

Cystic intraventricular schwannoma: case report and review of the literature

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Summary

Intraventricular schwannoma is an exceedingly rare tumour with only 6 cases described in the literature.

One case of a cystic intraventricular schwannoma operated on at our Institution is analyzed and the other cases reported in the literature are reviewed.

Complete removal was achieved and no recurrence was noted after a follow-up period of 10 years.

Intraventricular schwannomas are rare tumours that are amenable to complete surgical removal, having a good prognosis without the need of adjuvant therapy.

KEY WORDS: Intraventricular; Schwannoma; Surgical removal.

Schwannoma quístico intraventricular

Resumen

El schwannoma intraventricular es un tumor extremadamente raro del que se han descrito tan solo seis casos en la literatura.

Se describe un caso de schwannoma intraventricular operado en nuestro servicio y se analizan los casos reportados en la literatura.

En nuestro paciente conseguimos una extirpación total y no hemos observado recidiva después de 10 años de seguimiento.

Los schwannomas intraventriculares son tumores raros cuya extirpación total se asocia a buen pronóstico sin necesidad de terapias coadyuvantes.

PALABRAS CLAVE: Schwannoma. Intraventricular. Cirugía.

Introduction

The occurrence of intracerebral schwannoma is quite

uncommon¹⁷. Only 44 cases have been reported up to date^{2,3,5-8,12,18-20,23}. A schwannoma in an intraventricular location is even rarer, with only 6 cases being described in the literature^{3,5,9,13,15,16}.

We describe a new case in which a cystic schwannoma of the right lateral ventricle was totally removed, with an excellent result, and a review of the literature is done.

Case report

A 13-year-old girl presented in 1990 with a history of chronic headache which lasted for 3 years, accompanied by intermittent sensory focal seizures localized in her left hand.

Physical and neurological examination were unremarkable, with no stigmata of neurofibromatosis.

A CT-scan showed an irregular isodense mass lesion localized in the right ventricular atrium, with a huge cystic component and reactional parenchymal edema. The solid component of the lesion showed a marked enhancement with contrast administration (fig 1).

Magnetic resonante (MR) imaging demonstrated, in T1-weighted images, an iso to slightly hypointense signal mass lesion with a cauliflower appearance, with a mixed

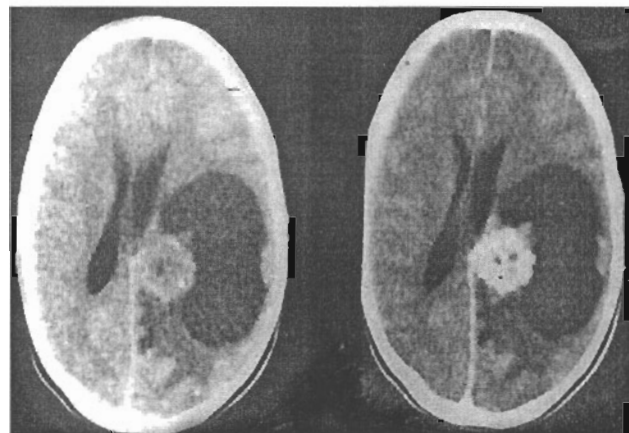


Fig.1- CT scan before and after contrast administration, showing an isodense lesion with a large cyst in the right ventricular atrium, with intense contrast enhancement.

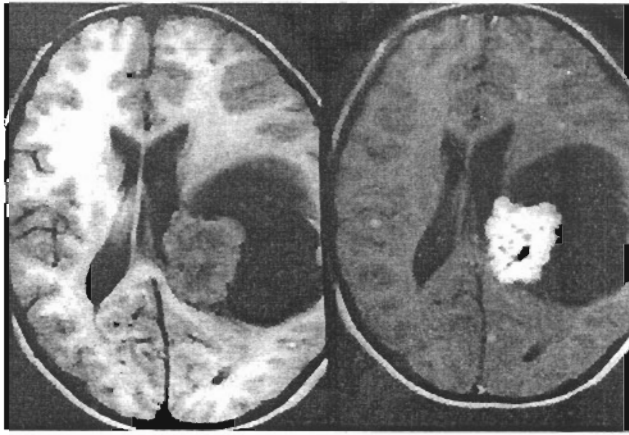


Fig 2- T1 before and after Gadolinium injection, showing the isointense cauliflower-like lesion, an associated huge cyst, and marked contrast uptake.

