G. SA (1, 4), F. BONNEVILLE (1), J. POIRIER (2), M. LOPES (3), D. DORMONT (1), J. CHIRAS (1)

(1) Department of Neuroradiology, (2) Department of Pathology, (3) Department of Neurosurgery, Groupe Hospitalier Pitié-Salpêtrière, 43 boulevard de l’Hôpital, 75013 Paris, France. (4) Department of Neuroradiology, Hospital Geral de Santo António, Porto, Portugal.

INTRODUCTION

Solitary fibrous tumour (SFT) is a rare neoplasm that has initially been described in the pleura and subsequently reported in almost every organ [5, 7]. SFT of the meninges is rare [7, 10] and appears as an extra-axial tumour with variable patterns of signal, homogeneity and contrast medium uptake. We present here a unique case of a giant bilobed meningeal SFT which unusually demonstrated two different solid components, resulting, to the best of our knowledge, in the largest meningeal SFT ever described in the literature. MR imaging and pathological features of this rare entity are presented and discussed along with those of other extra-axial tumours.

CASE REPORT

A 42-year-old man presented with a six-month history of progressive headaches and visual impairment. During the last fifteen days, he experienced rapid worsening of his symptoms and complained of severe headaches. Neurological examination was otherwise normal, without motor or sensory deficit.

MRI revealed a voluminous well-circumscribed right temporo-occipital extra-axial tumour, measuring 9×8×6 cm. This lesion clearly demonstrated two distinct solid components based on the signal and enhancement behaviour. The anterior-inferior portion, located immediately above the petrous bone, showed mainly low signal intensity on FLAIR images and isointensity on unenhanced T1-weighted images (figure 1a and 1b) with marked enhancement after gadolinium administration (figures 1d and 1f). On the other hand, the posterior-superior component was heterogeneously hypointense on T1-weighted images, hyperintense on FLAIR (figure 1a and 1c) images and showed moderate contrast enhancement (figure 1d and 1f). No dural tail sign (i.e. no dural enhancement adjacent to the tumour) was observed. This large mass was surrounded by only mild vasogenic oedema but produced an important mass effect on the ipsilateral temporal lobe and cerebral peduncle. Considering the imaging features of this extra-axial tumour, a presumptive diagnosis of atypical meningoima or haemangiopericytoma (HPC) was considered.

At surgery, the tumour was strongly attached to the petrous bone and massive bleeding occurred, hence allowing partial resection of the posterior-superior component of the lesion only. A second procedure was performed six months later, and allowed subtotal removal of the remaining portion...
of the tumour, that was attached to the petrous bone. Post-operative course was uneventful.

The histopathological examination showed a biphasic pattern (figures 2a, 2b and 2c). Several regions of the tumour were hypercellular (figure 2a) and composed of sheets of densely packed small cells with a rounded or ovoid nucleus containing a fine chromatin, inconspicuous nucleoli and a small indistinct and eosinophilic cytoplasm. These cells were embedded in a fine fibrillary collagenous matrix. Blood vessels were surrounded by connective tissue. Other areas of the tumour showed a different pattern, composed of interlacing bundles of spindle cells, in with an elongated nucleus was embedded in a more conspicuous collagenous stroma (figure 2b). Apoptotic nuclei were numerous, but mitotic figures were scarce and necrosis was absent. Immunocytochemistry showed that tumour cells were negative for cytokeratina, EMA, GFAP, PS100 and actin, but a high proportion of cells were positive for CD34 (figure 2) and BCL2. The proliferation rate was low, 3% of the nuclei of the tumour cells were positive with the antibody directed against Ki67. Finally, the histological features of both components of the tumour typically met criteria for solitary fibrous tumour.

**DISCUSSION**

SFT is a rare mesenchymal tumour that was first described in the pleura, by Kemperer and Rabin in 1931 [4]. Since then, it has been increasingly reported with a predilection for intrathoracic locations such as mediastinum or pericardium, but also in almost every other organ [2, 8]. Close to the central nervous system, it can occur in paracranial sites such as the orbit [1], paranasal sinus [11] and spine [5] but SFT is very uncommon in the central nervous system itself [10].
Our case is original in two aspects. First, it is to the best of our knowledge, the largest strictly intracranial SFT ever described. Indeed, Ahn et al. reported a large SFT, with longest diameter of 9 cm too, but it included a significant extracranial component in the orbit [1]. Second, the MR imaging pattern of our case demonstrating two distinct solid parts, with no macrocystic component, is very unusual, and has been reported only once [6]. In the literature, intracranial SFT are globally heterogeneous, like fibrous meningioma or HPC, with possible cysts [7], but a bilobed giant mass with two well defined different solid components, is very unusual. As a matter of fact, SFT, fibrous meningioma and HPC share imaging features, namely extra-axial location, hypointense areas on T2-weighted images, and moderate to strong enhancement on post contrast T1-weighted images, and moderate to strong enhancement on post contrast T1-weighted images. However, most of the HPC are described as hypointense or slightly hyperintense on T1, hyperintense with focal hypointensities on T2, and frequent peripheral or internal serpentine flow voids related to vessels [3]. On the other hand, presence of dural-tail and bone erosion, which have been described only in a small number of SFT [7], are more suggestive of HPC, as well as the localization close to the superior sagittal or transverse sinuses [3, 9]. Our case illustrates that the absence of flow-voids, dural-tail sign, hemorrhagic components and focal necrosis, in an extra-axial mass with distinct solid components, one of them strongly hypointense on T2 and intensely enhancing after gadolinium administration, could be in favour of SFT, but may not be pathognomonic, as evoked by Nawashiro et al. [6]. However, it reinforces that idea because we were able to establish an imaging-histology correlation, supported by the combination of surgery specimens, imaging features on T2 weighted-images and histopathological findings. The strong hypointense areas are composed of interlacing bundles of spindle cells with a more conspicuous collagenous stroma and the hyperintense areas corresponded to hypercellular regions, resembling HPC.

Discrimination of SFT from HPC is especially relevant because it carries significant therapeutic and prognostic implications. HPC is an aggressive tumour that requires additional radiotherapy following surgery because it tends to recur after resection, and occasionally metastasizes extra cranially [3, 10]. On the other hand, SFT is usually a slow growing lesion with rare malignant transformation or extra cranial metastasis [2, 7].

Whereas SFT, HPC and fibrous meningioma share imaging features, reliable distinction of these
three lesions can be made using pathologic immunohistochemical analysis. From the last studies and WHO classification, SFT in the central nervous system emerges as a distinct clinical pathologic entity, although the macroscopic, microscopic and immunohistochemical patterns may demonstrate similarities with both fibrous meningioma and HPC. However, unlike meningioma, SFT shows a prominent reticulin network and the cytoplasm of its cells do not stain with PAS. In addition, strong and diffuse immunohistochemical positivity for CD34 with negative staining for EMA and S100 protein is characteristic of SFT. Although immunostaining for CD34 can be positive in HPC, it is then focal and weak. Finally, the stag horn vascular pattern is usually more extensive and widespread in HPC than in SFT.

In conclusion, our case of giant meningeal SFT emphasizes that differential diagnosis remains difficult between SFT, HPC and fibrous meningioma based on MR imaging only. SFT should however been suspected with gross extra-axial masses that demonstrate multiple solid but different components, especially with areas of marked low T2 signal intensity that strongly enhance after gadolinium administration, and without focal necrosis nor flow voids nor dural-tail signs. Nevertheless, final diagnosis still relies on the histological and immunohistochemical analyses.

RÉFÉRENCES