Letter

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SUPRATENTORIAL INTRAVENTRICULAR SCHWANNOMA OF THE CHOROID PLEXUS

Lucas Perez de Vasconcellos[1], Américo Rubens Leite dos Santos[2], José Carlos Esteves Veiga[3], Igor Schilemann[4], Carmen Lúcia Penteado Lancellotti[5]

Intraventricular tumours represent around 10% of central nervous system tumours[1]. A variety of intraventricular tumours can be found, with the differential diagnosis depending upon the location in the ventricular system[2-4]. In the lateral ventricles, the more common intraventricular tumours are meningiomas, astrocytomas and ependymomas, whereas choroid plexus papillomas and carcinomas, subependymomas and dermoid cysts are rare[3,5].

We report a case of intraventricular schwannoma of the choroid plexus in the lateral ventricle, presenting with headache.

CASE

A 21-year-old female who had been previously healthy with no significant past medical history presented with a constant, dull, aching generalized four-week headache without improvement after conservative treatment. Brain MRI (Fig 1) revealed a solitary 4 cm mass centred on the trigone of the lateral ventricle. The lesion appeared to have a pedicle attached to the choroid plexus of the left lateral ventricle. A diagnosis of choroid plexus papilloma was made due to its aspect and location.

On examination, the patient was awake and alert and normal upon neurological examination; in particular, there were no features of increased intracranial pressure. There was no stigmata of a neurocutaneous syndrome and there was no history in her family of inheritable neurological disorder, such as neurofibromatosis.

She underwent a left parietal craniotomy and transcortical microsurgical excision of the mass. The operatoty findings were of an encapsulated mass arising from the choroid plexus of the left lateral ventricle. There was a good plane around it and a complete resection was achieved with progressive circumferential mobilization around its margins. Postoperatively she awoke without any deficits. She was later discharged on day 7, remains well and returned to normal social activities.

Histological examination of the resected specimen (Fig 2A) was typical of schwannoma surrounded by choroid plexus. There was a tumour composed of neoplastic Schwnan cells, and forming two basic patterns in varying proportion: areas of compact, elongated cells with occasional nuclear palisading (Antoni A pattern) and less cellular, loosely textured cells with indistinct processes and variable lipidization (Antoni B). The tumour cells were

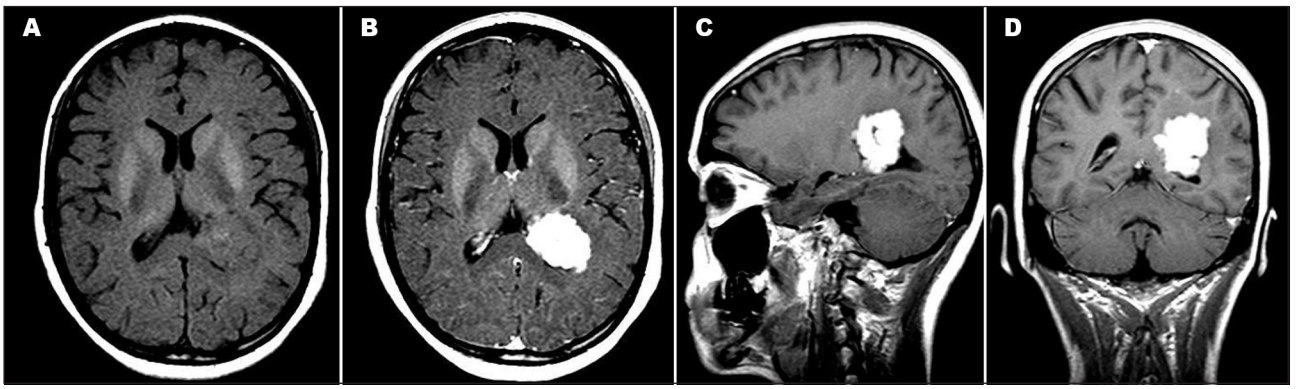

Fig 1. Brain T1-weighted MRI with a solitary 4 cm mass centred on the trigone of the left lateral ventricle. [A] Axial without gadolinium. [B] Axial with gadoliniumand. [C] Sagital with gadolinium. [D] Coronal with gadolinium.

