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# INTRAVASCULAR PAPILLARY ENDOTHELIAL HYPERPLASIA OF THE LATERAL THIGH : A CASE REPORT

General Surgery

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## ABSTRACT

**Background:** Intravascular papillary endothelial hyperplasia (IPEH), or Masson's tumor, is a rare benign vascular lesion that commonly mimics malignancy both clinically and radiologically. Accurate identification is essential to prevent unnecessary aggressive therapy. **Case Presentation:** We describe the case of a 15-year-old female presenting with a painless vascular mass on the lateral thigh. Radiological investigations revealed a highly vascular lesion suspicious for soft tissue sarcoma. Definitive diagnosis was established through histopathological and immunohistochemical analysis revealing characteristic intravascular papillary proliferations without atypia or mitotic activity. **Discussion:** IPEH poses diagnostic challenges, especially in atypical locations such as the thigh. Its pathogenesis involves endothelial proliferation triggered by thrombus formation. Imaging features are nonspecific, necessitating tissue diagnosis for accurate differentiation from malignant neoplasms. Complete surgical excision provides excellent prognosis with minimal risk of recurrence. **Conclusion:** Awareness of IPEH and multidisciplinary diagnostic evaluation are paramount to avoid misdiagnosis and overtreatment. This case adds to the limited literature on lateral thigh IPEH and underscores the importance of histopathological confirmation in managing vascular soft tissue masses.

## KEYWORDS

Intravascular Pappilary Endothelial Hyperplasia, Masson's Tumor, Masson's Hemangioma

## INTRODUCTION

Intravascular papillary endothelial hyperplasia (IPEH), or Masson's tumor, is an uncommon benign vascular condition characterized by the abnormal proliferation of endothelial cells within a blood vessel. Typically representing about 2% of vascular tumors involving the skin and subcutaneous tissue, it is most frequently observed in the extremities. However, its occurrence near the thigh and knee joint, although less commonly documented, adds to its clinical intrigue. IPEH poses significant diagnostic challenges due to its potential to mimic malignant vascular tumors, such as angiosarcomas, on clinical, radiological, and pathological evaluations. Misinterpretation as a sarcomatous lesion can complicate management, highlighting the importance of recognizing its distinct features for accurate diagnosis.

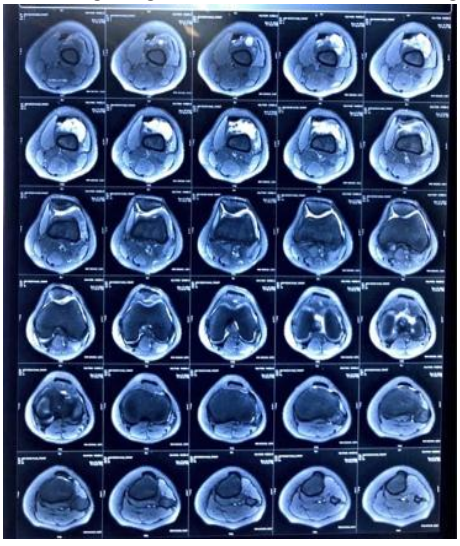

**Image-MRI Image 1**

**Image-MRI Image 2**

### Case Report

A 15-year-old female presented to our department with a swelling over the left lateral thigh, just proximal to the knee joint. She reported no history of recent trauma, prior surgical interventions, systemic complaints, chronic illnesses, or prolonged medication use. On examination, her vital signs were within normal limits. A physical evaluation revealed an otherwise unremarkable condition, except for a palpable, tender mass localized to the distal lateral thigh. There were no associated skin changes such as pallor, and neither inguinal nor para-aortic lymphadenopathy was detected. The mass was fixed to the underlying tissues, and no bruits or signs of infection were observed.

