Intravascular Papillary Endothelial Hyperplasia Presenting as a Cystic Mass in the Scalp with Underlying Bone Involvement: A Rare Entity

Abstract
Intravascular papillary endothelial hyperplasia (IPEH) also known as Masson’s hemangioma is a rare benign reactive vascular lesion of the skin and subcutaneous tissue. It is usually confined to the lumen of preexisting vessels/vascular malformations. It is characterized histologically by papillary fronds lined by proliferating endothelial cells. This lesion is often misinterpreted as soft-tissue tumors and other benign and malignant lesions. We present a case of 13-year-old female with IPEH of scalp causing bony defect because of its rarity and diagnostic challenge, it posed due to nonspecific clinical and radiological findings.

Keywords: Hemangioma, intravascular, Masson’s, papillary, scalp

Introduction
Intravascular papillary endothelial hyperplasia (IPEH) is a benign vascular lesion consisting of reactive proliferation of endothelial cells existing in organizing thrombus. It occurs mainly in the head and neck region and extremities.[1] It may occur as a primary or pure form developing in a distended vessel, or it can be associated with hemangiomas, pyogenic granulomas or lymphangiomas.[2] Lesions presenting as scalp swelling with bony defect is very rare, and only a few cases have been reported in literatures till now.[3] Here, we report a case of IPEH presenting as a scalp swelling with involvement of underlying bone, laying special emphasis on differential diagnosis, and review of literature.

Case Report
A 13-year-old female patient noticed a scalp swelling on the left side of temporal region 1 year back with no history of previous injury. The swelling gradually increased in size with moderate pain in the temporal region for the past 1 week duration. On examination, the patient was conscious, alert with intact cognitive functions. On physical examination, there was a nodular soft swelling on the left temporal region of size 2.5 cm × 2.0 cm. There were no abnormal pulsations, and the swelling was nonreducible, nonfluctuant, and freely mobile. The overlying skin was normal with no secondary changes. Computerized tomography (CT) revealed a well-demarcated oval isodense to hyperdense lesion in the left temporoparietal region in the subcutaneous plane measuring 2.2 cm × 2.0 cm with scalloping of underlying outer table of the skull. Irregular cortical erosion in the inner table of skull also was seen [Figure 1]. Magnetic resonance imaging (MRI) brain also showed the presence of 2.2 × 2.0 cm-sized lesion in the left temporoparietal region. The lesion was hypointense on T₁ and heterogeneously hyperintense on T₂-weighted images. The lesion involved the underlying skull bone creating a bony defect and extending into the extradural space. En bloc removal of the tumor with wide resection of margins was performed and received in the department of pathology. Grossly, the specimen was well circumscribed, gray-brown to tan and predominantly cystic. It measured approximately 2.0 cm × 2.0 cm. External surface was congested. On cutting, it shows a cyst, lumen of which was filled with blood clot and small papillary fronds [Figure 2]. Histopathological examination of hematoxylin- and eosin-stained smears revealed a dilated blood vessel with lumen having multiple, small papillary structures lined by single to multiple layers of plump endothelial cells. These papillae were fine, forming anastomosing channels in a...
loose connective tissue. There was no evidence of mitotic activity and atypia of the endothelial lining [Figure 3]. Immunohistochemical stain for CD34 showed positivity in endothelial cells [Figure 4]. A final diagnosis of IPEH was made. Postoperatively, the patient was normal and discharged. A repeat MRI on follow-up after 2 months showed no tumor.

Discussion

Pierre Marson (1923) described an intravascular papillary formation within the lumen of hemorrhoidal plexus in a man and named as “Hemangioendothelioma vegetant intravasculaire.” He believed it to be a neoplastic lesion.\(^4\) Hanschen also noticed this lesion in nasal and laryngeal vessels and considered it to be a reactive process rather than a neoplasm.\(^5\) Various terminologies were used by different authors for this lesion including papillary fibroendothelial intravascular endothelioma, Masson’s tumor, Masson’s pseudoangiosarcoma, intravascular angiomatosis, and papillary fibroendothelioma, but the most accepted term being benign IPEH which was first used by Clearkin and Enzinger.\(^6\)

Its pathogenesis is poorly understood. Now, it has been concluded to be a vascular reactive proliferation following traumatic vascular stasis.\(^7\) IPEH has been classified into three types.\(^2\) Type I (most common) – Primary or pure form that occurs within dilated vascular spaces.

Type II – Secondary or mixed form occurring in preexisting varices, hemangiomas, pyogenic granuloma, lymphangioma, or arteriovenous malformation.

Type III (least common) – This is found in an extravascular location in the bed of a hematoma and trauma is usually a prerequisite. In our case, IPEH is of type I.

This lesion occurs most commonly in the head and neck region (23%), lower extremities (77%), and fingers (16%) mainly in the skin and subcutaneous tissue.\(^8\) This lesion is found to be more common in females and occurs more
In our case, the tumor was completely intravascular and superficial; complete resection was performed. No recurrence was reported on follow-up on MRI after 2 months.

Microscopically, the endothelial cells proliferate in a papillary pattern toward the lumen of an enlarged blood vessel without atypia or mitotic activity. The core of the papillae is composed of fibrous connective tissue which is frequently hyalinised and hypocellular. On histopathological examination, IPEH can be distinguished from other differential diagnoses on the basis of certain key features as summarized in Table 1.[9]

One of the close differential diagnoses is Dabska tumor, also known as endovascular papillary angioendothelioma. These tumor cells are positive for Von Willebrand factor, CD31, CD34, and vascular endothelial growth factor receptor-3 (VEGFR-3) on immunohistochemical staining.[10] In our case, endothelial cells did not show any atypia (nuclear hyperchromasia, high nuclear-cytoplasmic ratio, etc.) or cytoplasmic vacuolation. No mitotic figures were present.

Immunohistochemically, IPEH shows positivity with vimentin, factor VIII, CD31, CD34, and ULEX europaeus agglutinin indicating its endothelial origin. CD34 stain showed positivity in endothelial cells in our case.

The treatment of IPEH includes complete surgical excision.[9] In our case, the tumor was completely intravascular and superficial; complete resection was performed. No recurrence was reported on follow-up on MRI after 2 months.

Table 1: Showing various differential diagnoses and their comparison with IPEH

| Diagnosis                      | Intravascular location | Papillary architecture | Solid areas and necrosis | Piling of endothelial cells | Cellular pleomorphism |
|--------------------------------|------------------------|------------------------|--------------------------|-----------------------------|-----------------------|
| IPEH                           | Common                 | Common                 | No                       | Rare                        | Rare                  |
| Angiosarcoma                   | Rare                   | Rare                   | Common                   | Common                      | Common                |
| Dabska tumor                   | Common                 | Common                 | Rare                     | May be present              | Common                |
| Kaposi sarcoma                 | Rare                   | No                     | Uncommon                 | Rare                        | Common                |
| Cavernous/capillary hemangioma | No                     | No                     | No                       | Rare                        | Rare                  |

IPEH: Intravascular papillary endothelial hyperplasia

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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