CASE REPORT

Supra and infratentorial ectopic schwannoma mimicking a meningioma

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SUMMARY

We report the case of a supra and infratentorial ectopic schwannoma, mimicking a meningioma, that was surgically resected on a 75-year-old man, with a history of progressive visual loss and headaches. MRI was in favour of an occipital, cystic, tentorial-based meningioma extending to the posterior fossa, with signs of intratumoral haemorrhage. This is a unique case because of the rare type of lesion, in a very atypical location, completely unexpected by the preoperative imaging analysis, and to the best of our knowledge it is the oldest patient reported in the literature with this kind of tumour.

BACKGROUND

Schwannomas correspond to 8% of all primary brain tumours. The great majority of these tumours (80–90%) can be found in the cerebellopontine angle in association with the vestibular nerve.¹

When there is no relation to cranial nerves, intracranial schwannomas of the brain can be considered to be intracranial ectopic schwannomas (IES), which are extremely rare, and often associated with neurofibromatosis type 2 (NF2).²

Since Gibson *et al*³ reported the first case of an intracerebral schwannoma located at the temporal lobe in 1966, about 70 cases have been reported in the English literature.^{2 4 5}

Most of the reported cases of IES are supratentorial in location, either in the cerebral hemispheres or lateral ventricles. To the best of our knowledge, there is no case in the literature of a simultaneous supra-infratentorial location of IES and besides that, this patient is the oldest one described with this type of tumour.

CASE PRESENTATION

A 75-year-old man with no relevant medical history presented with a slowly progressive visual impairment in the last 10 months and a recent 2 months daily headache. No nausea or vomiting was associated.

On admission, neurological examination revealed right homonymous hemianopia. No oculomotor paralysis, diplopia or nystagmus was observed.

No cutaneous stigmata of neurofibromatosis was identified.

INVESTIGATIONS

Brain MRI revealed a left medial occipital multicystic extra-axial mass lesion, traversing the duramater of the tentorium and insinuating into the posterior fossa. After gadolinium administration (intravenously), the lesion had strong peripheral and intralesional

multiseptal contrast-enhancement. There was significant mass effect over the occipital lobe and left cerebellum and also moderate perilesional oedema. No calcifications were observed (figure 1A–C). Visual fields test confirmed right homonymous hemianopia.

DIFFERENTIAL DIAGNOSIS

The image characteristics strongly suggested as the most likely hypothesis of a multicystic supra and infratentorial meningioma.

TREATMENT

The patient underwent an occipital paramedian left craniotomy, and a total microsurgical excision of the lesion was achieved, including its dural base and infratentorial component. The lesion was reddish with a smooth surface and a well-defined plane of cleavage. It was multicavitated, filled with yellow liquid and rests of blood coagulum (figure 2A, B). His postoperative course was uneventful.

Histopathological examination surprisingly revealed the diagnosis of schwannoma, showing compact Antoni A cellular areas mixed with Antoni B paucicellular areas (figure 3). There was also evidence of extensive areas of haemorrhage in reabsorption with intra and extracellular hemosiderin deposits (figure 4). Immunohistochemical examination showed strong positivity for S-100 and collagen IV, variable expression of calretinin and negativity for epithelial Membrane antigen (EMA) and glial fibrillary acidic protein (GFAP) (figure 5).

OUTCOME AND FOLLOW-UP

At 3 months follow-up, the patient had improved vision and a complete remission of headaches. MRI showed total resection of the lesion with no evidence of recurrence (figure 6A–C).

DISCUSSION

Schwannoma tumours with no relation to cranial nerves can be described as IES. These lesions are

Figure 1 T1-weighted axial (A), coronal (B) and sagittal (C) MRI depicts an extra-axial, supra and infratentorial left, multicystic enhancing mass lesion; displacing the occipital lobe and cerebellum. Moderate perilesional oedema is present. No calcifications were observed.

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Figure 2 Macroscopic aspect of the lesion removed with a reddish smooth surface (A) and an inside cystic cavity containing blood clots (B).

Figure 3 Photomicrograph (H&E) showing compact fascicles of fusiform cells, arranged in palisading pattern (Antoni A cellular areas) mixed with loosely arranged cells in a myxoid matrix (Antoni B paucicellular areas).

