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# Intravascular Papillary Endothelial Hyperplasia (Masson's Tumor) of the Supraclavicular Region: Management of a Rare Case

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Data Collection B  
Statistical Analysis C  
Data Interpretation D  
Manuscript Preparation E  
Literature Search F  
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**Patient:** Female, 24-year-old  
**Final Diagnosis:** Hemangioendothelioma  
**Symptoms:** Swelling  
**Medication:** —  
**Clinical Procedure:** —  
**Specialty:** Otolaryngology • Surgery

**Objective:** Rare disease

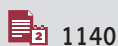
**Background:** Masson's tumor, also known as IPEH (intravascular papillary endothelial hyperplasia), is a rare benign vascular process, comprising approximately 2% of vascular tumors of the skin and subcutaneous tissue. IPEH presenting as a neck mass is rare, with only 5 reports of anterior neck mass and 7 cases of lateral neck lesions, of which 1 was in an external jugular vein aneurysm. To the best of our knowledge, the localization of intravascular papillary endothelial hyperplasia in the supraclavicular region is rarely reported.

**Case Report:** We describe our management of a non-traumatic and non-painful mass on the right supraclavicular region in 24-year-old woman. Ultrasound revealed a heterogeneously hypochoic mass with intense vascularization. Magnetic resonance imaging (MRI) showed a formation with lobulated contours and closely related to muscular layers of serratus anterior muscle. The findings of ultrasound-guided biopsy (FNA) were inconclusive. Complete removal of the mass was performed. Histopathological examination showed a well-circumscribed lesion with numerous small papillary structures. The papillae had hyalinized hypocellular cores covered by flattened endothelium. Immunohistochemical analysis showed endothelial positivity for CD34 and CD31. These features are typical of IPEH. No recurrence had occurred at 12 months after surgery.

**Conclusions:** The differential diagnosis of malignant tumors like angiosarcoma is important because the prognosis significantly differs. Surgical excision is the treatment of choice. In our experience, the size of the lesion and its critical landmarks have not influenced the outcomes of the surgical procedure in terms of potential functional damages.

**Keywords:** Vascular Neoplasms • Head and Neck Neoplasms • Subcutaneous Tissue • Neck • Antigens, CD34 • Vascular Diseases

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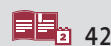
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## Background

Intravascular papillary endothelial hyperplasia (IPEH) is a rare vascular lesion comprising approximately 2% of benign and malignant vascular tumors of skin and subcutaneous tissues [1-4]. It can occur in all age groups and has no race or ethnic group preferences [5] but there is a slight female predominance, which can be related to involvement of hormonal factors [6]. It can affect any blood vessel in the body but has a predilection for the head and neck region, fingers, trunk, and upper and lower extremities [7]. Most reported head and neck cases have involved skin and subcutaneous tissues of face and scalp [8-10]; less common sites are buccal mucosa [4,11], tongue [12,13], pharynx [14], larynx and hypopharynx [15], masseter muscle [16] and parotid gland [17], maxillary sinus [18], mandible [19], orbit [20], and ocular adnexa [21]. IPEH presenting as a neck mass is rare, with only 5 reports of anterior neck mass [22-25] and 7 cases of lateral neck lesions [26-32], of which 1 was in an external jugular vein aneurysm [30].