signal in T2-weighted images, with considerable enhancement after Gadofinim (Gd-DTPA) injection, and an associated cystic component reaching the parietal cortex (fig.2).

A right parietal craniotomy was performed and through a thin transcortical incision the cystic cavity was entered. 60cc of a xanthochromic liquid were aspirated and the solid portion of the tumour lying in the ventricular atrium, irregular in shape, easily bleeding, and attached to the choroid plexus, was totally removed by microsurgical technique, with the help of the ultrasonic aspirator.

An external shunt was left in place and was converted to a ventriculo-peritoneal shunt one week after the first procedure.

Histopathological examination of the operation specimen showed a tumour composed of spindle-shaped cells, initially diagnosed as a fibroblastic meningioma and posteriorly changed to schwannoma, based on immunohistochemical and ultrastructural findings.

The tumour was characterized by a moderate cellularity, a predominant fascicular pattern and extensive pericellular reticulin. The cells were elongated and bipolar, with obscured cell borders and long club-shaped or oval nuclei. There was also some loose textured areas consisting of cells with round nuclei and indistinct cytoplasm where microcysts and xantoma cells were apparent. Some nuclear pleomorphism and hyperchromasia were apparent, but no mitotic figures were identified (fig. 3).

Tumour cells were strongly and uniformly immunoreactive for S-100 protein and vimentin. Extensive areas of glial fibrillary acidic protein (GFAP) reactivity were also present and epithelial membrane antigen (EMA) was negative.

Ultrastructural examination demonstrated fusiform cells rich in cytoplasmic processes, surrounded by a thin basal lamina. Poliribosomes were the predominant cytoplasmic

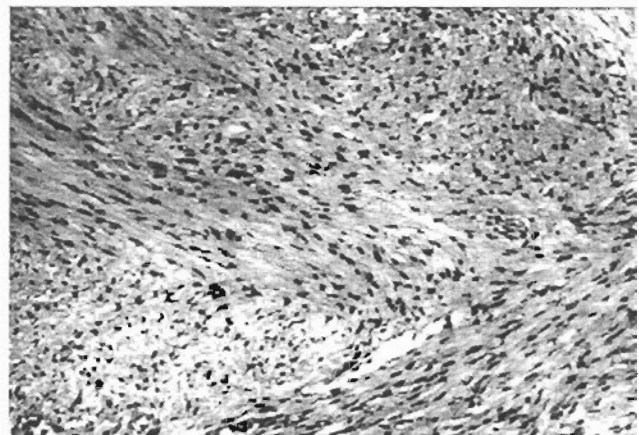


Fig 3- Photomicrograph of an Hematoxylin/Eosin stained slide at 200 x magnification, showing a cellular fascicular pattern of spindle cells with elongated nuclei.

organelles. Luse bodies were not found.

Clinically the patient was well, but an MR study showed a right fronto-parietal sub-dural hematoma.

The shunt was clipped and the hematoma removed. A new CT-scan did not show signs of hydrocephalus and the shunt was then removed.

She has been followed up for over 10 years now, with clinical and neuropsychological evaluation, and she is doing very well, finishing University. The last MR study shows no signal of tumour recurrence (fig 4).