<1% of surgically treated schwannomas of the central and peripheral nervous system.⁶

About 70 cases of IES have been reported in the English literature,^{2,4,5} and these lesions seem to be mainly supratentorial in location, either superficial or periventricular. More rare locations of ectopic schwannomas have also been described such as third⁷ and fourth⁸ ventricle, intrasellar⁹ and pontomedullary cistern tumours with extension into dorsal spinal cord.¹⁰

This special type of schwannoma, according to the previous literature reports, affects mainly the younger population, presenting essentially below the age of 30.^{11–14} In the biggest series described in the literature, the oldest patient had 72 years-old.²

Our case has some unique characteristics. To the best of our knowledge, it is the first case reported of a simultaneously supra and infratentorial ectopic schwannoma and with signs of intratumoral haemorrhage on MRI. Haemorrhagic areas and hemosiderin deposits in pathological analysis confirm the presentation of this ectopic intracranial schwannoma case with intratumoral haemorrhage. The presence of a dural tail sign pointing more likely towards the diagnosis of a tentorial meningioma is also a rare feature. Additionally, it seems to be the oldest patient (aged 75 years) with this kind of tumour described in the literature.

The most common clinical presentation includes headaches, seizures and focal deficits. In our case, the patient's first sign was hemianopia followed by strong headaches.

CT and MRI are not pathognomonic of IES; however, there are some frequent features, namely cystic formation,

Figure 4 Photomicrograph (H&E) depicting extensive areas of haemorrhage in reabsorption with intra and extracellular hemosiderin deposits. In the inferior margin of the picture, the compact basophilic cells represent the dura mater.

Figure 5 Photomicrograph (immunohistochemical analysis) showing strong positivity for S-100 and collagen IV, variable expression of calretinin and negativity for EMA and GFAP.

Figure 6 Postcontrast T1-weighted axial (A), coronal (B) and sagittal (C) MRI obtained 3-month after surgery: total resection of the lesion, no evidence of residual tumour.

calcifications and peritumoral oedema. Our patient's MRI showed a cystic lesion, but no calcifications or peritumoral oedema. Its tentorial base and the presence of a dural tail lead to a presumed diagnosis of a meningioma, a common mistake, with some similar cases described in the literature.^{11–13} The main differential diagnosis of IES is pilocytic astrocytoma, pleomorphic xanthoastrocytoma, ganglioglioma and meningioma.

Other non-tumour hypothesis such as tuberculomas must also be taken into consideration for differential diagnosis of intracranial schwannomas, as they are very common in particular regions such as South Asia. However, supratentorial tuberculomas are often multiple instead of solitary lesions and are mainly located at the grey–white matter junction.¹⁵

It is often extremely hard to differentiate between fibroblastic meningioma and solitary fibrous tumour (Antoni type A cells predominant schwannoma), and pilomyxoid/pilocytic astrocytoma

(Antoni type B cells predominant schwannoma) based only on histopathological analysis. These have very similar histological features. This way, either electron microscopy or immunohistochemical analysis is essential for the differential diagnosis.¹⁶

As most of the described cases, in our case the tumour had positive staining for S-100, collagen IV and calcitonin, with negative staining for EMA and GFAP. There was no association with NF2.

Regarding the origin of these tumours, there are several proposed theories that can be resumed in two categories, the non-development and the development theories.⁴

Proponents of the non-development theories consider that intracranial schwannomas develop from Schwann cells that are normally present in nearby structures, namely the perivascular nerve plexus,¹⁷ the meningeal branch of the trigeminal or anterior ethmoidal nerves innervating the anterior cranial fossa and olfactory groove,¹⁸ or the *fila olfactoria*.¹⁹

Development theories postulate that the mesenchymal pial cells convert into ectodermal Schwann cells or eventually the neoplasm can grow from aberrant neural crest cells found within the central nervous system.^{1 20}

Some authors believe in a congenital origin for some schwannomas based on the young age of the majority of patients reported in the literature (mainly under 25 years of age) and the tendency of these neoplasms to arise from midline structures.²¹

For IES, surgery should be the first line treatment, as a total removal can be curative and the long-term outcome is favourable. In our case, we could achieve total removal, as it was confirmed by the 3-month postoperative MRI.

Learning points

- ▶ Intracranial schwannomas are very rare lesions, and preoperative diagnosis is almost impossible but it should be considered in the differential diagnosis, especially in dural-based lesions.
- ▶ Diagnosis of intracranial ectopic schwannoma (IES) is only possible with histological and immunohistochemical examinations since imaging is clearly insufficient because there are no constant pathognomonic radiological characteristics.
- ▶ Although several theories have been postulated, IES histogenesis remains to be determined and more molecular and translational research is in order.
- ▶ Complete surgical resection can be curative and associated with good prognosis.

Contributors PRDC had the idea and wrote most of the case report, and JLA and AR contributed to conception and revision of the work.

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