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Case Report

Intravascular Papillary Endothelial Hyperplasia (Masson's Hemangioma) Presenting as Soft Tissue Mass in Left Scapular Region-An Unusual Presentation

Meenu Gill, Ashima Batra, Deepika Jain, Veena Gupta, Rajeev Sen, Sumiti Gupta

Department of Pathology, Pt. B.D. Sharma PGIMS, Rohtak, India

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Corresponding Author:
Deepika Jain
Department of Pathology,
Pt. B.D. Sharma PGIMS,
Rohtak, India
drdeepika5087@gmil.com

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Abstract

Intravascular papillary endothelial hyperplasia (IPEH) or reactive vascular endothelial lesion is a rare benign condition and is also known as Masson's hemangioma. It may appear as a primary or pure form developing in a distended vessel, or it can be associated with hemangiomas, pyogenic granulomas, or lymphagiomas. It is usually confined to the lumen of preexisting vessels or vascular malformations. Nearly all the lesions are intimately associated with a thrombus in various stages of organization. The main significance of IPEH is its clinical and histological resemblance to soft-tissue sarcoma and possible misinterpretation as such. We hereby report a case of IPEH clinically and radiologically misdiagnosed as soft tissue sarcoma, in a 47-year-old man, presenting as a soft tissue mass in the left scapular region.

INTRODUCTION

Intravascular papillary endothelial hyperplasia (IPEH) is an uncommon vascular lesion and first described within an inflamed hemorrhoidal plexus by Pierre Masson in 1923 as hemangioendotheliomevegetant intravasculaire [1]. IPEH is an unusual benign vascular lesion, comprising approximately 2% of the vascular tumors of the skin and subcutaneous tissue [2]. A clinicopathological study of 91 cases was done by Hashimoto et al in 1983 [3]. They noted that it simulates angiosarcoma because of the presence of papillary formation, anastomosing vascular channels and plump endothelial cells. It is identified because of the exclusive intravascular nature of the process, the lack of necrosis, bizarre cells and atypical mitosis, the characteristically fibrinous and/or hyaline (deeply eosinophilic) appearance of the papillary stalks and the frequent finding of residual organizing thrombi. Today, it is considered to be a reactive vascular proliferation, following traumatic vascular stasis [4]. Essentially, all cases are cured by simple excision. We are reporting a case of IPEH clinically and radiologically mimicking a soft tissue sarcoma.

CASE REPORT

A 47 year-old man was admitted to our hospital, with the complaint of swelling in the left scapular region. The mass had been gradually increasing over the past two months and was painful on movement of arm. There was no remarkable past medical history and his overall health condition was normal. Physical examination revealed a firm, smooth, round and slightly painful mass on the lateral surface of left upper back, densely adherent to the underlying musculature. There was no lymphadenopathy in axillary or cervical region. He had no history of recent trauma or surgical procedure. The patient reported no systemic complaint, no known illness nor continuous drug usage. General physical examination and systemic examination was within normal limits. Routine hematological and biochemical investigations were within normal limits.

Ultrasound (US) performed demonstrated a hypoechoeic collection with a size of 6.5x5.2 cm, located in the left scapular region, extending to the deeper tissues. Primary radiologic diagnosis was a soft tissue sarcoma. A magnetic resonance imaging (MRI) was planned for further evaluation. MRI, performed at

our institution two days later, revealed an isointense mass on T1 weighed images and hyperintense mass on T2 images (Figure 1). The mass showed homogenous enhancement on contrast enhanced magnetic resonance (CEMR). In the light of these findings, a soft tissue sarcoma could not be ruled out. Fine needle aspiration cytology revealed blood only. The mass was excised and sent for histopathology examination.

We received an encapsulated nodular grey brown soft tissue mass, measuring 7x4x2.5 cm (Figure 2a and 2b). External surface was congested. The cut surface was variegated with multiple hemorrhagic and cystic areas (Figure 2b). Histological examination revealed an encapsulated mass mainly composed of large to medium sized vascular spaces, containing papillary proliferations with fibrinous and hyaline stalks, lined by plump reactive endothelial cells (Figure 3a and 3b). Organizing thrombi within anastomosing vascular channels were also evident in some areas (Figure 3d). There was no evidence of necrosis, cytological atypia or atypical mitosis or any solid spindle cell areas even after extensive sampling, excluding a malignant vascular lesion. A diagnosis of IPEH (Masson's hemangioma) of the forearm was made. The lining cells were positive for CD 34, confirming its vascular nature (Figure 3c). Post-operative period was uneventful and the patient is still under follow-up, without any evidence of recurrence.

