



## Case Report

# Masson's intravascular hemangioma in a child: Uncommon histopathological entity in breast

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### ABSTRACT

Masson's hemangioma is an uncommon, benign and rare vascular tumor. It's often misdiagnosed as soft tissue sarcoma such as angiosarcoma or Kaposi sarcoma. It is also known as intravascular papillary endothelial hyperplasia (IPEH) accounting for 2% of skin and soft tissue vascular tumors. IPEH characterized by exuberant endothelial proliferation within the lumen of medium sized veins. The most common sites affected are head, neck, fingers and trunk. But the occurrence of IPEH in breast is rarely reported in literature.

We diagnosed this case in a 2 month old child presented with right breast bud swelling. On histopathology diagnosed as IPEH/ Masson's hemangioma. The aim of this case report to highlight the important role of clinical suspicious of such rare entities and histopathological diagnosis to avoid unnecessary aggressive line of treatment for better management of patient care.

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## 1. Introduction

Masson's hemangioma or intravascular papillary endothelial hyperplasia (IPEH) is a rare, benign tumor accounting for 2% of skin and soft tissue vascular tumors.<sup>1</sup> In 1923 it was first described by Pierre Masson in an ulcerated hemorrhoidal vein in a man and named as 'hemangioendothelioma vegetant intravasculaire'.<sup>2</sup> Later on it has been referred by various names as Masson's hemangioma, Masson's tumor, IPEH, or reactive papillary endothelial hyperplasia.<sup>3</sup> The present terminology i.e. IPEH was proposed by Clarkin and Enzinger in 1976.<sup>2</sup> It is presented as a firm to tender nodule or slightly elevated mass that appeared as red or blue on the skin with sharp demarcation and slow growth.<sup>4</sup> We report this case here in view of its rarity and misdiagnosed as malignant mass and to highlight the role of histopathology in the final diagnosis of this benign, reactive lesion to avoid invasive treatment to the patients.

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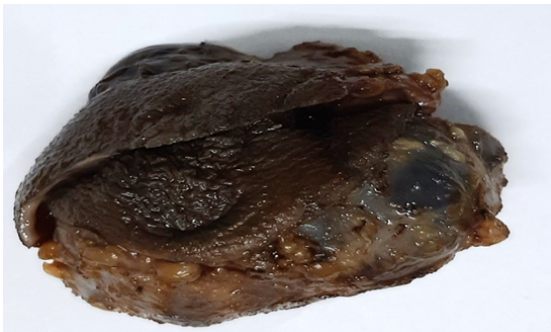
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## 2. Case Report

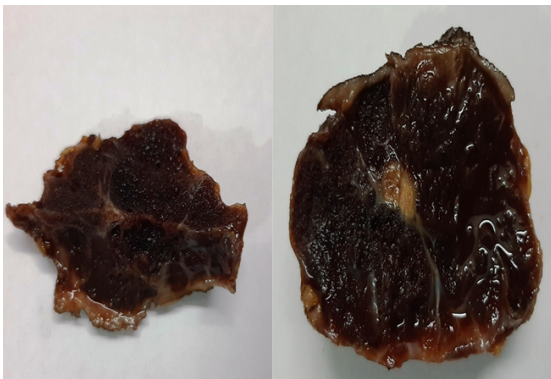
A 2 month old male child presented to the pediatric department of our hospital with chief presentation as soft tissue swelling in right breast bud since birth. The mass was gradually increasing in size to the present size of 6x6 cms. Personal and family history was not contributory. Clinically, the mass was firm, painless and with reddish blue discoloration of overlying skin. Local examination showed the mass measures 6x6x4 cms, not adherent to the underline structures with no signs of inflammation. Vascular malformation was clinically suspected and wide local excision was planned. All the biochemical, serological and hematological parameters were within normal limits. Wide local excision was performed and the resected specimen sent for histopathology. The post operative period was uneventful. Gross findings-We received excised soft tissue mass measuring 5.5.x5x3 cms. E/S-covered with elliptical piece of skin with grey brown, circumscribed and nodular areas (Figure 1). C/S-showed mesh like spongy network of brownish to black material with blood clots admixed with

grey white areas (Figure 2).

Light microscopy- Showed tissue bit lined by stratified squamous epithelium and subepithelium and superficial dermis showed well circumscribed vascular tumor. (Figures 3 and 4). The tumor composed of dilated vessel contains multiple papillary proliferations of plump endothelial cells without atypia around a fibrinous core (Figure 5). Fibrin deposition and thrombi was seen. Variable pleomorphism and stratification noted with absent marked atypia, mitotic figures, and necrosis. Proliferating endothelial cells form solid islands in the dermis with hyaline core. Final histopathological diagnosis was given as IPEH/Masson’s hemangioma of right breast bud.



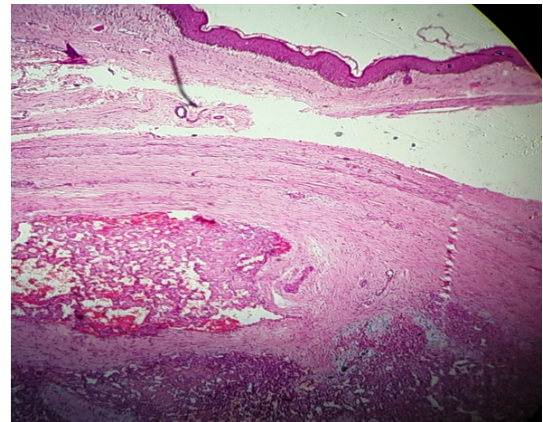
**Fig. 1:** Gross image of resected mass with piece of skin with grey brown, circumscribed and nodular areas.



