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ENT

KEY WORDS:

MASSON'S TUMOR ON THE FOREHEAD: A RARE CASE REPORT

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ABSTRACT

Intravascular papillary endothelial hyperplasia (IPEH) or Masson's tumor is a rare benign entity commonly found on the head, neck, and upper extremities. Typically, IPEHs cause no symptoms and present as slowly growing soft-tissue masses. We present a 50-year-old female, a case of Masson's tumor over the forehead. Masson's tumor, with the involvement of the forehead is rare.

INTRODUCTION:

Masson's tumor is a rare, benign, reactive proliferating vascular lesion, also termed as intravascular papillary endothelial hyperplasia (IPEH) contributing only 2% of all skin and soft-tissue vascular tumors[1,2].The clinical presentation of Masson's tumor as a skin-colored-to-bluish nodule may be confused not only with other benign and malignant vascular lesions but also with nonvascular lesions. We report this case to increase awareness regarding this uncommon and benign tumor.

Case Report:

A 50-year-old female presented with an asymptomatic swelling over the forehead for 1 year. The lesion was completely asymptomatic, initially small, and slow-growing. On examination, 3 cm × 3 cm soft, cystic, painless, nonindurated, skin-colored nodular swelling over the forehead was present without any skin surface changes. The swelling was compressible, nonpulsatile, and not fixed to any underlying or overlying structures. After detail history and examination written informed consent were taken and under all aseptic precautions and under local anesthesia painting and drapping was done. The lesion was completely excised with keeping in differentials as appendegeal tumor and neurofibroma. Excised tissue sent for HPE. Histopathology of the excised specimen showed normal epidermis and upper dermis. The reticular dermis showed intravascular thrombotic tumor with hyalinized eosinophilic material. Anastomosing vascular channels with intraluminal papillary projections (fronds) were also seen. There was no nuclear atypia. The absence of nuclear atypia, solid areas, or necrosis ruled out malignancy.

Classical histopathological appearance with clinical correlation confirmed the diagnosis of Masson's tumor. We didn't performed Immunohistochemical markers because of unavailability of facility.

DISCUSSION-

The masson's tumor was first described in 1923 by the French pathologist Pierre Masson within a thrombosed hemorrhoid. He thought the disorder was a benign neoplasm. Masson theorized that the proliferation of benign cells led to hemorrhoid thrombus formation[3,4]. Masson's tumor accounts for 2% of the vascular tumor of the skin and soft tissue[2]. Masson's tumor affects the skin and subcutis of the head and neck region (23%), lower extremities (77%), and fingers (16%) commonly[5]. Less than 250 cases in the head and neck have been reported over the past 35 years, with <5% involving the forehead[6]. Clerkin and Enzinger coined the most widely accepted histological term intravascular papillary endothelium proliferation tumor (IPEH) in 1976[1].Various possible mechanisms have been postulated regarding etiopathogenesis, a vascular injury leads to inflammation and stasis with the release of endothelial basic fibroblast growth factor by macrophages plays a important role in the formation of intravascular papillary endothelium proliferation tumor (IPEH)[6].

In the rare instances in which arterial involvement has been reported, IPEH tends to be associated with aneurismal degeneration of the vessel in both the peripheral and intracranial circulations[3]. Hashimoto *et al* classified IPEH into three types; Type I, a "pure," primary type, which arises *de novo* in dilated vessels; Type II, a secondary, "mixed" form, which occurs within pre-existing vascular anomalies that may exhibit thrombosis; and Type III or the undermined type, an extravascular type, which occurs in hematomas and is rare[7]. The first type is the most common[8]. In our case, IPEH is of type I. Clinically, the primary lesions are usually tender nodules less than 2 cm in size. Clinically, the lesion needs to be differentiated from hemangioma, lymphangioma, angiosarcoma, hematoma, Kaposi sarcoma, traumatic fibroma, traumatic neuroma, and neurofibroma[8]. These tumors are typically positive for CD31, CD34, smooth muscle actin, and factor VIII-related antigen. CD105 (a marker for primary endothelial neoplasms) is typically negative and can help differentiate a Masson tumor from an angiosarcoma[9].

Spontaneous, atraumatic STA aneurysms are a rare entity, representing approximately 8% of all STA aneurysms[10]. Aneurysmal dilation of the STA is typically associated with trauma, to which the vessel is predisposed because of its superficial course within connective tissue. Typically, post-traumatic aneurysms develop 2 to 6 weeks after blunt head trauma[11,12].The majority of these are pseudoaneurysms[11].

The imaging findings in intravascular papillary endothelium proliferation tumor (IPEH) are usually, doubtful; histopathology usually confirms the diagnosis. Characteristic histopathology of intravascular papillary endothelium proliferation tumor (IPEH) shows the intravascular nature of the process consisting of a mass of anastomosing vascular channels with a variable degree of hyaline papillary projections lined by prominent endothelial cells without any atypia ruled out malignant nature of lesion.

Masson's tumor is a reactive condition on the basis of histopathology the differential diagnosis is Kaposi's sarcoma, and malignant angiosarcoma, endovascular papillary angioendothelioma or Dabska's tumor since the lesion is, usually, curable by surgical excision alone[13]. Masson's tumor shows commonly a papillary architecture confined to the intravascular location with an absence of solid areas and

necrosis without any evidence of cellular pleomorphism, unlike angiosarcoma[8]. CD105 is positive in primary vascular neoplasms, differentiating IPEH from angiosarcoma[6].

The prognosis of intravascular papillary endothelium proliferation tumor (IPEH) is excellent. Complete resection is believed to be curative and should be meticulously undertaken as residual tumor can aggressively recur[4]. Complete excision was done in our case with computed tomography-scan and local ultrasonography showing no extension to surrounding tissue. The patient is followed up for any evidence of recurrence.

CONCLUSION:

Masson's tumor is a benign tumor that may get confused on histopathology with malignant tumors. Several markers are available to differentiate masson's tumor from other primary endothelial neoplasms.

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Conflicts of interest:
There are no conflicts of interest.

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