

An intravascular papillary endothelial hyperplasia of the hand radiologically mimicking a hemangiopericytoma: A case report and literature review

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Abstract

Intravascular papillary endothelial hyperplasia is a rare benign vascular lesion of the skin and subcutaneous tissues, characterized by a reactive proliferation of endothelial cells that can present de novo in normal blood vessels (primary intravascular papillary endothelial hyperplasia), but it can also develop from a pre-existing vascular process (type II intravascular papillary endothelial hyperplasia), or it can arise in an extravascular location from a post-traumatic haematoma. The differential diagnosis between intravascular papillary endothelial hyperplasia and malignant vascular tumours can be challenging, due to the lacking of a specific radiologic description. We present a case of intravascular papillary endothelial hyperplasia of the hand radiologically mimicking a hemangiopericytoma.

Keywords

Intravascular papillary endothelial hyperplasia, Masson's tumour, hemangiopericytoma, hand, musculoskeletal soft tissues, sarcoma

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Introduction

Intravascular papillary endothelial hyperplasia (IPEH), also known as Masson's tumour, is a rare benign vascular lesion of the skin and subcutaneous tissues, characterized by a reactive proliferation of endothelial cells. Hashimoto et al.¹ classified IPEH into three types: type I IPEH presents de novo, in normal blood vessels; type II IPEH can develop from a pre-existing vascular process, that is, a haemangioma, a pyogenic granuloma or a haematoma, while type III IPEH, which is the least common variant, has an extravascular location and generally arises from a post-traumatic haematoma.^{2,3}

The concern of this neoplasm is the differential diagnosis with malignant vascular tumours, that is, hemangiopericytoma, angiosarcoma and Kaposi's sarcoma, to avoid an unnecessary aggressive management, including demolitive surgery and regional radiation.

In this article, we present and discuss an unusual case of an IPEH of the hand, radiologically mimicking a hemangiopericytoma.

Case report

A 61-year-old man was referred to our Department with a 5-month history of a mass at the thenar eminence of the right hand. The patient did not report local antecedent trauma, neither systemic complaints nor recent weight loss. The past medical history and the social history were non-contributory; the patient only reported to drink no more than one glass of

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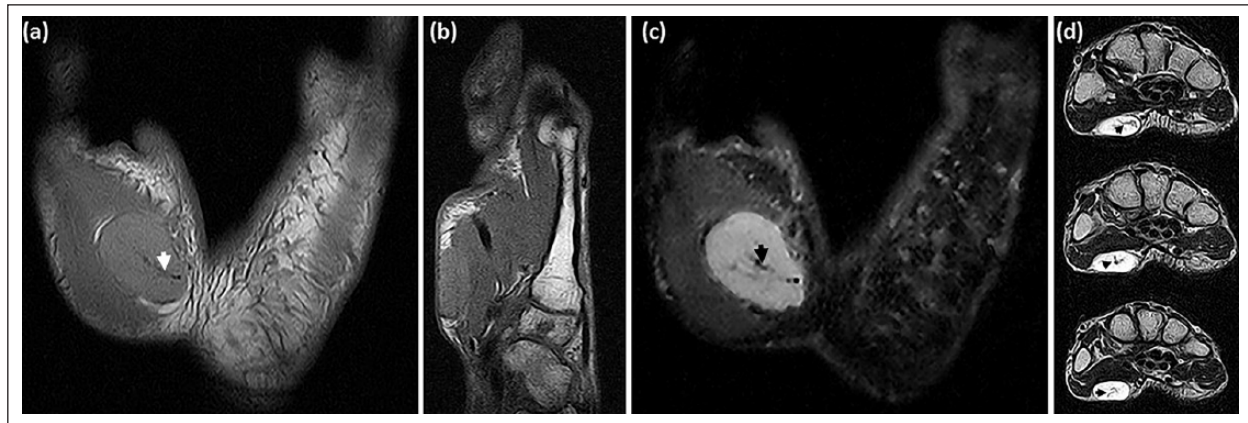


Figure 1. MRI images: (a) coronal T1-weighted image (TR = 680.0 TE = 20.0), (b) sagittal T1-weighted image (TR = 520.0 TE = 20.0). A heterogeneous mass, almost isointense to muscle, with a central marked hypointense area (white arrow) is shown. (c) Coronal STIR image (TR = 1780.0 and TE = 25.0), (d) axial T2 images (TR = 4340.0 TE = 100.0) conducted at three different levels. A heterogeneous hyperintense lesion with an internal low-signal septation, compatible with intralesional calcification (black arrows) is shown.

wine a day. Physical examination of the hand showed a $3 \times 1.5 \text{ cm}^2$ mobile, pulseless, ovular mass in the soft tissues of right thenar eminence. The overlying skin appeared normal. The mass was not painful on palpation, and there were no sensory changes. No regional or axillary adenopathies were observed. Laboratory tests were performed, including routine full blood count, serum biochemistry, inflammatory markers (erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP)), electrolytes, tumour markers, coagulation tests, liver function tests and kidney function tests; no pathological values were observed.

