FACIAL INFANTILE HEMANGIOPERICYTOMA RESEMBLING AN ARTERIOVENOUS MALFORMATION

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SUMMARY

Malignant highly vascularized tumors such as hemangiopericytomas (HPC) may mimic a benign arteriovenous malformation (AVM) which is sometimes still referred to as “angioma”. We describe the clinical and radiological findings of a facial hemangiopericytoma in comparison to an AVM in order to avoid misdiagnosis between these two pathologies since evolution and therapeutic management are completely different. Because hemangiopericytomas in children show malignant behavior requiring aggressive management, early and accurate diagnosis is of significant importance for the clinical outcome.

Key words: child, hemangiopericytoma, arterio-venous malformation, vascular tumor.

INTRODUCTION

HPC is a rare (less than 1% of all tumors) soft tissue tumor predominantly involving adults often with a protracted course. Children are affected in approximately 5 to 10% of all cases [3, 8]. The head and neck region is the main site of tumor location (15 to 30%), while involvement of lower limbs, lung, pelvis and visceral organs occurs less frequently. In 10 to 20% of patients metastases are observed at the time of initial diagnosis. Because of its high-flow characteristics, this tumor can be misdiagnosed as a meningioma or an AVM depending on its topography [2]. To distinguish a HPC from an AVM can be of crucial importance because therapeutic management and clinical outcome of both lesions are totally different.

CASE REPORT

A 6-year-old boy was referred to our department with an initial diagnosis of superficial facial AVM. The lesion was present since the age of 3, and had progressively increased in size, altering facial morphology. Clinical examination revealed a large firm throbbing mass underneath normal skin, involving the right cheek with a palpable thrill in the parotid area. Neither the right orbit nor the mouth showed any particular abnormality. On CT-scan, the lesion was hypodense relative to adjacent muscles, with homogeneous enhancement after intra-venous injection of iodinated contrast material. Ultrasonographic evaluation showed a high-flow vascular lesion with arteriovenous shunting. The angiogram disclosed the existence of a tumoral blush. The lesion was supplied by the right transverse facial artery and the right jugal arteries originating from the right internal maxillary artery and the right facial artery. There was early venous drainage, predominantly involving dilated branches of the right facial vein (figures 1a, 1b). MR imaging included axial and coronal spin-echo (SE) T1 and T2-weighted sequences followed by axial and coronal SE T1-weighted sequences after i.v. injection of gadolinium chelates (Dotarem; Laboratoire Guerbet, Aulnay-sous-Bois, France). MRI showed a well defined right jugal mass extending upwards to the skull base through the infra-temporal fossa, downwards to the parotid region, anteriorly towards the greater sphenoid wing, inwards to the pterygoid plate with erosion of its lateral part, and finally out-
wards to the mandibular ramus which was also eroded. The lesion did not extend into the subcutaneous fat. It remained isointense to the adjacent muscles on all SE T1-weighted images (figure 1c) and showed homogeneous enhancement following contrast injection, with serpentine signal voids corresponding to high-flow vessels (figure 1d). On T2-weighted images, the lesion was frankly hyperintense (figure 1e). These MR findings, combined with clinical signs of a high-flow lesion and angiographic findings, were not consistent with the initial diagnosis of AVM. Thus, a tumor was suspected and a surgical biopsy was undertaken. In order to avoid excessive bleeding, selective transarterial embolization using calibrated polyvinyl-alcohol particles (Embospheres; Biosphere Medical Inc, Rockland, USA), was performed prior to biopsy. Pathologic examination disclosed a dense proliferation consisting of round-shaped and fusiform cells disposed around collapsed capillaries (figures 2a, 2b) with only a few mitoses but a rich network after reticulin staining. These findings associated to immunohistochemical staining lead to the final diagnosis of hemangiopericytoma.

**DISCUSSION**

HPCs are highly vascularized potentially malignant tumors of the soft tissues most commonly involving the head and neck region. Published data on HPCs suggest that, although the childhood and adult forms of this tumor are similar in terms of their clinical presentation and pathology, they show significant differences in their natural history and response to treatment [4]. Hemangiopericytomas in children are usually less aggressive and respond more favorably to chemotherapy than in adults. Rodriguez-Galindo et al. [7] reported 12 cases of hemangiopericytoma in children and infants. Three (3/12) of their patients died of disease progression. One patient (1/12) was found to have a HPC of the lung at birth. After surgical tumor resection, a recurrence was treated with chemotherapy. In this
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CASE, treatment with chemotherapy resulted in “maturation” of the HPC to a lesion that was indistinguishable from a capillary hemangioma. Pathologically, infantile hemangiopericytoma demonstrates multilobular growth pattern, branching vasculature, and two types of cells: elongated spindle cells forming fascicles and micronodules, and more primitive, immature perivascular cells with round or oval pale nuclei, and indistinct pale cytoplasm [7, 9]. Long-term follow-up is recommended as local recurrence and distant metastasis may develop even after many years of recurrence-free survival.