Given the persistence of the mass despite antibiotic treatment, further imaging was conducted. Ultrasonography and color Doppler studies

revealed a non-cystic, heterogeneous, hypervascular solid lesion measuring 3 × 3 × 1 cm. The mass extended into the deeper tissues and demonstrated significant arterial and venous blood flow (Figure 1). It was located anterior to the vastus lateralis muscle and lateral to the rectus femoris muscle.

Primary radiological diagnosis was soft tissue sarcoma with quary, MRI performed at our institution two days later showed the mass was minimally heterogeneous onT1- weighted images (T1WI), mostly isointense to muscle (Fig. 2A). T2 weighted images (T2WI) demonstrated aheterogeneous hyperintense mass with centrally low signal intensity, consistent withthrombosed vessels (Fig. 2B). so in the light of these findigs soft tissue sarcoma could not be ruled out and surgery was planned for accurate diagnosis.

During surgery, a well-defined, vascular mass with a reddish-blue coloration was identified in the left lateral thigh and completely excised. Gross examination of the resected specimen revealed an irregular, hemorrhagic, and lytic mass measuring 3 × 3 × 1 cm. Microscopic analysis demonstrated papillary proliferation of endothelial cells without any signs of atypia, mitotic activity, or necrosis. Hyalinized stromal tissue was noted beneath the endothelial cell lining within the vascular lumen. The final diagnosis was intravascular papillary endothelial hyperplasia (IPEH), also known as Masson's hemangioma, characterized by intravascular papillary proliferations.

IPEH can develop in various locations throughout the body, with a preference for the deep dermis and subcutaneous tissues of the head, neck, fingers, trunk, and lower limbs. It does not exhibit any specific age preference and shows a slight predominance in females, with a male-to-female ratio of approximately 1:1.2. The patient recovered well postoperatively without any complications.

**Image-histopathological Image 1**

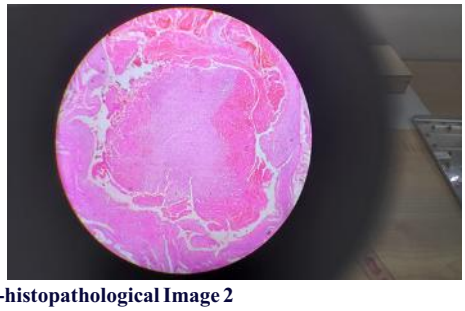

**Image-histopathological Image 2**

## DISCUSSION

Intravascular papillary endothelial hyperplasia (IPEH), also known as Masson's tumor, remains a subject of considerable clinical and pathological interest due to its unique nature as a benign intravascular lesion that can clinically mimic highly aggressive malignancies, particularly angiosarcoma. This resemblance challenges even experienced clinicians and pathologists, therefore emphasizing the importance of comprehensive understanding of its pathogenesis, morphology, differential diagnosis, and management strategies (1-3).

IPEH is widely recognized as a reactive proliferative process rather than a true neoplasm. The prevailing hypothesis suggests that it arises as an exaggerated reparative response to vascular injury and thrombus formation. Endothelial proliferation occurs within the lumen of a blood vessel in reaction to stasis and thrombosis, creating characteristic papillary fronds enveloped by endothelial lining and supported by fibrous connective tissue cores (1,7). The role of growth factors, particularly basic fibroblast growth factor (bFGF), released by macrophages during thrombus organization, has been implicated in the autocrine and paracrine stimulation of endothelial cells, propagating this locally exuberant but non-neoplastic proliferation (8,33). This mechanism helps distinguish IPEH from malignant vascular tumors, which demonstrate autonomous growth unrelated to thrombotic events.

Histologically, the lesion is composed of well-formed papillae protruding into the vascular lumen, lined by endothelial cells showing minimal atypia. Mitotic figures are rare or absent, and necrosis is characteristically lacking, contrasting sharply with angiosarcoma and other sarcomas where these features predominate (1,33). Immunohistochemical staining adds diagnostic precision, with positive markers including CD31, CD34, and factor VIII-related antigen confirming endothelial origin, and low Ki-67 proliferation indices supporting the benign nature of the lesion (1,7,37).

