CASE REPORT

Giant frontal intraparenchymal schwannoma: unexpected diagnosis

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Professor Assunta Scuotto,
assunta.scuotto@unina2.it**SUMMARY**

We report the case of an intraparenchymal frontal schwannoma, surgically resected in a middle-aged, otherwise healthy woman. Magnetic resonance imaging was ineffective for the definitive diagnosis and was instead afforded by histology. This case is unique because the rare lesion presented as a *giant* mass; to the best of our knowledge, such a finding has not been previously reported.

BACKGROUND

The described case represents a rare manifestation of an unusual tumour.

Intracranial schwannomas in the adult population account for 8% of all primary brain tumours, most of which involve the VIII cranial nerve.

While schwannomas arising from other cranial nerves are rare and usually associated with Von Recklinghausen's disease, intracranial—intracerebral schwannomas (IS), without any apparent connection to cranial nerves, are exceedingly rare. Approximately, 70 cases¹ of schwannomas unrelated to the cranial nerves have been reported so far in the literature.

CASE PRESENTATION

A 45-year-old woman presented with a long-standing history of depression, unsuccessfully treated by psychologists. She was referred to our department because of the onset of headache and progressive visual impairment in the last 3 months.

On admission, the results of the physical examination were unremarkable. No cutaneous stigmata of neurofibromatosis was observed. Neurological examination revealed no significant findings.

A fundoscopic examination disclosed light papilloedema.

INVESTIGATIONS

Brain magnetic resonance imaging (MRI) detected a frontal giant primarily solid mass without perilesional oedema (figure 1A).

DIFFERENTIAL DIAGNOSIS

On MRI, definitive evidence of neither intra nor extra axial location was found, while signal intensity variations and intense postgadolinium enhancement suggested the presence of a meningioma

TREATMENT

The patient underwent a frontal bilateral craniotomy and total microsurgical excision of the lesion, which was fleshy, highly vascular, with a well-defined plane of cleavage, well suckable.

Postoperative course was uneventful.

Histopathological examination (figure 1B) was consistent with the diagnosis of schwannoma, showing compact Antoni A areas admixed with paucicellular Antoni B areas.

OUTCOME AND FOLLOW-UP

At 1-year follow-up, neurological and fundoscopic examinations were normal, and MRI showed no recurrence (figure 1C).

DISCUSSION

It is an exceedingly rare event for a schwannoma to occur within the neuraxis, with intraspinal location being far more likely than the intracerebral one.

As Schwann cells are not normally present in the central nervous system, the origin of IS remains an

Figure 1 (A) Postcontrast T1-weighted axial magnetic resonance imaging (MRI) depicts a frontal giant, intensely enhancing mass with a small necrotic component (black arrow), displacing the lateral ventricles. No perilesional oedema is present. (B) Photomicrograph showing compact fascicles of fusiform cells with elongated nuclei and indistinct cytoplasmic borders arranged in a palisading pattern (Antoni A areas) admixed with haphazardly oriented loosely arranged cells in a myxoid matrix (Antoni B areas; H&E, $\times 140$). (C) Postcontrast T1-Weighted Fat saturation axial MRI obtained 1 year after surgery: no evidence of residual tumour.

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enigma.² Hypotheses put forward to explain their intracerebral occurrence can be considered under two groups, such as developmental and non-developmental. According to the developmental theory, aberrant Schwann cells in the brain parenchyma may occur due to the transformation of the pial mesodermal cells, or from displaced neural crest cells that form the foci of Schwann cells non-developmental theories base their assumption on the fact that Schwann cells are present within the perivascular nerve plexus and large arteries in the subarachnoid spaces.

IS mostly occur in children and young adults with no gender bias. The most common signs and symptoms include headache, seizures and focal deficits.

Although the neuroradiological characteristics of IS are thought to be of high frequency calcifications, cystic formations and peritumoral oedema,³ these findings are not the rule.

MRI characteristics are variable, with T1 isointensity and hypointensity and mixed T2 isointensity and hyperintensity. They show intense but often inhomogeneous contrast enhancement depending on calcification, cystic and/or necrotic component.

Mild to moderate peritumoral oedema is common with IS.

The best imaging modality is MRI; CT scan is useful to better detect calcification.

The differential diagnosis of IS includes several other neoplasms that may occur in children and young adults: pilocytic astrocytoma, pleomorphic xanthoastrocytoma, gangliogliomas and meningiomas.

It is sometimes very difficult to differentiate between these neoplasms based on the histological examination. Thus, either immunohistochemical or electron microscopic examination could be required for the differential diagnosis.

The treatment of choice for IS is total surgical removal. In fact, these tumours are benign, do not require adjuvant therapy and do not recur when total excision was carried out.

In the presented case, calcifications and perilesional oedema were lacking; only a small necrotic area manifested on the lateral tumour edge.

We did not achieve the diagnosis preoperatively: the intracerebral location was not thought and the schwannoma was not postulated. Microsurgical dissection allowed a total tumour removal with a favourable outcome.

Learning points

- ▶ Although intracerebral schwannomas are rare neoplasms, they should be considered in the differential diagnosis to properly select the treatment.
- ▶ Their neuroradiological features are not systematically distinctive: the differential diagnosis could be a true challenge.
- ▶ Either immunohistochemical or electron microscopic examination could be required in addition to histological study for the differential diagnosis.
- ▶ Surgical excision is curative and long-term prognosis is good.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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