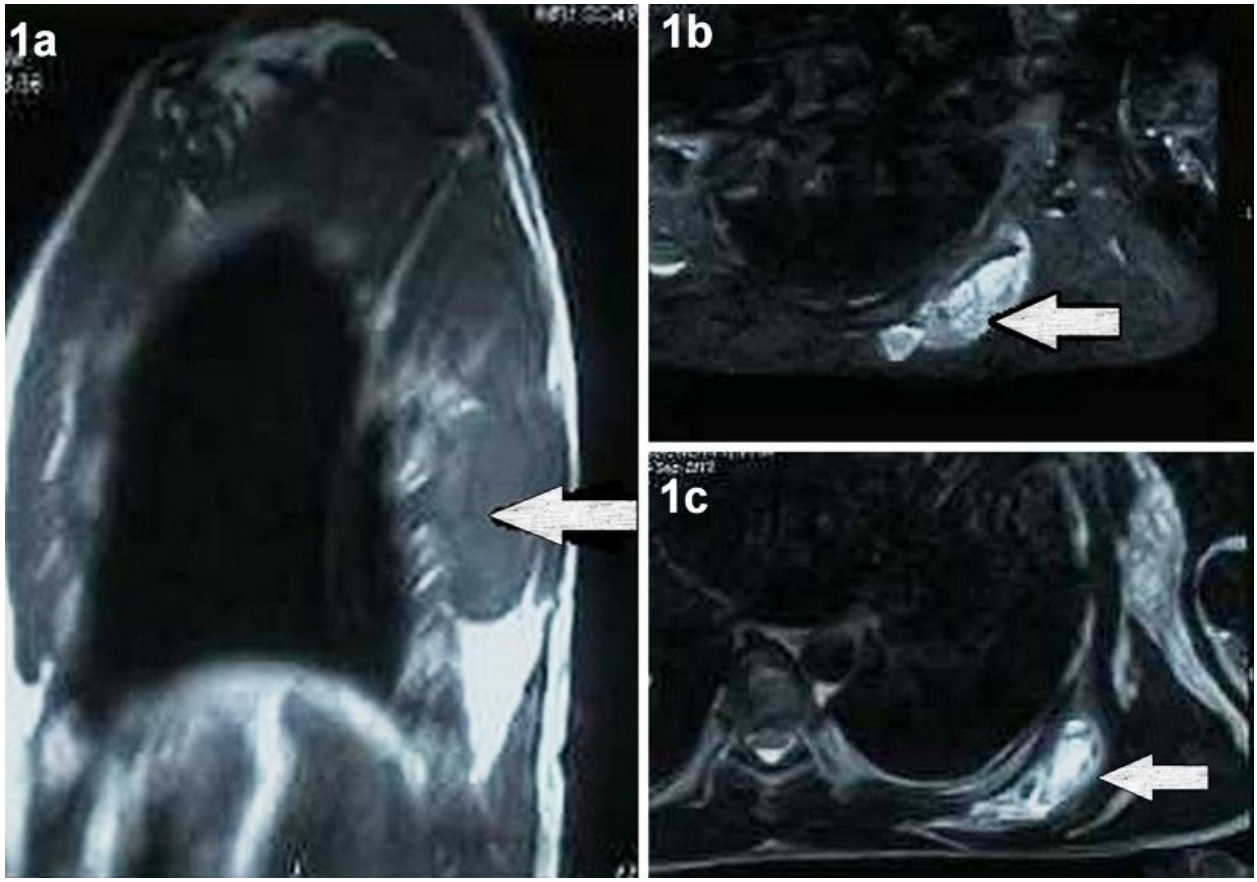

Figure 1: Saggital T1 (1a), axial STIR (Short TI Inversion Recovery) (1b), axial T2 (1c) magnetic resonance imaging showing an isointense mass (marked by arrows) on T1 weighed images and hyperintense mass on T2 images.