IPEH accounts for approximately 2–4% of all benign and malignant vascular tumors involving the skin and subcutaneous tissues (1,2). It affects a broad demographic range but most commonly presents in middle-aged adults, with no strong gender predilection, though some studies have observed a slight female predominance (4,6,46). The lesion frequently arises in the extremities, particularly the fingers, head, and neck, but less commonly in deeper structures such as the oral cavity, bladder, central nervous system, and internal organs (5,43,46). This broad anatomical distribution accounts for its diverse clinical manifestations and often complicates timely diagnosis.

Clinically, IPEH typically manifests as a slow-growing, well-circumscribed mass, often painless but occasionally causing discomfort due to local mass effect or inflammation (4,36). The lesion is often firm and fixed to underlying tissues but usually lacks overlying skin involvement, ulceration, or regional lymphadenopathy (2,4). When located near joints or deep soft tissues, patients may present with symptoms mimicking synovial sarcoma or aggressive vascular malformations, further complicating clinical assessment (9,31).

Diagnosis of IPEH is notoriously challenging due to its clinical and radiological similarity to malignant tumors. Imaging modalities such as ultrasound and magnetic resonance imaging (MRI) play crucial roles but often fail to provide definitive diagnosis. On ultrasonography, IPEH lesions appear as hypoechoic or mixed echogenic masses with variable vascular flow. MRI reveals lesions that are iso- to hypointense on T1-weighted images and hyperintense on T2-weighted images, with enhancement patterns ranging from homogeneous to heterogeneous (9,10). Central low-intensity areas corresponding to thrombosed vessels or organized thrombi are common (10).

Despite these features, imaging is devoid of pathognomonic signs, and the inability to exclude malignancy radiographically frequently leads to biopsy or excision for definitive histopathological evaluation (29,31). In some cases, extensive vascularity noted on imaging may lead clinicians to suspect arteriovenous malformations or high-grade sarcomas, particularly if rapid growth or pain is reported (31).

The histopathological differential diagnosis is broad and includes benign and malignant vascular lesions. Hemangiomas often share clinical and imaging characteristics but are distinguished by their lobular architecture and endothelial proliferation without marked papillary formations (1). Pyogenic granulomas may resemble IPEH grossly but lack the organized papillary fronds with associated thrombi.

The most critical differential is angiosarcoma, a malignant endothelial tumor with poor prognosis. Angiosarcoma differs histologically by showing marked cellular atypia, multilayered atypical endothelial cells, frequent mitoses, and necrosis (1,33). The Ki-67 proliferation index and immunohistochemical profiles assist in differentiation (33). Misdiagnosis of IPEH as angiosarcoma can lead to unnecessary aggressive treatment including radical surgery, chemotherapy, or radiation, hence the importance of multidisciplinary team discussion and referral to specialized pathology centers when diagnosis is uncertain (8,12).

Other vascular malformations such as arteriovenous malformations or capillary hemangiomas can mimic IPEH clinically, especially on imaging, but these lack the endothelial reactive papillary proliferation hallmark of IPEH (31).

Surgical excision remains the cornerstone of treatment for IPEH.

Adequate excision with clear margins typically results in excellent local control and negligible morbidity (3,7,39). Incomplete excision is the primary risk factor for recurrence, underscoring the importance of meticulous operative technique and careful histological margin assessment (20). Recurrence rates have been reported as less than 10% in most large series (6,20,41).

Alternative approaches such as sclerotherapy or radiotherapy have limited roles but may be considered for lesions in challenging anatomical sites where complete excision risks significant functional or cosmetic morbidity (4,10). Observation or active surveillance has been described in isolated cases with asymptomatic lesions, especially when biopsy confirms the diagnosis and the lesion demonstrates indolent behavior over months (14,18).

Long-term follow-up is advocated since rare delayed recurrences are described, often years after initial surgery (20,41). Patient education on symptom recurrence and regular clinical examination are therefore essential components of care.