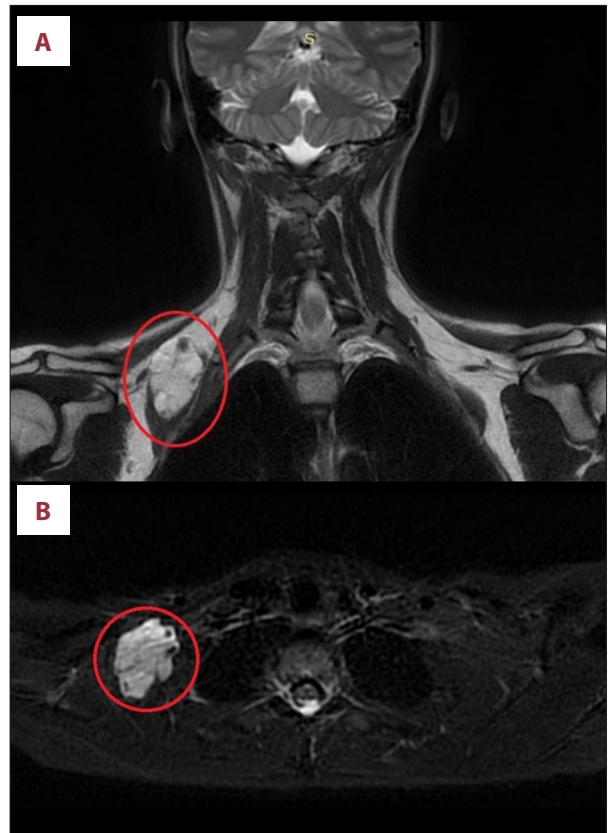
To the best of our knowledge, the localization of intravascular papillary endothelial hyperplasia in the supraclavicular region has rarely been reported [33,34]. Only 1 case in close proximity of serratus anterior muscular fibers was reported in the English language literature and treated by a multidisciplinary team.

Here, we report a clinical case of IPEH located in subcutaneous soft tissue of the supraclavicular region that was successfully managed by otolaryngologists without postoperative complications and with no recurrence of the disease 12 months after the surgical procedure.

## Case report

A 24-year-old woman presented with non-traumatic and non-painful swelling on the right supraclavicular region that was noticed 1 year ago and was progressively increasing in size. At the dermatological examination, the lesion appeared mobile, non-tender, non-compressible, with fibroelastic consistency, without alterations of the overlying skin and pulsatility. Ultrasound revealed a heterogeneously hypoechoic mass and Doppler US recognized in it an intense vascularization.

T1-weighted and T2-weighted magnetic resonance imaging (MRI) was conducted, showing a 43×30×49 mm formation with lobulated contours and closely related to muscular layers of serratus anterior muscle (Figure 1A, 1B). The signal is mostly fluid (likely serohematic content) and few calcifications are described. After intravenous administration of contrast (gadobutrol) there is a septal enhancement.



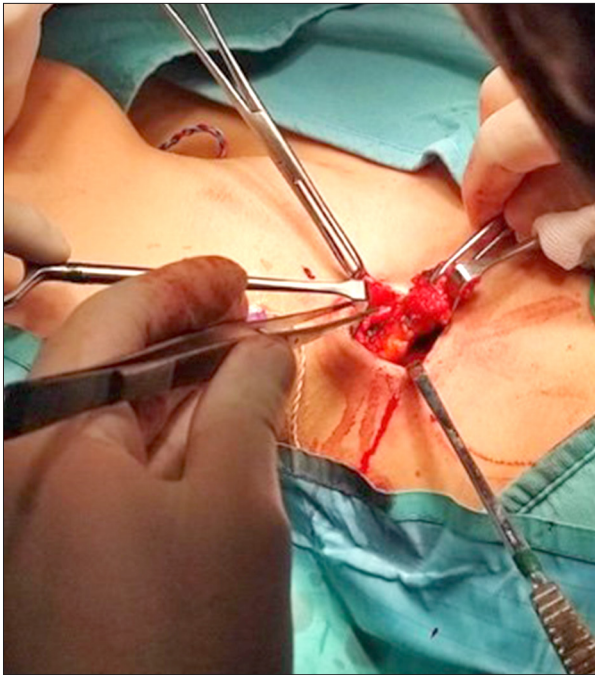
**Figure 1.** (A) Coronal T2 HR FSE Propeller; (B) axial T2 STIR. A 43×30×49 mm formation with lobulated contours is closely related to muscular layers (circles). The signal is mostly fluid (likely serohematic contents) and few calcifications are present.

An ultrasound-guided biopsy was performed, but the findings were inconclusive.

The presence of a lymphangioma was hypothesized and the mass was radically excised for histopathological evaluation.

Intraoperative neurophysiologic monitoring has been used. The mass was completely removed. Macroscopically, the mass was 5.5×3×3.5 cm in size (Figures 2, 3). After surgery, a drainage tube has been placed. The patient had a fast and uncomplicated recovery. There was no evidence of recurrence after 12 months.