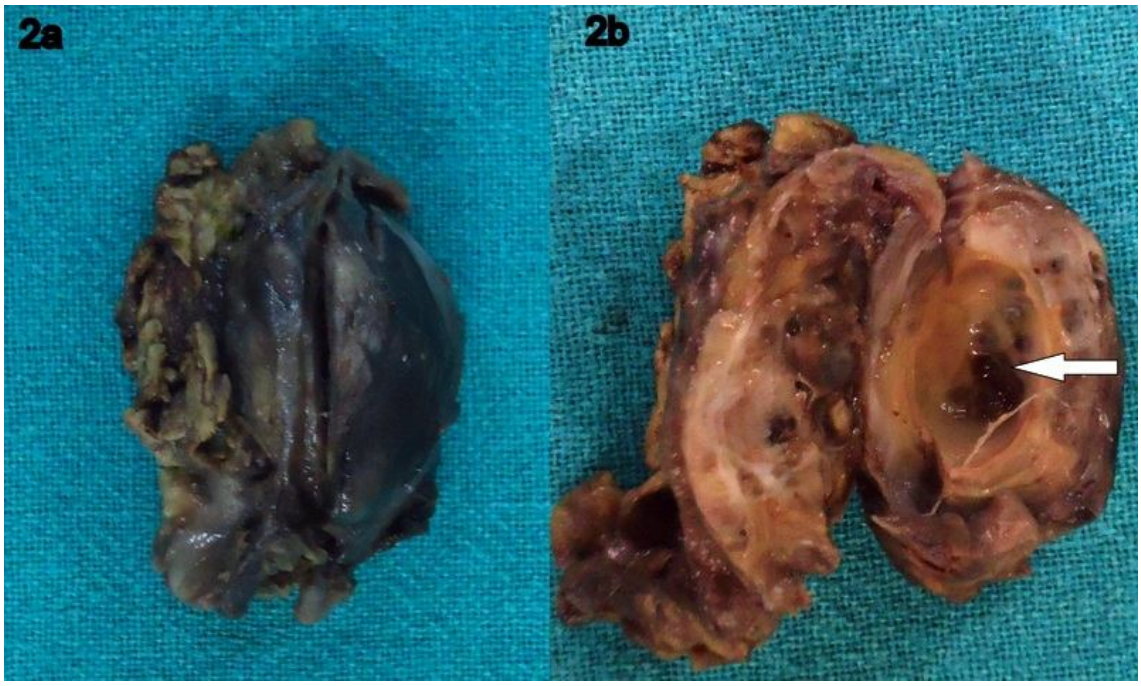

Figure 2: Gross appearance of the mass: **(a)** External surface of one half of the mass showing grey brown smooth surface. **(b)** Cut surface was grey brown with multiple cystic spaces (arrow).

