · S. Chibbaro · S. Bouazza · B. George
Department of Neurosurgery, Lariboisiere University Hospital, Paris, France

G. Mirone (✉)
Service de Neurochirurgie, Hopital Lariboisière,
2 rue Ambroise-Paré 75475,
Paris, France
e-mail: peppemirone@fastwebnet.it

Introduction

Pylocitic astrocytoma accounts for 5% to 10% of all gliomas and is the second most common pediatric brain tumors. It occurs most often during the first and second decades of life, but this kind of tumor most commonly occurs around the third and fourth ventricules, in optic chiasm, and hypothalamus. The literature suggests that all but seven gliomas, of any sort, encountered in the cerebellopontine angle (CPA) have been exophytic extensions of a brain stem glioma [1].

Only two other cases of primary extra-axial pilocytic astrocytoma (PA) of the CPA are reported: Beutler et al [2] reported a single case of a pilocytic astrocytoma arising from the eighth cranial nerve, completely separate from the brain stem. Five additional extra-axial nonpilocytic CPA gliomas have been reported arising from the eighth nerve. All seven, except the last, occurred in adults.

We recently encountered a CPA angle pylocitic astrocytoma arising from the eighth cranial nerve completely separated from the brain stem. Preoperatively, neuroimaging, even if unusual, suggested a schwannoma with internal auditory canal (IAC) enlargement.

Case report

History and examination

A right-handed 12-year-old Caucasian boy was referred to our department with the chief complaint of right ear fluctuating hearing difficulty and mild intermittent morning headaches lasting for 3 years. His medical, surgical, and family histories were all unremarkable.

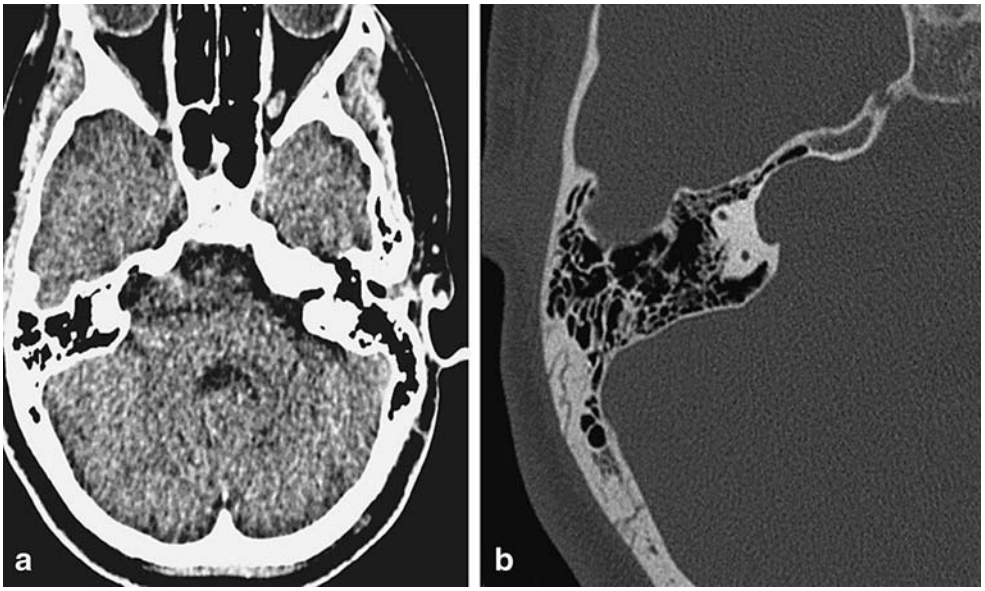

Fig. 1 **a** Unenhanced axial cerebral CT scan of demonstrating a right hypodense CPA tumor (*asterisk*) with IAC enlargement (**b**)

On admission, neurological examination result was negative except for a right ear hypoacousia. He had no other neurological or otological signs and symptoms, particularly no tinnitus, no vertigo, or facial weakness or numbness. On physical examination, *café au lait* spots or other signs of neurofibromatosis were not present. Audiological testing was performed before surgery. Pure-tone audiometry showed a sensorineural hearing loss in the right ear. The auditory brain stem-evoked response on the right side showed no identifiable waves, with normal stimulation parameters. When the stimulus intensity to right ear was raised at 80 db, a V wave at 9 ms was identified. The absence or delay of a V wave indicated a retrocochlear lesion on the right side.