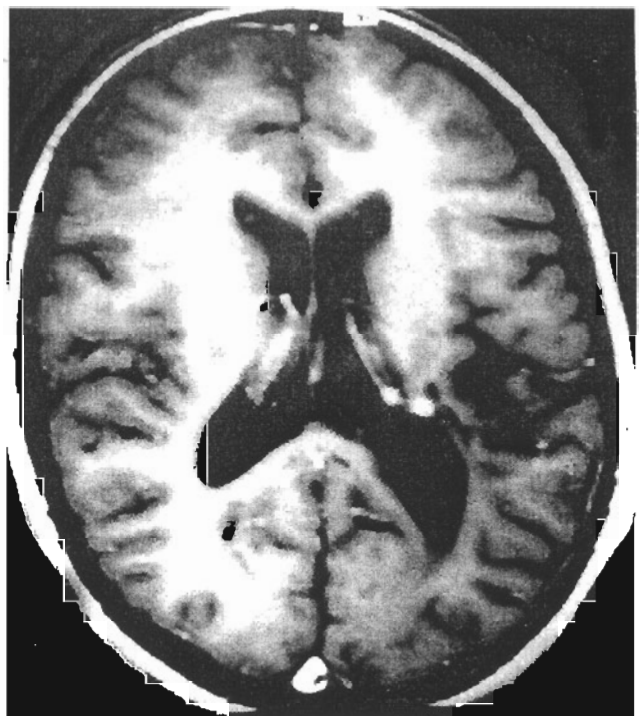


Fig 4. MR ten years after surgery, with no signs of recurrence.

Discussion

Intracranial schwannomas represent 8% of all primary intracranial tumours, and are usually associated with cranial nerves^{6,7,8,12,16,23,24}.

Intraventricular schwannomas are exceedingly rare. They were described for the first time by David³ in 1965. In that very same year Kernohan described as well the first intraparenchymal schwannoma. Since then only 37 more cases of intracranial parenchymal schwannomas^{2,6,7,8,12,13,14,18,19,20,23} and 5 cases of pure intraventricular schwannomas^{5,9,13,15,16} descriptions are found in the literature.

Six of the intraparenchymal schwannomas described have a close relationship with the ventricles, being difficult -from the authors description- to differentiate, at least in some of them, between an intraparenchymal tumour with ventricular extension, or the opposite, i.e., an intraventricular schwannoma with brain parenchyma invasion^{2,7,19,23}.

The origin of intraventricular schwannomas is thought to be from the autonomic nervous system supplying the choroid plexus, which has Schwann cells^{4,9,11,13,15,22}.

Intraventricular schwannomas are not associated with neurofibromatosis, while intraparenchymatous tumours, infrequently, may be a part of a von Recklinghausen disease⁹.

A malignant variant of these schwannomas has been described^{9,20}.

The clinical features of the intraventricular schwannomas, including our case are summarized in Table 1. The intraventricular schwannoma is more frequent in males than in females, with a proportion of 5:2.

It has no age preference - from 7 to 63, with an average of 27 years of age.

Clinically they more frequently present with short duration symptoms and signs of intracranial hypertension, with cranial nerves affected and motor and sensory deficits in some of them. Focal seizures appeared in two cases and in these patients the clinical course was prolonged.

CT scan and MR despite being useful in demonstrating these lesions, fail in showing specific imaging characteristics, as is a rule in intraventricular mass lesions. For the differential diagnosis of intraventricular tumours the age of the patient and the precise location of the mass inside the ventricle is of primordial importance²¹.

Except for the case where the tumour was located in the 4th ventricle¹⁶, all these schwannomas are located in the lateral ventricles -5 in the right & 1 in the left- usually posterior to the foramen of Monro^{3,5,9,13,15}. This may justify the fact that hydrocephalus is not a common feature accompanying these tumours, appearing in just two of the described cases^{3,15}.

The only consistent finding of intraventricular schwannomas on CT scanning is the marked heterogeneous contrast enhancement. The lesion itself may be iso or hyperdense. It is usually irregular and a cystic component is frequent. In some cases there is peripheral edema. Hydrocephalus appears in just 1/3 of the cases and calcification is quite rare^{9,13,15,16}.

MR imaging is the best diagnostic tool in these tumours because it demonstrates the intraventricular position and its relationship with the choroid plexus.