Ultrasound (US) revealed an oval, well defined hypoechogenic $2.8 \times 0.8 \text{ cm}^2$ mass, with intralesional calcifications; the mass appeared confined and encapsulated, and colour Doppler evaluation showed the hypervascular nature of the lesion. The ecographist concluded that the mass was an intravascular tumour of equivocal interpretation, maybe a hemangiopericytoma.

Magnetic resonance imaging (MRI) documented, in the soft tissues of thenar eminence, an ovular, encapsulated $2.7 \times 1.7 \times 0.9 \text{ cm}^3$ mass. The adjacent structures, included the flexor pollicis brevis muscle, were not invaded by the tumour. T1-weighted sequences depicted a heterogeneous mass, almost isointense to muscle, with a central marked hypointense area (Figure 1(a) and (b)). Short T1 inversion recovery (STIR) and T2 images showed a heterogeneous marked hyperintense nodule, without intralesional fat (Figure 1(c) and (d)). Coronal STIR images (Figure 1(c)) revealed a hyperintense lesion with an internal low-signal septation, compatible with intralesional calcification. The radiologist too hypothesized a hemangiopericytoma and suggested the surgical removal of the lesion.

Based on these clinical findings, we decided to perform a percutaneous needle biopsy that diagnosed an IPEH. Consequently, an elective resection of the nodule with histologic examination was executed. A longitudinal palmar

incision centred over the nodule revealed a well-encapsulated, red to bluish, tumour. The specimen sent for pathological examination consisted of a nodular, grey, tense-elastic $1.7 \times 1.2 \times 0.7 \text{ cm}^3$ mass, surrounded by fibroadipose tissue. The section of the nodule revealed a central calcification. Microscopic examination showed an intravascular single layer endothelial proliferation with a papillary architecture, in absence of cytologic atypia (Figure 2(a)–(c)).

At 6-month follow-up, the patient reported a satisfactory aesthetic and functional outcome, without signs of recurrence.

Discussion

IPEH was originally described in 1923 by Masson,⁴ who reported in a 68-year-old man, a papillary endothelium neoplasm associated with thrombosis and fibrin deposits, in the context of a painful, ulcerated haemorrhoidal vein.³ The reactive, rather than neoplastic, nature of this lesion was first recognized 9 years later by Henschen.⁵ The histologic definition of IPEH, coined by Clerkin and Enzinger⁶ in 1976, is currently used to best describe this lesion.

IPEH represents approximately 2%–4% of benign and malignant vascular tumours of the skin and subcutaneous tissues.⁶ It usually occurs most often in adults aged 30–40 years and is slightly most common in woman, just as haemangiomas, suggesting a hormonal factor in development of this kind of lesions.⁷ The most common locations are the hand fingers, followed by the head and neck, although a few cases have been identified at the foot district.² IPEH rarely develops intracranially, where it may present as mass lesion, thus mimicking a malignant neoplasia.⁸ Intracranial IPEH could have a significant morbidity and should be considered in the differential diagnosis for mass lesions in patients that have undergone gamma knife radiosurgery.⁸

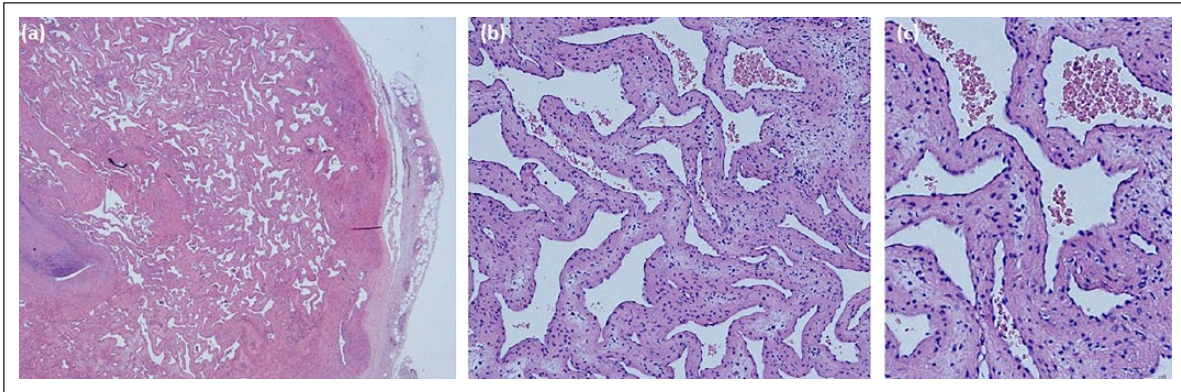


Figure 2. (a) H&E 20x: the section shows a nodular, pseudoencapsulated lesion composed of papillary formations with hyaline or fibrous bundles and (b and c) H&E 100x, H&E 500x: at high magnification, it is possible to observe some vascular formations, with a labyrinthine structure, lined by plump endothelial cells. The underlying fibrous stroma appears oedematous; necrosis, cellular atypias, or atypical mitotic cannot be detected.

Clinically, IPEH presents as a palpable soft-tissue mass, often located within normal or dilated vascular spaces and characterized by slow growth.² An unusual case with rapid growth was recently reported by Corni et al.,⁹ but they did not observe recurrence or complications at 6 months follow-up. Furthermore, though in the recent literature 15% of recurrence rate was noted;⁹ all these cases had a benign natural history.