Neuroimaging

Computed tomography (CT) scan showed a nonheterogeneously enhancing mass in the right CPA with, using a bone algorithm, marked internal auditory meatus enlargement (Fig. 1a,b). Head magnetic resonance imaging (MRI) revealed a lesion (2.18×2.23×2.37) with cystic appearance contacting the brain stem. It was hyperintense in FLAIR and hypointense in T2 sequence (Fig. 2c), isointense in T1 peripherically and hypersignal in the center (Fig. 2a). It slightly enhanced heterogeneous after administration of gadolinium–diethylenetriamine penta-acetic acid (Fig. 2b). The brain stem adjacent to the tumor did not reveal any

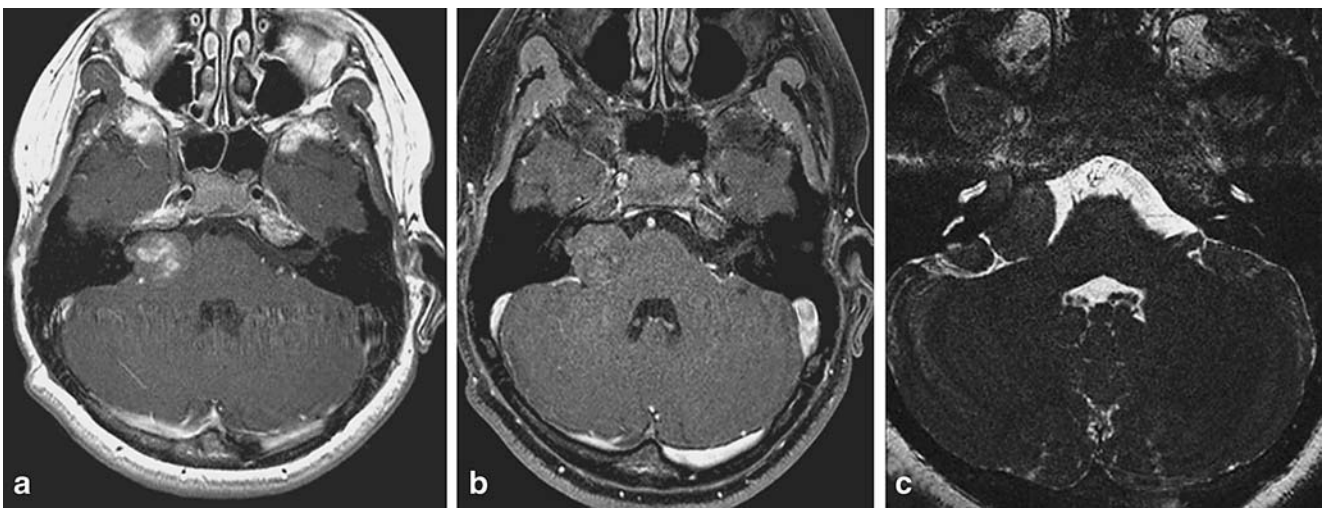

Fig. 2 **a** Noncontrast axial T1-weighted MR image revealing a nonhomogeneously hypointense right CPA tumor. **b** Same image after contrast enhancement revealing a non enhancing tumor. **c** A T2-weighted MR image revealing a hypointense in the right CPA cistern

definite signal alteration. Because of the atypical clinical and neuroradiological findings for a schwannoma and for a differential diagnosis with a vascular lesion, an angiography was carried out, showing only a mild tumor blushing arising from right anterior inferior cerebellar artery (AICA).

Intraoperative findings

Facial nerve electromyography and brain stem auditory-evoked response were utilized for intraoperative monitoring. The tumor was exposed through a right retromastoid suboccipital craniectomy. After gentle medial retraction of the cerebellum, we encountered a gray-white, apparently encapsulated, relatively poorly vascularized mass, abutting the brain stem and adherent to the seventh and eighth cranial nerve complex. At this time, frozen section biopsy surprisingly returned a diagnosis of "schwannoma" despite an unusual macroscopic appearance. A clear arachnoid cleavage plane was found along the interface between tumor and the cerebellum, middle cerebellar peduncle with evident separation from the pons. The anterior inferior cerebellar artery and the brain stem were very well separable from the tumor, but the tumor clearly invaded the area of the eighth cranial nerve starting about 3 mm far from the brain stem. The tumor apparently arose from the eighth cranial nerve, but the cochlear and vestibular nerves were so encased by the tumor and it was decided to sacrifice them. The internal auditory canal was opened with a high-speed drill so that the tumor could be followed laterally. Using a nerve stimulator, facial nerve was identified and, by arachnoid dissection, successfully preserved at the end of the operation. Peace-meal resection was performed, resulting in a gross total excision.