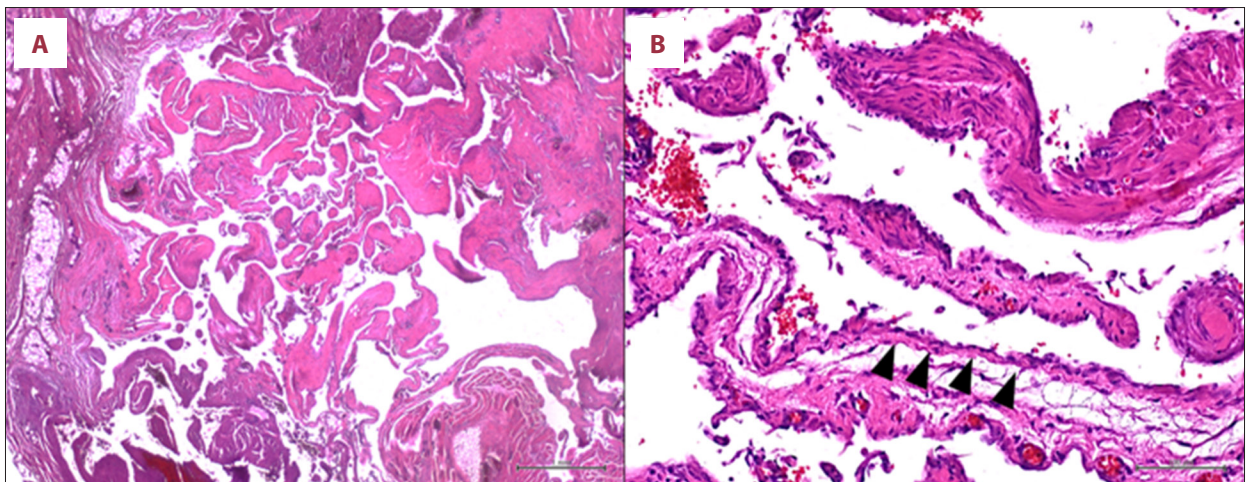
Under pathologic examination, the lesion appears well-circumscribed and presents numerous small papillary structures. The papillae have a hyalinized hypocellular cores covered by flattened endothelium (Figure 4). Immunohistochemical analysis showed endothelial positivity for CD34 and CD31 (Figure 5). These features are typical of IPEH.



**Figure 2.** Surgical field. Small skin incision; a mass in the left supraclavicular fossa can be seen.



**Figure 3.** Resection “en-bloc” of the mass. Macroscopically, the mass was 5.5×3×3.5 cm in size.



**Figure 4.** (A) Vascular malformation containing organizing thrombus with papillary endothelial hyperplasia (H&E, 2.5× magnification, scale bar 1mm). (B) Papillae with hyalinized hypocellular cores covered by flattened endothelium (black arrow-head) (H&E, 20× magnification, scale bar 100 µm).

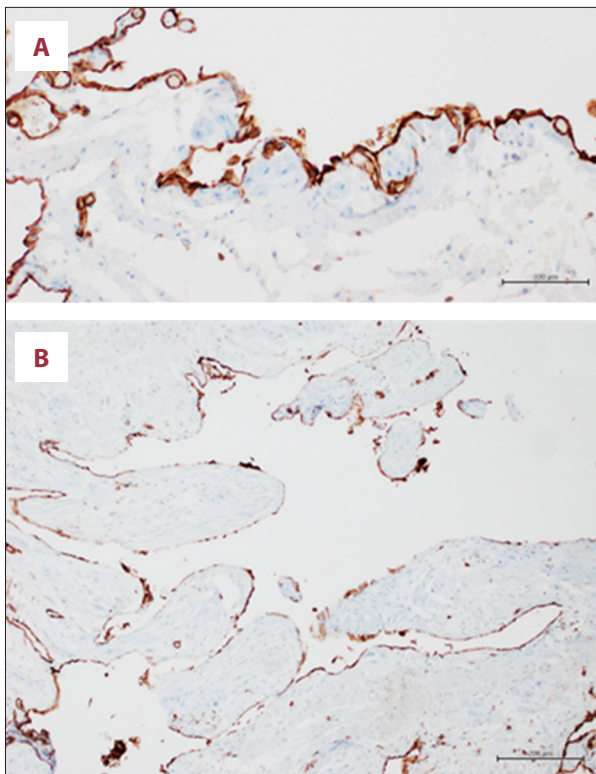
## Discussion

The complexity of the anatomy of the supraclavicular fossa allows for an extensive differential diagnosis for regional pathology, which categorizes into neoplastic, congenital, and infectious etiologies. According to some authors, the majority of supraclavicular masses biopsied via FNA were found to be the result of a malignant process, with metastatic spread being far more likely than lymphoma [35,36].

