



## Intravascular Papillary Endothelial Hyperplasia in the Nose - Case Report and Review Of Literature

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### Abstract

Intravascular papillary endothelial hyperplasia is a rare vascular neoplastic lesion in the setting of an unusual thrombus formation. Its significance is in its resemblance with angiosarcoma. It is classified into 3 types - Pure form, mixed and extravascular form. In view of its close histological similarity and subtle differences with angiosarcoma, the role of immunohistochemistry has evolved in the diagnosis of this condition in the recent past. Here we discuss one such case in the nasal cavity where the tumor was attached to the middle turbinate and how it was managed. This is probably the 6<sup>th</sup> case report worldwide of such a tumor in the nasal cavity not involving the sinuses.

**Keywords:** Intravascular; Papillary; Endothelial; Hyperplasia; Masson; Tumor; Angiosarcoma

### Level of Evidence

5.

### Introduction

Intravascular papillary endothelial hyperplasia is a benign tumor of rare occurrence in the nasal cavity. It is important to report it, in view of the pathological similarity to angiosarcoma and the potential to over treat this condition. In this paper, we discuss one such case wherein there was a confusion between IPEH and a malignancy and the features which help discriminate them.

### Case Discussion

A 69 year old man presented to our hospital with a 6 month history of right sided nasal obstruction and recurrent epistaxis. He had a 6 month history of using a pseudoephedrine nasal spray for his breathing difficulties. He was a non-smoker and had no allergies. On endoscopic examination, a large vascular lesion was seen in the right nasal cavity.

### Imaging

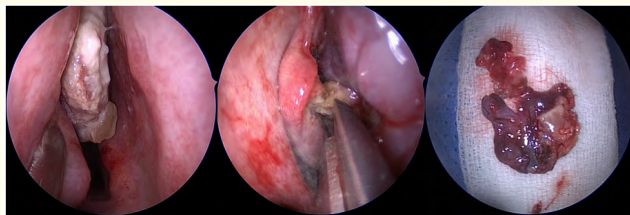
A contrast enhanced computed tomography scan of the paranasal sinuses was then obtained which showed a mass occupying the right nasal cavity and patchy changes within the anterior and posterior ethmoid sinuses on the same side. The frontal, maxillary and sphenoid sinuses on the right and all the sinuses on the left were within normal limits.

Magnetic resonance imaging showed a smooth walled lesion measuring 3.1 cms craniocaudally and 2.5 cms anteroposteriorly, anterior to the middle turbinate, superior to the inferior turbinate and abutted the nasal septum and lateral wall of the nasal cavity. There were no signs of tumor infiltration into the surrounding tissues.

### Management

Patient underwent endoscopic resection of the lesion. The tumor had an attachment to the medial lamella of the middle turbinate and another attachment to the lateral wall of nose just

above the axilla of the middle turbinate (Image 1). A wide resection of the tumor was done and sent for histology.



**Image 1:** A) Tumor in the right nasal cavity; B) Tumor attachment to the medial lamella of the middle turbinate being resected; C) Tumor after being excised.

**Histology**

Expanded mucosa was lined by focally, thin flat stratified squamous epithelium. The expansion was effected by a non-encapsulated, widespread ‘proliferation’ of variously ectatic, meandering, hyperaemic vascular channels lined by a single endothelial layer. Sub-endothelial fibrosis, micro-thrombi and intraluminal hyalinised papillary projections lined by endothelium were focally seen. These projections reflect response (organization) to thrombus formation and were conventionally described as papillary endothelial hyperplasia.

**Follow up**

Patient was followed up for a year with no evidence of recurrence. His epistaxis had completely resolved and he hadn’t had further bleeds ever since. His breathing had improved as well and he was happy with the outcome of the procedure.

**Discussion**

**Nomenclature**

Intravascular papillary endothelial hyperplasia (IPEH) is a benign tumor with a significant vascular component. It was first described by Masson in 1923 as ‘Hemangioendothelioma vegetant intravasculaire’ and later renamed by Clearkin and Enzinger to the present terminology of IPEH in 1976 [1,2].

**Incidence**

IPEH have been described in various locations of the body from head to toe [3-5]. However, nasal cavity IPEH is of extremely rare occurrence and at the time of writing this paper, only 4 reports

(5 cases) have been described worldwide despite an exhaustive literature search [6-9]. Hiroshi., *et al.* reported 91 cases of IPEH from different locations and found that nearly 40% of their specimens were from the upper limb, most commonly the fingers, followed by about 26% in the head and neck region [10].