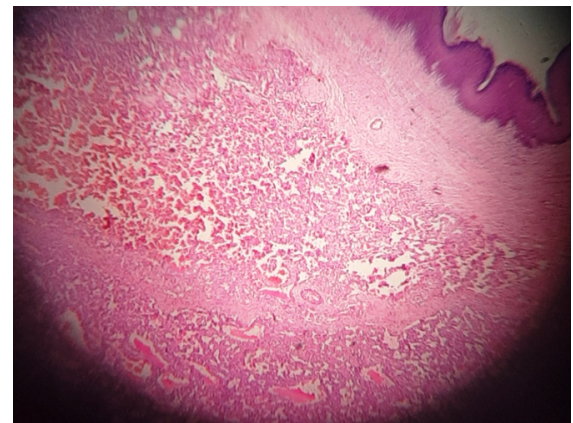
**Fig. 2:** Cut section of the soft tissue mass with mesh like spongy network of brownish to black material with blood clots.

**3. Discussion**

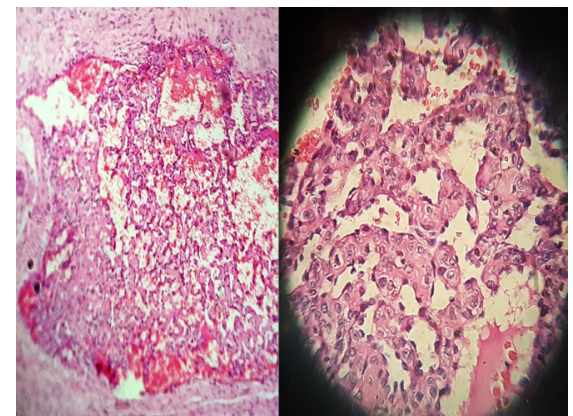
IPEH consist of reactive proliferation of endothelial cells with papillary structures and thrombus formation. In view of its morphology simulates various benign and malignant vascular entities.<sup>5</sup>Masson’s hemangioma is mostly an intravascular lesion but extra vascular hematogeneous organization may be present.<sup>6</sup> Clinically, IPEH is extremely important because it can be confused with malignant conditions like angiosarcoma or Kaposi sarcoma.<sup>7</sup>We can



**Fig. 3:** Photomicrograph showed tissue bit lined by stratified squamous epithelium with subepithelial nodular vascular tumor (H & E stain, x100)



**Fig. 4:** Subepithelium and superficial dermis showed well circumscribed vascular tumor with features of IPEH (H & E stain, x100)



**Fig. 5:** The tumor composed of dilated vessel contains multiple papillary proliferations of plump endothelial cells without atypia/mitosis around a fibrinous core. (H & E stain, x400)

easily differentiate with condition from various reactive, benign and malignant ones. Preoperatively its differential are varies from fibrolipoma, ganglion, soft tissue sarcoma to angiosarcoma. IPEH has no age/gender predilection and has been frequently reported from head, neck, fingers, trunk and cutaneous veins.<sup>8</sup> Grossly usually presented with small, firm, superficial mass with bluish red discoloration of the overlying skin.<sup>8</sup>

Histopathological examination is the gold standard of the excised mass for the final diagnosis of IPEH and important to rule out various differentials.<sup>8</sup> The specific criteria to diagnose Masson's hemangioma on histopathology are – well circumscribed or capsulated mass with endothelial proliferation in the vessel. The multiple papillae are composed of fibrohyalinsed type of two or more endothelial cell layers. Proliferative process with fibrin thrombi in the vessels. The endothelial cells have hyperchromatic nuclei but extreme nuclear atypia and mitotic figures and necrosis are not seen.<sup>9</sup> Microscopically, IPEH classified into three categories as pure, intermediate and mixed forms. The pure form is mostly seen fingers, elbow, hands and neck. It's occurred within dilated vascular space.<sup>10</sup> Such morphology is seen in our case. The mixed form is associated with the changes in pre-existing hemangioma, vascular malformation or pyogenic granuloma. The intermediate form is rare and not belong to either of two entities and has extra vascular origin.<sup>10</sup> However, these pathological entities doesn't have any clinical significance since the treatment is surgical excision. The recurrence rate is very rare if the mass is resected completely.<sup>10</sup>

Pathogenesis of the Masson's hemangioma is variable. Masson's states that the endothelia proliferation occurs and the thrombus arise secondary to endothelial proliferation.<sup>2</sup> Second theory by Clearlens and Enzinger suggested that this papillary stratification occurs after a pre-existing thrombus organizes. Now days, it's considered as reactive vascular proliferation following traumatic vascular stasis due to release of fibroblastic growth factors.<sup>2</sup>

Regarding differential diagnosis of IPEH the main entities are angiosarcoma and Kaposi sarcoma. Angiosarcoma is rarely intravascular and without papillary architecture. The nuclear atypia, pleomorphism and piling of endothelium is common with necrosis and solid areas in angiosarcoma. In Kaposi sarcoma, the intravascular location is rare with no papillae formation and necrosis with presence of nuclear atypia. Thus IPEH is easily differentiates from malignant conditions. In undifferentiated and complicated cases, CD 34 and 31 helpful as immunohistochemical markers for endothelial cells.

#### 4. To conclude

The diagnosis of Masson's hemangioma is almost important for the surgeons as it simulates malignant tumor like angiosarcoma to avoid aggressive line of management for the betterment to the patient's health.

#### 5. Source of Funding

None.

#### 6. Conflict of Interest

None.

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