While current understanding positions IPEH as reactive in nature, emerging molecular studies hint at complex endothelial biology involved in thrombus-induced proliferation (33). Further molecular characterization may elucidate pathways amenable to medical therapy, potentially reducing the need for surgery in select cases. Genetic and proteomic analyses are ongoing to identify biomarkers predictive of aggressive behavior or recurrence (33,37).Multicenter registries and collaborative research are warranted to gather larger cohorts, given the lesion's rarity and diverse presentations, to refine diagnostic criteria, optimize management, and develop targeted therapies (39).

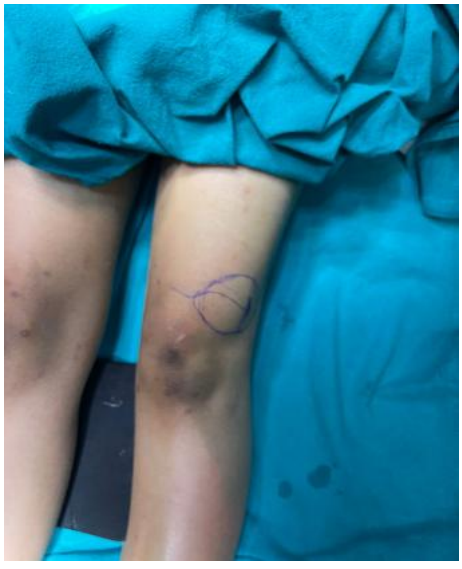

**Image- Pre Operative Image**

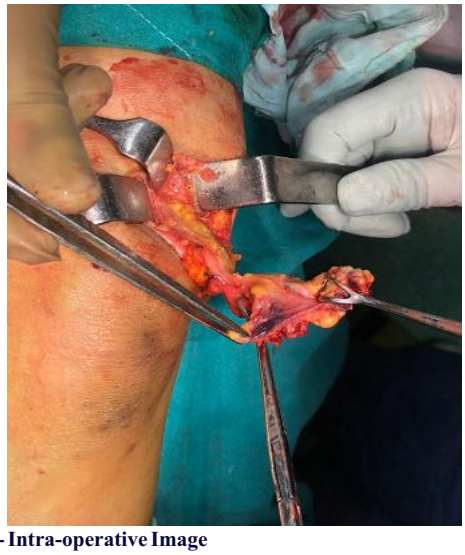

**Image- Intra-operative Image**

**Image- Post Operative Image**

## CONCLUSION

Intravascular papillary endothelial hyperplasia (IPEH), or Masson's tumor, is a rare benign vascular lesion that often masquerades clinically and radiologically as malignancy. Accurate differentiation from angiosarcoma and other aggressive vascular tumors is vital, relying heavily on thorough histopathological evaluation and immunohistochemical staining. IPEH typically arises as a reactive endothelial proliferation associated with thrombus formation, distinct from true neoplastic processes.Complete surgical excision remains the mainstay of treatment and offers excellent prognosis with minimal recurrence risk. The lesion's benign nature necessitates cautious interpretation of imaging findings to avoid unnecessary aggressive interventions. Multidisciplinary collaboration comprising radiologists, pathologists, and surgeons optimizes diagnostic precision and patient management. Extended follow-up is advised to detect rare cases of recurrence.This case of IPEH presenting as a mass in the lateral thigh contributes valuable insights into the clinical, imaging, and pathological features of this unusual entity in a less commonly affected site. Enhanced awareness and recognition of IPEH enhance clinical outcomes by preventing misdiagnosis and overtreatment. Further multicenter studies and molecular research are warranted to better elucidate the pathogenesis and explore potential non-surgical therapeutic options for challenging cases. In conclusion, IPEH exemplifies the necessity of integrating clinical acumen, imaging, and pathology to ensure accurate diagnosis and curative treatment of this rare but important vascular lesion.

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