CASE REPORT

Usefulness of T2*-weighted MR sequence for the diagnosis of subfrontal schwannoma

Apport de la séquence T2* pour le diagnostic de schwannome sous-frontal


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KEYWORDS
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Abstract Subfrontal schwannomas are rare tumors that are usually diagnosed during surgery. They are often misdiagnosed as meningioma or esthesioneuroblastoma because of their similar clinical and radiological features. We report a case of schwannoma arising from the floor of the anterior cranial fossa that had radiological features similar to that of meningioma. However, T2*-weighted MR imaging revealed multiple foci of low signal intensities within the tumor related to microbleeds, which suggested a diagnosis of schwannoma that was confirmed by histopathology. This case report demonstrates the usefulness of T2*-weighted sequence in distinguishing meningioma from schwannoma, especially in cases where the tumor has an unusual location.

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MOTS CLÉS
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Introduction

Subfrontal schwannomas are extremely uncommon tumors that are often misdiagnosed as meningioma before surgery. The origin of these tumors is widely debated. There are no characteristic imaging features described as yet for these tumors. We report here on a rare case of subfrontal schwannoma that was correctly diagnosed preoperatively, with subsequent histological confirmation. We also describe the imaging findings of the presence of microbleeds within the tumor.

Case report

A 30-year-old female patient presented with multiple episodes of generalized tonic-clonic seizures, with mild frontal headache, in June 2007. There was no relevant family history and she had no signs of neurofibromatosis. Other than hyposmia, her neurological and general examinations were normal, as were the routine laboratory investigations.

Brain CT scans revealed a large, well-circumscribed, hypodense, extra-axial mass (5.0 × 5.1 cm) in the floor of the anterior cranial fossa that was displacing the falx to the left and splaying both frontal horns of the lateral ventricles. There was erosion of the crista galli. Necrotic areas were evident in the tumor (Fig. 1, a, b). MRI showed a lobulated, heterogeneous, intensely enhancing extra-axial mass displacing the falx and splaying the frontal horns with subsequent mass effect. The tumor appeared hypointense on T1-weighted sequence, and heterogeneously hyperintense on T2-weighted and FLAIR sequence. Focal high signal intensity on T1-weighted imaging was suggestive of intratumoral hemorrhage. The T2*-weighted gradient-echo MRI sequence showed multiple, small hypointensities within the tumor that were presumed to be due to microbleeds as CT revealed no calcifications. Apparent diffusion coefficient (ADC) mapping showed increased tumor diffusion. A thin band of contrast enhancement over the jugum sphenoidal—the so-called ‘dural tail sign’—was present. The adjacent ethmoidal sinuses were normal (Fig. 2a-f).

The preoperative diagnostic possibilities considered were subfrontal schwannoma and olfactory-groove meningioma. The former was considered because of the presence of microbleeds. This finding is common with schwannoma, but extremely rare with meningioma. The patient underwent bifrontal craniotomy and the tumor was found to be moderately vascular, attached to the dura over the cribiform plate. The olfactory nerve was not identified.

Histopathological study of the resected tumor showed Antoni A and Antoni B areas of spindle cells arranged in periodic rows with loosely textured, sparsely cellular, neoplastic cells, confirming the diagnosis of schwannoma (Fig. 3a and b).

Discussion

Schwannomas arise from the Schwann cells of the peripheral nervous system. Schwannomas have been reported in very unusual locations such as the olfactory groove, falx cerebri, sella, intraventricular sites and cerebral parenchyma [14]. Subfrontal schwannomas are rare: a recent review could find only 26 cases in the world literature [1]. These schwannomas arise at an earlier age than most tumors (mean age of 32 years) and affects mostly men. Common initial presentations include headache, seizures and hyposmia or anosmia [1].

Various developmental and non-developmental theories have been proposed to explain the origin of these tumors, which remains enigmatic and largely speculative. Developmental theories include conversion of mesenchymal pial cells into Schwann cells—called ‘schwannosis’—or that they arise from aberrant neural-crest cells within the nervous system [9,14]. Non-developmental theories suggest origin from the Schwann cells in the neural plexus around vessels, in the meningeal branch of the trigeminal nerve, and in the anterior ethmoidal nerve normally innervating the anterior cranial fossa and olfactory groove [9,12]. Olfactory filaments can acquire a Schwann-cell sheath approximately 0.5 mm beyond the olfactory bulb and possible origin of tumor from this has also been postulated [1,8,13].

Schwannomas comprise 6-8% of all brain tumors. They usually appear isodense to hypodense on CT scans, and hyperintense on T2-weighted images with MRI. They show intense heterogeneous contrast enhancement, as cystic degeneration is common. Calcifications and dural tail signs are rare [3,4].

Adachi et al. [1] has summarized the clinical, radiological and intraoperative findings of subfrontal schwannomas in the literature to date. Of the reported cases, most tumors were solid or cystic and showed heterogeneous intense enhancement [1]. Calcifications were reported in two cases of subfrontal schwannoma [1,2]. There were no cases of radiologically detected microbleeds described in subfrontal schwannoma prior to our case. This is probably because T2*-weighted gradient-echo sequence was not used in these cases. Because of its rarity, this tumor was
not considered preoperatively and, in every instance, was diagnosed as meningioma.

In our case, although the location, enhancement pattern and dural tail sign favored the diagnosis of meningioma, the presence of microbleeds prompted the diagnosis of schwannoma in the differential diagnosis, as such a finding has been previously described for vestibular schwannoma [6].

Kuzeyili et al. [7] reported that the incidence of hemorrhage in meningioma is rare—it was found in only two of their cases (1.5%). Calcifications are very common in meningioma and these may be misinterpreted as foci of hemorrhage. Krishnamoorthy et al. [6] reported the presence of microhemorrhages in the majority of vestibular schwannomas (94%) and suggested that this might help to
differentiate it from other cerebellopontine-angle tumors. Acute intratumoral bleeding can occur within acoustic neuromas, possibly due to their high vascularity and fragile vessels [11]. The dural tail sign has been described as specific to meningiomas. A recent study reported the presence of this sign in 58% of meningiomas [10]. However, its occurrence is also reported in other intracranial pathologies such as pituitary adenoma, fungal abscess, lymphoma, neuromas and other tumors and, thus, it is not specific to meningioma [5,10].

Hemangiopericytoma, esthesioneuroblastoma and metastases are the other unusual tumors of the subfrontal region. The presence of flow voids may be a helpful sign in the case of hemangiopericytoma, whereas esthesioneuroblastoma frequently shows paranasal sinus invasion and extensive bone destruction.

Although subfrontal schwannomas are rare, imaging signs such as microbleeds should be considered uncommon in the usual meningioma. The possibility of an unusual tumor such as schwannoma should then be considered in such situations. Our case is unique in that this is the first reported case of a subfrontal schwanna with microhemorrhages. Also, it appears to be the first case to be diagnosed preoperatively and confirmed by histology.

Conclusion

Subfrontal schwannoma is a rare tumor that is often not diagnosed preoperatively. MRI findings such as microbleeds within the tumor should prompt consideration of the diagnosis of schwannoma in the differential diagnosis. T2*-weighted gradient-echo sequence can help in the diagnosis.

References