Hystopatological examination

Gross pathological examination showed multiple small pieces of pale, gelatinous tumor. The cells were bipolar elongated spindle-shaped and had small, oval-shaped nuclei in the background of strongly eosinophilic fibrillary glia with many Rosenthal fibers. The blood vessels showed mild hyalinization. These histopathological findings suggested the diagnosis of pylocitic astrocytoma. Immunohistochemistry was performed on paraffin-embedded sections by using the peroxidase–antiperoxidase technique for glial fibrillary acidic protein GFAP and S-100 protein. The tumor was reticulin negative, excluding the diagnosis of schwannoma.

Postoperative course

The patient sustained a transitory mild right facial paralysis but did otherwise well in the postoperative period, and no further treatment was given. He was discharged from the hospital 5 days after surgery. No clinical and neuroradiological evidence of recurrence was noted on follow-up MRIs during the past 6 months (Fig. 3a,b).

Discussion

CPA lesions are more commonly found in adults in which they account for 5–10% of all intracranial tumors. With the exception of patients with neurofibromatosis type II, pediatric extraparenchymal CPA tumors of any sort are extremely uncommon, with an incidence of approximately 0.2–3%. Tumors of glial series are, in general, very

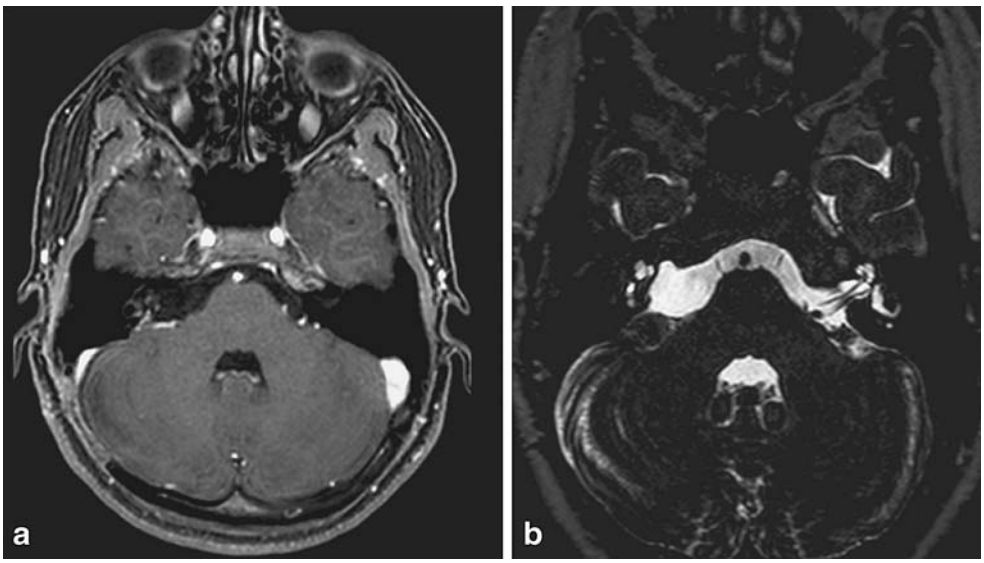

Fig. 3 Postoperative T1-weighted (**a**) and T2-weighted (**b**) contrast-enhanced axial MR image demonstrating gross total tumor removal and relief of brain stem compression

exceptional findings in this anatomic region and are, thus, very rare in pediatric age. A few gliomas presenting as CPA tumors have previously been reported in either children or adults, for most either fibrillary or gemistocytic astrocytomas [5, 7, 9]. Of these cases, the majority appears to be a secondary exophytic extension of a primary brain stem and/or cerebellar tumor mimicking the radiographic picture of an extra-axial lesion. For a few CPA gliomas, however, an exclusively extra-axial location has been documented [4–6]. In our patients, preoperative neuroimaging did not demonstrate an intra-axial component. This finding was confirmed during surgery by the identification of a clear tissue plane between the tumor capsule and the brain stem; the tissue plane was only interrupted at the point where the cochlear and vestibular nerves emerged from the brain stem. We consider that our case was strictly extra-axial CPA pylocitic astrocytoma arising from VIII nerve complex.

