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Hemangiopericytoma: A Rare Intracranial Tumor Mimicking a Meningioma: Case Report and Review of the Literature

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ABSTRACT

Intracranial hemangiopericytoma is an extra axial tumor often mistaken for meningioma. Clinical symptoms are not specific. On the MRI they are iso-intense or hypo-intense on the T1-weighted sequences, hyperintense on T2 WS with early and intense heterogeneous enhancement. There is sometimes an extension of the contrast to adjacent meninges.The treatment is based on surgical excision and complementary radiotherapy. The evolution is marked by a high rate of recidivism and distant metastasis. We report the case of intracranial hemangiopericytoma in a 55 year old man.

Introduction

Intracranial hemangiopéricytoma (HP) is a rare vascular tumour. The clinical symptomatology is not specific and depends on the location and size of the tumor. The main differential diagnose is meningioma.

Imaging allows in some cases to avoid the diagnosis. Diagnostic certainty remains histological, mainly based on immunocytochemical study. The treatment is based on surgery followed by radiotherapy but the evolution is marked by a high rate of recurrence and metastases [1,2].

Case report

A 55-year-old man, without any particular history, presented to the emergency department for a progressive right hemiplegia associated to headaches. Upon admission, the patient was conscious, well oriented, with a right motor deficit. The cutaneous examination found a left frontal and palpebral cutaneous lesion with exophytic development, related to an orbito palpebral hemangioma present since childhood.

Cerebral CT scans showed an extra-axial left frontal tumor spontaneously hyperdense with a large base of duremerian implantation, with a mass effect, associated with a left orbito palpebral tumor related to an hemangioma (figure 1). The brain MRI showed an extra axial left frontal tumor having heterogeneous signal on T2-weighted sequences containing areas of necrosis hyperintense on T2 WS, enhancing heterogeneously without "dural tail" sign, and presenting a contact with the lower part of the upper longitudinal sinus (figure 2). The radiological aspect evoked either atypical meningioma or intra cranial hemangiopericytoma.

A biopsy with immunohistochemical study confirmed the diagnosis of hemangiopéricytoma. The patient received exclusive radiotherapy. The evolution was marked by a little improvement in the motor deficit.

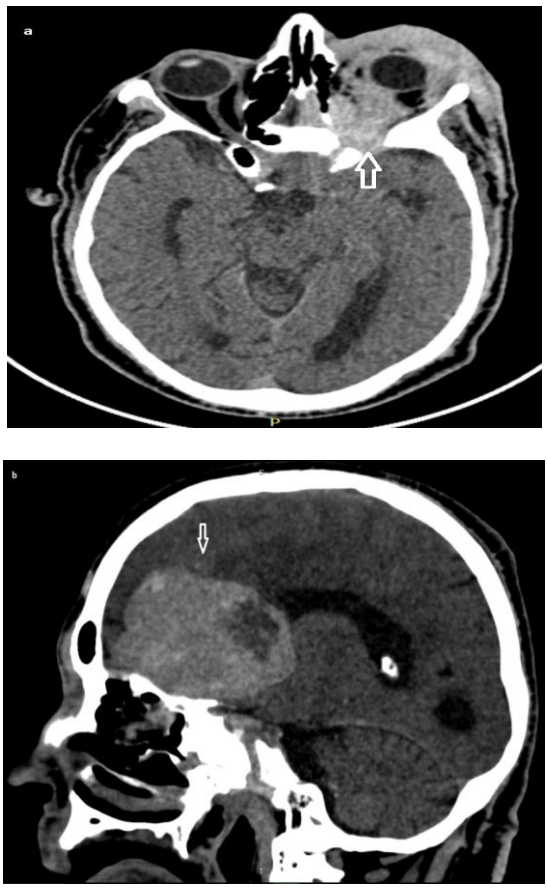

Figure 1. Non-contrast brain computed tomography (CT) scan in axial section (a) and coronal reconstruction (b), showing a spontaneously hyperdense frontal extra axial tumor, heterogeneous with larg base of implantation (b, arrow), associated with a left orbito palpebral tumor (a, arrow).