SCHWANNOMA SUPRATENTORIAL INTRAVENTRICULAR DO PLEXO CORÓIDEO

Santa Casa Medical School, São Paulo SP, Brazil: [1]MD, Medical Resident, Discipline of Neurosurgery; [2]MD, Medical Instructor Professor, Discipline of Neurosurgery; [3]MD, Associate Professor, Discipline of Neurosurgery; [4]MD, Medical Resident, Department Pathology; [5]MD, PhD, Full Professor, Department Pathology.

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Dr. Lucas Perez de Vasconcellos – Rua Desembargador Joaquim Barbosa de Almeida 368 - 05463-010 São Paulo SP - Brasil. E-mail: lucas.vasconcellos@terra.com.br

strongly and diffusely expressed with S-100 protein (Fig 2B), and expressed GFAP focally (Fig 2C).

Further management was discussed in the setting of a multidisciplinary group, where no further adjuvant treatment was elected. Serial imaging was performed, and a brain MRI 3 months after resection, showed no evidence of a residual or recurrent tumour.

DISCUSSION

Schwannomas account for approximately 8% of all intracranial neoplasms[6] . Most originate from the vestibular portion of the cranial nerve (CN) VIII. Others arise from the sensory fibers of CN V, VII, IX and XI. Nerve fibers of the central nervous system are not invested with a Schwann cell covering. Therefore, the occurrence of a nerve sheath tumour in central nervous tissue is unexpected[1,2]. There are eleven case reports of ventricular schwannomas[3,7-9]. The majority of these were located in either the lateral or fourth ventricles[4-7]. Most reported intraventricular schwannomas have been benign, with only two demonstrating malignant features[7,8] (Table).

Several theories attempt to explain the origin of intraventricular schwannomas. In 1874, Benedickt[3] identified nerve fibers in the choroid plexus of the fourth ventricle, an observation that was confirmed by Stohr[10] in 1922, suggesting that neoplastic transformation of these Schwann cells could result in an intraventricular schwannoma.

As the autonomic nerves are associated with Schwann cells, this has led to the theory that primary intraventricular schwannomas may arise from the autonomic nervous tissue of the plexus choroids[3-8]. It has also been suggested that intra-axial schwannomas could arise from neural crest cells displaced into the nervous system as a result of failed migration in embryonic life[1]. This disordered embryogenesis theory may account for the relationship between intraventricular schwannomas and other neurocutaneous syndromes such as neurofibromatosis, Hirschprung's disease, and Waardenburg's syndrome[3,6].

Schwannomas are usually solitary, unless associated with a specific genetic syndrome. Most schwannomas have aberrations of cromossome 22. This presumably leads to suppression of the neurofibromin 2 gene product, merlin, located at band 22q12. Genetic syndromes are suggested by the presence of either unusual histologic variants of schwannoma and multiple schwannomas. Plexiform schwannoma and schwannomatosis are characteristic of neurofibromatosis type II[3].

Tumours of the lateral ventricle commonly include meningiomas, ependymomas, choroid plexus papillomas and carcinomas, and astrocytomas. It is difficult to distinguish these from schwannomas based on imaging alone[4].

In summary, intraventricular schwannomas are rare tumours that may be indistinguishable from other benign intraventricular lesions on imaging. When possible, total resection is the treatment of choice, particularly if the lesion is causing mass effect or neurologic symptoms, as was done in this rare case of a supratentorial intraventricular schwannoma arising from the choroid plexus, shown by the histological examination.