Glial tumors of the brain stem, and especially pilocytic astrocytomas in young adults, can manifest as asymmetric expansion of the brain stem that can rarely be pedunculated and exophytic, invading the CPA and even mimicking a vestibular schwannoma by enlarging the porus acusticus. A high index of suspicion for a parenchymal tumor is warranted when evaluating a CPA mass in a child. In this age group, intra-axial lesions are relatively common, and schwannomas and meningiomas are quite rare. Intrinsic lesions of the brainstem and cerebellum typically are associated with more profound neurologic deficits (e.g., hydrocephalus, long tract signs, or rapidly evolving multiple cranial neuropathies) at the time of their diagnosis. It is important for the clinician to make the accurate diagnosis preoperatively, since the operative treatment for a benign extra-axial mass differs from that of a primary brainstem lesion. Decisive to obtain a correct diagnosis is the meticulous assessment of preoperative imaging. Some "red flags" could be: (1) blurring of the margin between the tumor and brainstem or cerebellum; (2) a degree of peritumoral hyperintensity on T2-weighted scans disproportionate to the size of the extra-axial mass; and (3) widening of the lateral recess of the fourth ventricle. Typically, the lack of IAC involvement is a marker for a nonacoustic lesion of the CPA. However, as in our case and other reports [14], the porus acusticus could be penetrated with IAC enlargement. Nevertheless, the diagnosis of such a rare tumor remains a challenge. MR display of the detailed regional anatomy is critical to planning the surgical approach. For this purpose, the 3D CISS sequence proved to be superior to other sequences [15]. This sequence is also very useful for delineating cranial nerve and the borders of the tumor within the cistern.

Gross total resection is the therapeutic target for benign extra-axial masses, such as acoustic neuromas and meningiomas, while surgery for most primary central nervous system lesions is more conservative, including tumor debulking to decompress neurologic structures and to reestablish cerebrospinal fluid pathways. Aggressive removal of an exophytic brainstem parenchymal tumor under the mistaken impression that it is an extra-axial mass may result in devastating neurological injury.

The pathogenesis of primary CPA gliomas is still uncertain. An interesting question raised about their origin: developmental and nondevelopmental theories have been proposed. Proponents of nondevelopmental theories postulate that CPA gliomas could arise from adjacent anatomic structures (as the medial velum of the lateral recess of the fourth ventricle) [7]. The appearance of such astrocytomas in the CPA cisterns should also suggest metastatic deposits. Low-grade gliomas seldom metastasize, and the chance of a pilocytic astrocytoma metastasizing is probably even more exceptional. There are only a few cases: a report by Kocks et al. [8], which described a pilocytic astrocytoma of the optic chiasm spreading by the cerebrospinal fluid to the lumbosacral region and a report by Obana et al. [11], discussing a patient with a pilocytic astrocytoma originating from the hypothalamic region and spreading to the cerebellar tonsil and lumbosacral region. In our case, no evident primary tumor was demonstrated. However, the possibility of an occult primary site in the spinal cord or in the brain should be always considered.

To justify a developmental theory, transformation of heterotopic nests of neuroglial tissue within the leptomeninges could be hypothized. These heterotopias, first described by Wolbach in 1907, consist of a mixture of neurons and glial cells (astrocytes, oligodendrocytes, microglia, ependymal cells). They were found in 1% of a series of 100 consecutive autopsies and in 25% of a series of cases associated with CNS malformations [10, 13]. Primary leptomeningeal glioma should be diagnosed when a glioma is found in the subarachnoid space, when there is no history of intraparenchymal glioma, and no part of the glioma can be detected in central nervous tissue (brain, spine, optic nerve) also postmortem, in a thorough autopsy.