In T1 weighted images the tumour may show iso or slightly hypointensity. In T2 it usually has a mixed signal. Gadolinium (Gd-DPTA) uptake is considerable. These MRI characteristics are similar to the ones found in intraparenchymal schwannomas^{9,13,16}.

Cerebral angiography was done in three of the cases of intraventricular schwannoma, revealing a hypervascular mass fed by the choroidal arteries in one of them⁹, while in the other two the tumours only a faint vascular blush was noted, and vascular displacement was the most relevant feature^{3,5}.

Surgical removal was the treatment of choice in all these tumours, but sometimes a complete removal has not been possible. A transtentorial approach was used in all cases, except for the fourth ventricle one, where a suboccipital craniectomy was performed^{3,5,9,13,15,16}.

The tumour appeared as a firm, irregular but well circumscribed yellow or whitish, easily bleeding mass, attached to the choroid plexus. The cyst may contain clear or xanthochromic fluid^{3,5,9,13,15,16}.

Microscopically these tumours may show areas of high (Antoni A type) or sparse (Antoni B type) cellularity. On the former areas, the cells, arranged in interlacing fascicles, are elongated, with long club-shaped or oval nuclei, occasionally showing palisading and obscured cell borders. On the second areas cells have round condensed nuclei and indistinct cytoplasm with frequent cystic degeneration; xanthoma cells are often seen^{1,3,5,9,13,15,16}.

Immunohistochemical studies are used to confirm the diagnosis, as these tumours could be quite difficult to distinguish from fibroblastic meningioma, if only optic microscopy is used. They show positivity for S-100 protein and vimentin. Glial fibrillary acidic protein staining may be found between cell clusters. Stains for cytokeratin and epithelial membrane antigen are negative^{5,9,13,15,16}.

The diagnosis confirmation may also be done by electron microscopy, which may reveal the so-called Luse bodies, which are extracellular collagen fibrils, and cell processes covered by a basal membrane^{9,15,16}.

In summary, schwannomas in an intraventricular location are rare, usually benign and amenable of successful surgical curative removal, without the need of any adjuvant therapy.

Table 1

Case n° Author	Age (yrs) Sex	Symptoms and signs	Duration	Location	Surgical removal Results	Follow-up period
1 David et al. ³	15 M	Headaches, vomiting, visual and gait disturbances. Bilateral papilledema, Mild left hemiparesis.	4 months	Right Lateral Ventricle	Total Left hemianopsia	?
2 Ghatak Et al. ⁵	63 F	Seizures. Facial paresis, hemiparesis, hyperreflexia, Babinsky sign, sensory inattention defect and homonymous hemianopsia on the left side.	40 years	Right Lateral Ventricle	Total Partial recover	1 year, no reccurrence
3 PimentelM Et al. ¹⁵	8	Headaches, vomiting left-sided weakness. Bilateral papilledema, left hemiparesis	2 months	Right Lateral Ventricle	Total	3 years, no reccurrence
4 Redekop Et al. ¹⁶	7 M	Bilateral oculomotor paresis and nystagmus, left facial palsy, left dysdiadochokinesia.	5 months	IV Ventricle	Subtotal Left-sided ataxia	18 months, no reccurrence
5 Ost And Mayer ¹³	44 M	Occasional finding of a right homonymous hemianopsia	?	Left Lateral Ventricle	Total (?)	?
6 Jung Et al ⁹	40 M	Headaches, vomiting. Drowsiness, bilateral papilledema, retinal haemorrhage. Headaches, vertigo. Confusion.	20 days 7 months	Right Lateral Ventricle Local reccurrence PC angles and cerebellar metastasis.	Subtotal Total (cerebellar and left PC angle)	reccurrence and metastasis 7 months Death 15 days (meningitis and pneumonia)
7 Present case	13 F	Headaches, sensory focal seizures.	3 years	Rigth Lateral Ventricle	Total	10 years, no reccurrence

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