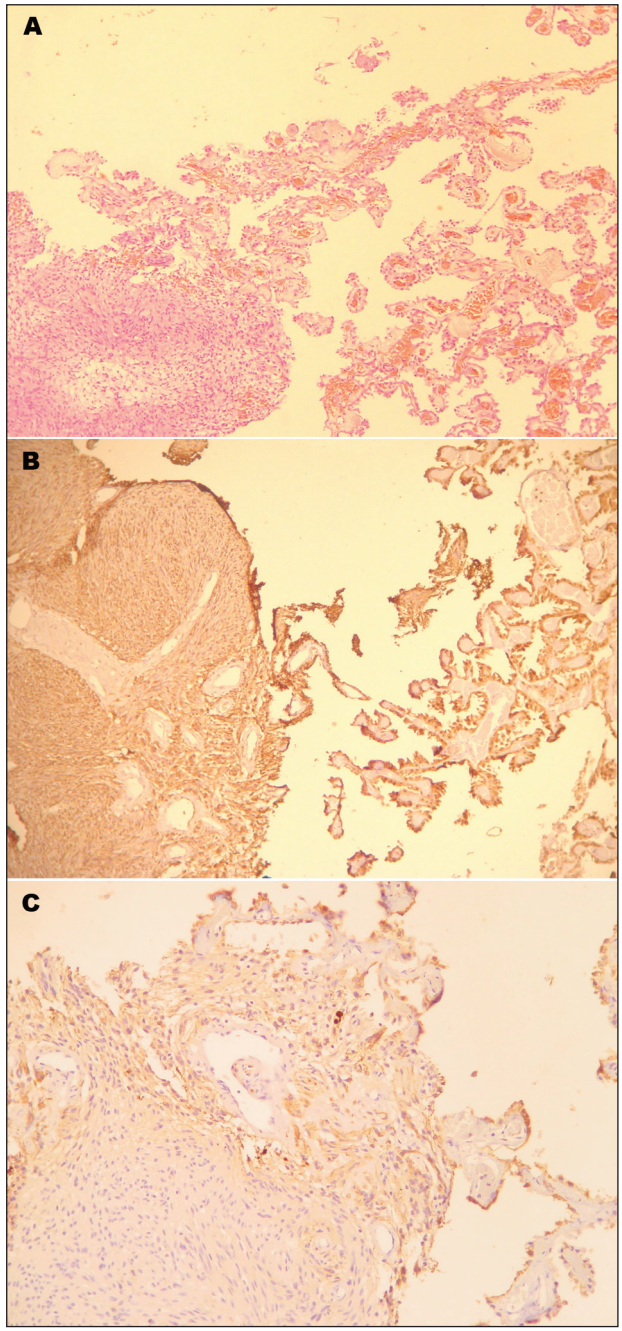

Fig 2. Histological features of the resected specimen. [A] Typical schwannoma surrounded by choroid plexus. Areas of compact, elongated cells with occasional nuclear palisading (Antoni A) and less cellular, loosely textured cells with indistinct processes and variable lipidization (Antoni B) (H&E). [B] Tumour cells stongly and diffusely expressing S-100 protein (S-100). C: Tumour cells focally expressing GFAP and choroid plexus cells stongly and diffusely expressing GFAP (GFAP).

Table. Different cases of intraventricular schwannomas in patients from several studies.

Author/year	Age/sex	Location of tumour	Presentation	Management	Outcome
David 1965 (3)	15/male	Right lateral ventricle	Headache, vomiting and left hemiparesis	Surgical resection	No recurrence at 1 year from surgery
Pimental 1988 (12)	8/male	Right lateral ventricle	Headache, vomiting and left hemiparesis	Surgical resection	No recurrence at 3 years from surgery
Redekop 1990 (14)	7/male	Fourth ventricle	Inward deviation of left eye, rotatory right head tilt, left facial palsy	Subtotal removal	Left ataxia; previous deficits persisted; no evidence of regrowth on CT imaging
Ost 1990 (10)	44/male	Occipital horn of left lateral ventricle	Right homonymous hemianopsia	Surgical resection	Not discussed in paper
Jung 1995 (6)	40/male	Right lateral ventricle	Headache, vomiting, mental status changes	Subtotal removal	Metastases to cerebellum
Barbosa 2001 (1)	8/male	Third ventricle	Headaches	Surgical resection	No recurrence at 6 months from surgery
Erdogan 2003 (5)	21/male	Right lateral ventricle	Left eye visual loss	Surgical resection	No recurrence at 8 years from surgery
Dow 2004 (4)	16/male	Right lateral ventricle	Asymptomatic papilledema	Surgical resection	No recurrence at 1 year from surgery
Messing-Junger 2006 (8)	21/female	Third ventricle	Tinnitus, vertigo, nausea	Surgical resection	Uneventful postoperative course
Benedict 2007	15/male	Right lateral ventricle	Headaches	Surgical resection	No recurrence at 1 year from surgery
Lévêque 2007 (7)	16/male	Right lateral ventricle	Seizures	Surgical resection	No recurrence at 14 months from surgery

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