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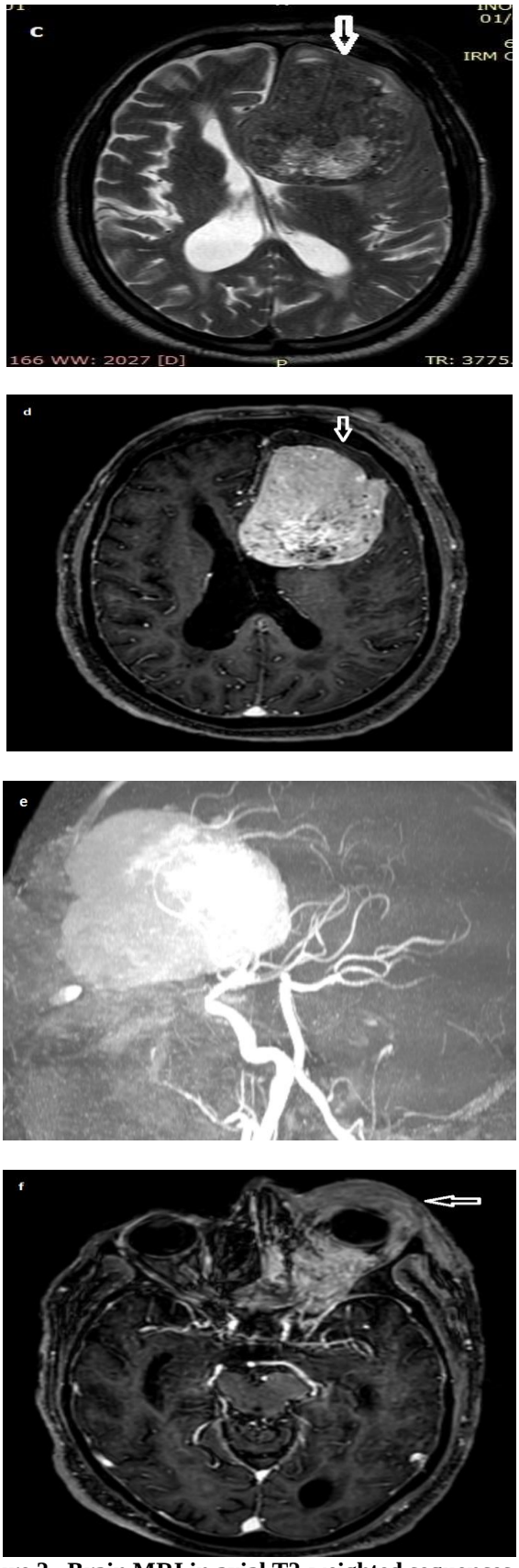

Figure 2 . Brain MRI in axial T2-weighted sequences (c), axial T1 WS after gadolinium injection (d, f) and 3D TOF, showing an extra axial left frontal tumor process in heterogeneous T2 signal with signal-void vessels (arrow, c), enhanced intensively and heterogeneously after gadolinium injection (arrow, d), associated with left palpebral orbito hemangioma enhancing intensively after injection (arrow, f).

Discussion

Intracranial hemangiopéricytoma (HP) is a rare vascular tumour accouting for less than 1% of all intracranial tumours [1, 2].

It was initially classified as a subtype of meningioma known as “meningiomesangioblastic” or “meningioma hemangiopéricytic”. This classification was controversial given the different origin of these two tumors: hemangiopéricytomas originate from the pericytes of the capillaries surrounding the meninges, while meningiomas develop from arachnoid cells [2,3]. Since 1993, the World Health Organization (WHO) has classified hemangiopéricytoma as a "mesenchymal meningothelial malignant tumor"[3].

Macroscopically, HP appears as a solid, generally encapsulated and well-circumscribed mass.Their size is variable. They are richly vascularized, and sometimes contain hemorrhagic, necrotic or cystic areas.

The microscopic aspect of HP is characterized by the presence of massive proliferation of pre-capillary cells Rouget, associated with a very dense reticulin fibers.The clinical symptomatology is not specific and depends on the location and size of the tumor. Supratentorial tumors usually present with headache, vomiting, visual disorder or motor deficit. Posterior fossa tumors may cause ataxia [3,4].

Immunohistochemistry confirms the diagnosis. HP only express mesenchymal markers, the tumour cells are marked by antibodies against CD 34, but are negative for antifactor antibodies VIII. Protein S-100, cellular adhesion proteins and progesterone receptors are never observed. There is an accumulation of basal membrane but there is no desmosome, interdigitation or gap junction [1,4].

Imaging guides the diagnosis. On computed tomography, hemangiopericytoma is generally presented as an extra-axial lobulated mass, spontaneously hyper-dense heterogeneous with a broad or narrow dural attachment rather than classical meningioma, associated with aggressive behaviour such as parenchymal invasion and bone erosion [1,3].

On the MRI they are iso-intense or hypo-intense on the T1-weighted sequences, an hyper signal is found in case of intra tumor hemorrhage. They are hyperintense on T2 WS with early and intense heterogeneous enhancement. There is sometimes an extension of the contrast to adjacent meninges [4,5].

Cerebral angiography is interesting to search the nourishing vessels of the tumor mass and to show the tumor’s limits with the large vascular axes in order to guide the surgical procedure [2,4].

The main differential diagnoses are meningiomas and malignant hemangioendotheliomas.

Meningiomas are more homogeneous than HP with generally homogeneous enhancement. Tumor calcifications are common. Reactive hyperostosis is often present in meningioma and absent in HP which is often associated with bone erosions. The tumor periphery of meningioma is multilobulate exceeding the rounded limits of the tumor giving a «mushrooming aspect [1, 2, 3].

As for malignant hemangioendotheliomas, they are very limited and spontaneously hyperdense at CT. At MRI, they are isointense on T1WS, with heterogeneous signal on T2WS, and sometimes have areas of intratumor hemorrhage [2,5].

The treatment is based on surgical excision and complementary radiotherapy. The evolution is marked by a high rate of recidivism and distant metastasis [2,3].

Conclusion

Hemangiopericytoma is a rare tumor, difficult to diagnose, often mistaken for meningioma. In some cases, imaging can evoke the diagnosis which must be confirmed by histology with immunochemistry study. When the tumor is extra axial, lobular , isointense ,containing signal void vessels and cystic areas, without calcification or dural tail sign, it is probably a hemangiopericytoma.

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