Although several cases of IPEH have been reported in the literature, its pathogenesis remains unclear. Masson⁴ originally described a reactive process that could be related to a trauma, with a subsequent thrombus organization.¹⁰ Other investigators¹¹ have argued that thrombosis occurs prior to papillary growths, and the following fibrin deposition acts as a substrate for the IPEH development. Levere et al.,¹² however, proposed an autocrine aetiology of post-traumatic IPEH, involving the fibroblast growth factor (FGF) secretion. The macrophages that reach the site of trauma release the FGF, which triggers IPEH; the endothelial proliferating cells, on their turn, release more FGF, thus activating a positive feedback loop of endothelial proliferation.¹²

The case we described shows that although the histological features of IPEH are well characterized, the imaging appearance of this lesion is still uncodified. Consequently, radiologists usually can recognize the vascular nature of the lesion, but they are unable to differentiate IPEH from other malignant vascular neoplasms.

Clifford et al.⁷ in 2004 already noticed that IPEH is not well described in the radiologic literature; they presented a case of IPEH simulating a soft-tissue sarcoma and defined the differential features between IPEH and angiosarcomas. Other studies, moreover, have stressed that the differential diagnosis of IPEH should include Kaposi's sarcoma, angioendothelioma, papular angioplasia, Kimura disease, bacillary angiomatosis, intravenous atypical vascular proliferation and sinusoidal haemangioma (SH).^{2,11} SH is a rare subtype of cavernous haemangioma, which generally develops subcutaneously in

the extremities, predominantly in women.¹³ Histologically, this benign lesion consists of dilated and interconnected thin-walled blood vessels; on cross section, pseudo-papillary structures covered by endothelial cells can be observed, thus resembling IPEH.¹³ The differential diagnosis between SH and IPEH lies in recognizing the truly papillary pattern which is a typical feature of Masson's tumour.

To the Authors' knowledge, however, none of the previous studies reported an IPEH resembling a hemangiopericytoma on US and MRI investigation.

The sonographic appearance of IPEH has been recently defined by Lysy et al.¹⁴ This neoplasm, generally, appears as a well defined echogenic mass, which may be confined but locally extended. It could be found within peripheral veins, thus resembling, at a first feeling, an endovascular thrombus, in the subcutaneous layer of the hand or within a muscle. Colour doppler sonography demonstrates the hyper-vascular nature of this lesion, provided with both venous and arterial flow patterns.¹⁴

The MRI findings of IPEH have been recently reviewed by Clifford et al.⁷ On T1-weighted sequences, the neoplasm may show an hypointense or heterogeneous signal, due to intralesional haemorrhages,^{7,14} on T2 images, IPEH appears hyperintense, with a low-signal internal septation and some hypointense areas reflecting thrombotic or haemorrhagic material.^{10,14} A diffuse enhancement on MRI has been also described.¹⁴

Hemangiopericytoma is a rare vascular tumour, originally described by Stout and Murray¹⁵ in 1942 and currently classified as a subtype of the extrapleural solitary fibrous tumour.¹⁶ It is typically a low-grade benign sarcoma, accounting for less than 2% of soft-tissue sarcomas and 1% of all vascular tumours;¹⁶ it is supposed to originate from perivascular pluripotent mesenchymal cells.¹⁷ Hemangiopericytoma is more frequent in the fifth and sixth decade, in both sexes, and it could be found most commonly in the soft tissues of the upper and lower extremities,

pelvis and retroperitoneal space.¹⁷ It generally presents as a painless mass, covered by normal skin.

On US, hemangiopericytoma appears as a lobulated hypoechogenic mass with frequent intralesional calcifications; colour Doppler sonography usually shows an intense vascularity, with prominent surrounding arterial vessels.¹⁷

MRI typically shows a lobulated lesion with hypointense or isointense signal on T1 sequences and heterogeneous hyperintense signal on T2 and STIR images;¹⁷ contrast-enhanced MRI sequences reveals a large enhancing of the tumour.

Hemangiopericytoma has an unpredictable behaviour, because of its malignant potential and its high recurrence rate; some cases of hemangiopericytoma behaving as high-grade sarcomas are reported.¹⁷ Radical surgical excision is the treatment of choice, while adjuvant chemotherapy and radiation therapy have shown limited success in patients with this malignancy.¹⁷

Currently, the differential diagnosis between IPEH and hemangiopericytoma can be only performed through histological examination, since the imaging can be misleading. It is important to remark the management of these two neoplasms could be different: IPEH can be treated with an elective resection of the nodule, whereas hemangiopericytoma may need a more aggressive surgery due to its unpredictable behaviour.

Conclusion

IPEH is correctly managed with surgical-wide margins resection, to avoid recurrence risk and subsequent pathological examination to confirm the diagnosis of the lesion. This report highlights the need of a radiologic univocal description of IPEH, in order to better recognize preoperatively this benign lesion, avoiding a misdiagnosis that may lead to an unnecessary aggressive management.

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Ethical approval

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Informed consent

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