Enlarged feeding arteries and draining veins due to intra-tumoral arteriovenous shunts can be found on MR-imaging of hemangiopericytomas. Unlike AVMs, a hemangiopericytoma has an associated parenchymal mass causing a low signal on SE T1-weighted images and a high signal on SE T2-weighted images. Enhancement following Gd-DTPA injection is frequently seen, but usually involves only a small portion of the lesion. Serpentine flow voids may be observed, accounting for high-flow in arterial or venous vessels, and may explain the initial misdiagnosis of AVM in our patient. However, the well-circumscribed mass on MRI was not consistent with a diagnosis of AVM. Selective arteriography can remain inconclusive, because even intra-tumoral fistulae can be found, simulating the appearance of an arteriovenous shunting malformation. CT is essential in assessing the extension of a hemangiopericytoma into surrounding bony structures but is less helpful in the differential diagnosis, because both lesions may erode adjacent bones [3, 9].

AVMs are high-flow vascular anomalies consisting of a nidus or a network of abnormal vascular channels between feeding arteries and draining veins. An AVM is mainly characterized by dilated feeding and draining vessels. These enlarged vessels can usually be identified as signal voids on spin echo MR images. The intervening nidus may or may not be visible, depending upon the size of channels. An AVM sometimes causes signal abnormalities of the tissue that it involves, possibly related to fibrofatty degeneration or edema, which may be evident on MR imaging. Signal of affected tissues may change related to sclerosis or lytic defects [1].

Hemangiomas are the most common tumors occurring in infancy. They are benign vascular tumors resulting from endothelial proliferation at the capillary level. Clinically, most often, hemangiomas grow as well-circumscribed, strawberry-like masses, but infiltrating lesions also develop. Their proliferation begins shortly after birth and can continue up until approximately 1 year of age after which they begin to involute. Most hemangiomas are observed without any specific therapy, while endangering hemangiomas are managed pharmacologically. Deep hemangiomas with normal overlying skin could mimic malignant vascular tumors or AVMs. Doppler US is nonspecific showing numerous vessels with high Doppler shift and low resistance. The MR imaging appearance of hemangioma is characteristic showing a well-defined tumor isointense to muscle on T1-weighted imaging and markedly hyperintense on T2-weighted imaging [6].

In order to avoid any delay in the appropriate therapeutic management of HPC, the definite diagnosis must be obtained as early as possible. To avoid major intra-operative bleeding, a biopsy should be performed after preoperative devascularization of the tumor. A screening for distant metastases is mandatory before any therapy is proposed. Complete surgical resection, if possible, performed after preoperative embolization is the treatment of choice in HPCs. In large invasive HPCs, or when resection remains incomplete, additional chemotherapy and/or radiotherapy should be considered. The treatment of a facial AVM is either surgical, endovascular or a combination of both depending on the size and extent of the lesion. Neither chemotherapy nor radiotherapy is able to stop the progression of a large superficial AVM in the head and neck area [5].

Fig. 2. – Pathologic findings. a) The tumor is composed of rounded and elongated cells, surrounding branched stag-horn like vessels (H & Ex64). b) Reticulin staining shows the characteristic dense reticulin meshwork surrounding vessels and cells (reticulin stain x 50).

REFERENCES


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**Analyse de livre**


L’évolution des connaissances sur la pathologie vasculaire intracrânienne a connu récemment une très importante progression ; cela est largement lié au développement des techniques d’imagerie diagnostique et interventionnelle. L’ouvrage de Michael Forsting comporte cinq chapitres consacrés successivement aux anomalies veineuses de développement, aux cavernomes et télangiectasies, aux malformations artérioïdèmes piales, aux malformations artérioïdèmes durales et aux anévrismes intracraniens. Les chapitres anatomopathologie, clinique, imagerie, thérapeutique en particulier endovasculaire, sont structurés de la même façon.

L'iconographie est à la fois riche, claire, et très adaptée.

Michael Forsting s’est entouré de collaborateurs de renommée internationale pour proposer cet ouvrage indispensable à tout spécialiste, neuroradiologiste confirmé ou non, neurologue neurochirurgien, radiothérapeute et neuropathologiste, désireux d’accéder facilement à une synthèse très actuelle de l’ensemble des connaissances sur les malformations vasculaires intracraniennes.