IPEH was first described in 1923 by Pierre Masson [37]. He described an intravascular papillary proliferation in the lumen of an inflamed hemorrhoidal plexus in a 68-year-old man and named it “hémangioendothéliome végétant intravasculaire”.

In 1932, Folke Henschen [38] reported a similar case and named it “endovasculite proliférante thrombopoiétique”, describing it as a reactive process instead of a neoplasm. The name of “intravascular papillary endothelial hyperplasia” (IPEH), reported by Clearkin and Enzinger in 1976 [39], is currently the most





**Figure 5.** Immunohistochemical analysis showed endothelial positivity for CD34 (A: 20× magnification, scale bar 100 μm) and CD31 (B: 10× magnification, scale bar 200 μm).

commonly used, since it underlines the non-neoplastic character of this disease.

Three types of IPEH were described: a primary type, the most common, which is a pure endovascular form that originates in dilated vascular spaces; a mixed or secondary form that occurs in pre-existing vascular abnormalities; and the undetermined type, which is a rare extravascular hemangioma form that arises from hematomas [25,29]. The pathogenesis of IPEH seems to be multifactorial [10]. In 30% of cases, it is related to trauma or vascular abnormalities (eg, hemangiomas, vascular malformations, pyogenic granulomas, blood stasis) that induce a perivascular inflammation and favor the onset of a reactive process by endothelial cells through a consistent secretion of FGF-β [5]. However, the trigger stimulus is still not clear, so the etiology remains unknown [5,15,40]. Salyer et al found a peculiar pattern of thrombus organization [28,41]. CD 31 and CD 34 were found to be the most efficient immunohistological markers to identify the vascular origin of IPEH, in comparison to FVIII, type IV collagen, SMA and MSA antibodies, CD105, and Ki-67 [28,42]. Clinical and radiological features are not adequate to guide the diagnosis; therefore, histological characterization of the lesion is necessary.

IPEH can be mistaken for other benign subcutaneous tumors, such as lipomas, angiomas, and cysts, but the differential diagnosis is more important with malignant tumors like angiosarcoma and Kaposi's sarcoma [7], since the prognosis will change.

Angiosarcoma is distinguished from Masson's tumor by the presence at histopathological examination of solid and necrotic areas, mitotic figures, or pleomorphic cells and by the absence of confined intralaminar location [10,26]. Other differential diagnoses to consider are pyogenic granuloma, mucocele, the intravascular form of hemangioma, and malignant endovascular papillary angioendothelioma [25,29].

Surgical excision is the treatment of choice, generally with a good prognosis. Recurrences are rare and mostly associated with incomplete excision [25,26].

The size in combination with the site of the lesion gave us some concerns in terms of potential complications, due to the uncertain histology and vascularization of the mass.

Intraoperative neurophysiologic monitoring is crucial since several nerve structures such as the brachial plexus, cervical plexus, and phrenic nerve are potentially at risk.

Bleeding control has a fundamental role too, since IPEH has a conspicuous vascularization and the vasculature within the region of the supraclavicular triangle is complex, with presence of major vessels like subclavian, transverse cervical, and suprascapular arteries and veins, as well as external and internal jugular veins and their branches.

## Conclusions

To the best of our knowledge, the localization of intravascular papillary endothelial hyperplasia in the supraclavicular region is rarely reported. The differential diagnosis with malignant tumors like angiosarcoma is fundamental since the subsequent prognosis will significantly change. Surgical excision is the treatment of choice. In our experience, the size of the lesion and its critical landmarks do not influence the outcomes of the surgical procedure in terms of potential functional damages.

## Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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