Arnautovic et al. [1] reported a primary cerebellopontine angle glioma arising from the V cranial nerve root entry zone. The root entry zone is the proximal portion of the cranial nerve, where there is the transition zone between central glia and neurilemma. This zone, as reported by different investigators [3, 12], has different lengths for the different cranial nerves excluding the olfactory and optic nerves. Cranial nerves 8 and 5 have the first and second longest root entry zone, respectively, of any cranial nerve. Even along the nerve beyond the root entry zone, isolated "glial islands" have been identified on histological sections of the peripheral segment.

Conclusion

We presented the first pediatric case of pure CPA pilocytic astrocytoma showing IAC enlargement and arising from the VIII cranial nerve. We believe that even if a rare occurrence, primary pylocitic astrocytoma, and generally, gliomas should be kept in mind in the differential diagnosis of CPA lesions with unusual neuroimaging characteristics. Pathogenesis is still to be clarified because of the rarity of the tumor, but the improvement of neuroradiological techniques will help in obtaining a correct preoperative diagnosis for CPA lesions with unusual findings and, possibly, contribute to answer the puzzling question about their origin. Total resection is of paramount importance regarding recurrence of free survival. The efficacy of postoperative radiation for low-grade astrocytoma has been debated, but it would seem that a surgery and periodic follow-up evaluation is the treatment of choice for most patients. However, for primary CPA pylocitic astrocytoma, there are still not an adequate number of cases to determine their progression and a stricter follow-up should be recommended.

References

1. Arnautovic KI, Husain MM, Linskey ME (2000) Cranial nerve root entry zone primary cerebellopontine angle gliomas: a rare and poorly recognized subset of extraparenchymal tumors. J Neuro-Oncol 49:205–212
2. Beutler A, Hsiang J, Moorhouse D, Hansen L, Alksne J (1995) Pilocytic astrocytoma presenting as an extra-axial tumor in the cerebellopontine angle: case report. Neurosurgery 37:125–128
3. Bridger MWM, Farkashidy JF (1980) The distribution of neuroglia and schwann cells in the 8th nerve of man. J Laryngol Otol 94:1353–1362
4. Cushing H (1917) Tumors of the Nervus Acusticus and the Syndrome of the Cerebellopontine Angle. Saunders, Philadelphia
5. Forton G, Verlooy J, Cras P, Parizel P, Van de Heyning P (1992) Problematisch dwarsfluitspel: Een otologisch probleem? Case report van een bijzonder cerebellair astrocytoma. Acta Otorhinolaryngol Belg 46:405–410
6. Kasantikul MD, Palmer JO, Netsky MG, Glasscock ME III, Hays JW (1980) Glioma of the acoustic nerve. Arch Otolaryngol Head Neck Surg 106:456–459
7. Kernohan JW, Woltman HW, Adson AW (1948) Gliomas of the cerebellopontine angle. J Neuropathol Exp Neurol 7:349–367
8. Kocks W, Kalff R, Reinhardt V, Grote W, Hilke J (1989) Spinal metastasis of pilocytic astrocytoma of the chiasma opticum. Childs Nerv Syst 5:118–120
9. Millen SJ, Campbell BH, Meyer GA, Ho KC (1985) Mixed glioma of the cerebellopontine angle. Am J Otol 6:503–507
10. Ng HK, Poon WS (1988) Primary leptomeningeal astrocytoma: Case report. J Neurosurg 88(3):586–589 Mar
11. Obana WG, Cogen PH, Davis RL, Edwards MSB (1991) Metastatic juvenile pilocytic astrocytoma. J Neurosurg 75:972–975
12. Russell DS, Rubinstein LJ (1989) Pathology of Tumors of the Nervous System, 5th edn. Williams & Wilkins, Baltimore, p 106
13. Sceats D, Quisling R, Rhoton A, Ballinger W, Ryan P (1986) Primary leptomeningeal glioma mimicking an acoustic neuroma: case report with review of the literature. Neurosurgery 19:649–654
14. Takada Y, Ohno K, Tamaki M, Hirakawa K (1999) Cerebellopontine angle pilocytic astrocytoma mimicking acoustic schwannoma. Neuroradiology 41(12):949–50 Dec
15. Yousry I, Muacevic A, Olteanu-Nerbe V, Naidich TP, Yousry TA (2004) Exophytic pilocytic astrocytoma of the brain stem in an adult with encasement of the caudal cranial nerve complex (IX-XII): presurgical anatomical neuroimaging using MRI. Eur Radiol 14(7):1169–73 Jul