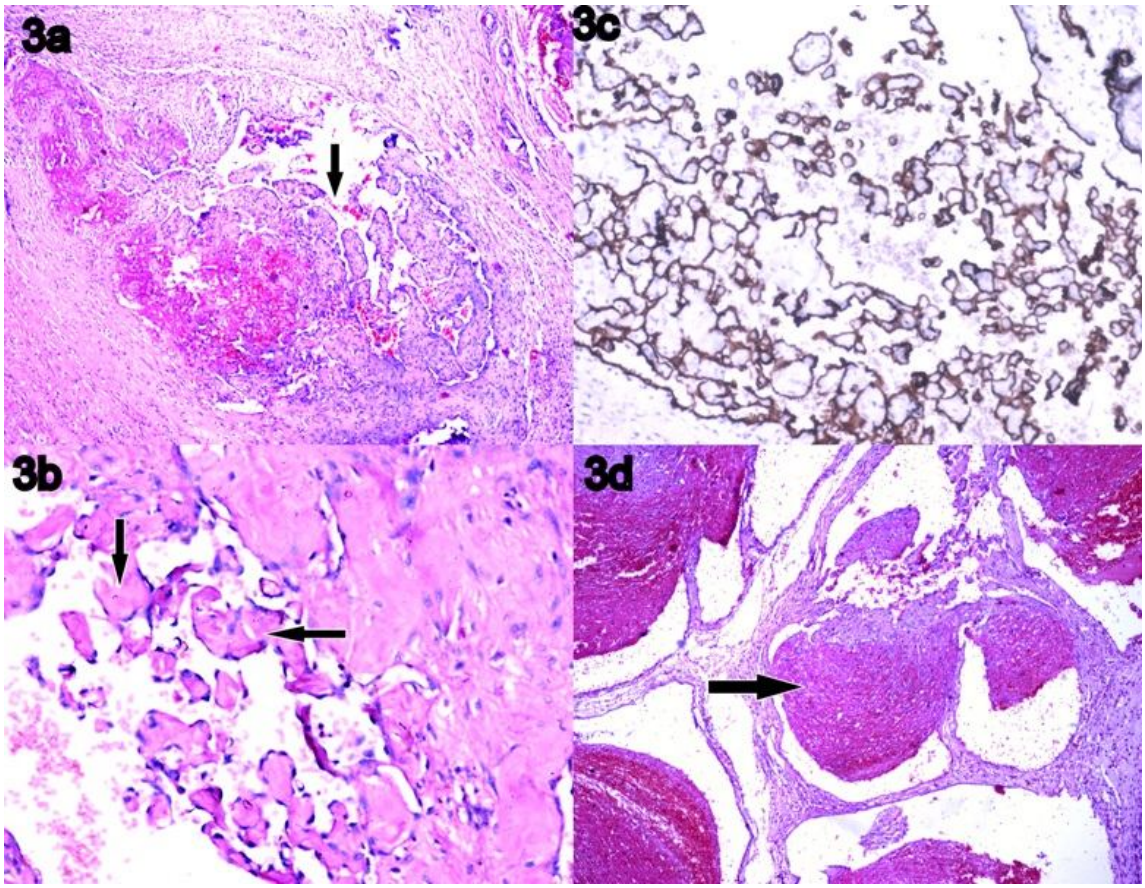

Figure 3: Microscopic appearance of the lesion: **(a)** Numerous papillary proliferations protruding (arrow) into vascular space (H&E, x40). **(b)** High magnification of papillary proliferations with fibrinous and hyaline stalks (arrow) lined by reactive endothelial cells (PAS, x100). **(c)** These lining cells were positive for CD34 antibody (Immunohistochemistry, x200). **(d)** Vascular spaces revealing organized thrombus (arrow) were also evident in other areas (H&E; x40).

DISCUSSION

Masson's hemangioma, which is first described in 1923 in an ulcerated hemorrhoidal vein, is a rare vascular disorder. It is a locally occurring benign tumor, almost always intravascular, and seems to be clearly distinct from an organizing thrombus. From the time of its initial description it has been referred to by various eponyms, including Masson's tumor, Masson's hemangioma, Masson's intravascular hemangioendothelioma, IPEH, or reactive papillary endothelial hyperplasia [1]. It occurs in cranium, thyroid, breast, neck, external jugular vein, tongue, and oral cavity [5]. IPEH can be divided into three different categories: 1) the pure form that occurs within a dilated vascular space, which is most frequently located in the finger, head and neck or in the region between the elbows and hands; 2) the mixed form that appears as a focal change in a hemangioma, vascular malformation, or pyogenic granuloma; 3) the undetermined form, belonging to neither of the first two categories, which has an extravascular origin [6]. The radiologic features of IPEH are not well documented in the literature. Usually after clinical evaluation, Ultrasonography is the first choice imaging method for soft tissue masses. Color doppler ultrasound (CDUS) is also very helpful for vascular conditions, but neither of the two modalities is specific for excluding differential diagnosis. Regarding MRI findings of IPEH, several authors have reported on intracranial IPEH, but only a few cases found in the literature concern the other regions of the body. Lee et al reported papillary endothelial hyperplasia of the extremities that showed heterogeneously isointense and hyperintense on T2WI and mostly isontense on T1WI [7].

Histologically, IPEH has a characteristic exuberant endothelial proliferation within the lumen of medium-sized veins. Microscopically, the tuft-like or papillary proliferation of endothelial cells is nearly always associated with a thrombus, and seems to represent a peculiar variant of an organizing process. The papillary structure and exuberant endothelialization of IPEH, necessitates ruling out the much more frequent angiosarcoma. The following features are important in the differential diagnosis: a) intravascular papillary endothelial hyperplasia is often well-circumscribed or encapsulated; b) the proliferative process is completely limited to the intravascular spaces; c) though the endothelial cells are hyperchromatic, extreme nuclear atypia and frequent mitotic figures cannot be seen; d) papillae are composed of fibro-hyalinized tissue of two or more endothelial cell layers without any covering; e) there is no true endothelial confirmation of IPEH; f) tangential sectioning may reveal pseudochannels, but no irregular or anastomosing blood vessels in the stroma; and g) necrosis is an unusual finding in IPEH [8].Other differential diagnosis include intravenous pyogenic granuloma, Kaposi's sarcoma, and vascular conditions such as hemangioma, angioendothelioma, papular angioplasia, Kimura's disease, bacillary angiomatosis, and intravenous atypical vascular proliferation [5].

Treatment of IPEH is complete surgical excision, which yields the best outcome. Recurrence has been noted in cases of subtotal resection. Nevertheless, IPEH may recur if it arises in a primary vascular lesion which may itself recur. The therapy in these cases should be planned according to the nature of underlying lesions. The prognosis of IPEH is excellent. Follow- up of some large series showed no evidence of metastasis [9].

As a result, achieving a correct diagnosis is essential to avoid subjecting a patient to unnecessarily aggressive therapy. IPEH in scapular region is unusual. Moreover the clinician, radiologist and pathologist should be aware of this benign disorder, which can be mistaken for as sarcoma.

CONFLICTS OF INTEREST

Authors declare that there are no conflicts of interest.

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