**Classification - IPEH is classed into 3 types -**

- Type 1 - Also known as the ‘pure’ form or primary IPEH, where in the papillary hyperplasia is seen in the intravascular spaces [11-13].
- Type 2 - Also known as the ‘mixed’ form or secondary IPEH, where in the hyperplasia occurs in the background of a pre-existing lesion such as lymphangiomas, vascular malformations, pyogenic granuloma or hemangiomas of the capillary or cavernous varieties [14,15].
- Type 3 - Is rare and the hyperplasia is extravascular rather than intravascular [16-18].

**Pathogenesis**

Although initially regarded as a neoplastic process as per Masson’s description, it is now thought that it is an unusual thrombus formation [1,19]. There is increasing evidence to support the hypothesis that a thrombus forms the ground substance for the papillary hyperplasia. It is thought that the inflammation and stasis of blood within the thrombus secondary to slowing of blood flow play a vital role in the development of IPEH [10,20].

**Clinical features**

The presenting symptoms of intranasal IPEH from all the previous case reports (including this one) are mentioned along with their frequency of occurrence (Table 1). Nasal obstruction and recurrent epistaxis were the most common presenting features in the case reports of nasal IPEH we studied, which was similar to the presenting features of our case [6-9].

Symptom	No. of cases	Percentage
Nasal obstruction	6/6	100
Epistaxis	4/6	66.67
Rhinorrhea	3/6	50
Headache	2/6	33.33
Hyposmia	1/6	16.67

**Table 1:** Different symptoms with frequency of occurrence in the 5 mentioned case reports including our case report [4-7].

**Histology and immunochemistry**

The tumor consists of papillary fronds within a vascular lumen with a single layered lining of endothelial cells around a fibrous connective tissue core. These cells are based around a thrombotic focus and the papillary growth is towards the vascular lumen. These endothelial cells lack hyperchromasia, nuclear atypia, piling of cells, mitotic foci and necrotic foci which are usually seen in an Angiosarcoma [10]. The papillary fronds are extravascular in the case of the latter which cannot be used as a differentiating feature from the extravascular variant of papillary endothelial hyperplasia.

IPEH is positive for CD 31 and CD 34 -both which are markers of angiogenesis and occasionally SMA [13]. However, it is negative for CD105 (endoglin) which is more commonly seen in Angiosarcoma or pyogenic granuloma. Endoglin is seen in neo-angiogenic tissue and not in pre-existing vascular endothelial cells which backs the theory that IPEH is a reactive process rather than a neoplastic one [19]. CD 105 could be used as a differentiating feature between the extravascular papillary endothelial hyperplasia variety and angiosarcoma (Table 2).

	IPEH	EPEH	Angiosarcoma
Presence of hyperplasia	Intravascular	Extravascular	Extravascular
Thrombus	Present	Present	Absent
Pleomorphism	Absent	Absent	Present
Mitotic activity	Absent	Absent	Present
Necrotic foci	Absent	Absent	Present
Endothelial piling	Absent	Absent	Present
Invasion of tissue	Absent	Absent	Present
CD 105	Absent	Absent	Present

**Table 2:** Differentiating features between intravascular papillary endothelial hyperplasia (IPEH), extravascular endothelial hyperplasia (EPEH) and angiosarcoma.

**Differential diagnosis**

IPEH needs to be differentiated from angiosarcoma, hemangioma, hemangiopericytoma, angiolymphoid hyperplasia with eosinophilia, Kaposi’s sarcoma, spindle cell hamangioma to prevent over management as would be done for a malignant tumor [10,13].

**Management**

Successful management of IPEH by surgery with close follow up has been reported by many previously with minimal chances of recurrence [8,9,11,12]. Recurrence is less with the pure/primary form of IPEH. The mixed/secondary forms and the extravascular forms have higher chances of recurrence [10].

**Conclusion**

IPEH remains a diagnostic dilemma for clinicians and pathologists alike. Clinically it presents as a solitary mass in the nose or sinuses and being a vascular lesion precludes a biopsy in most cases as it may bleed vigorously and could cause significant

blood loss especially if carried out in an outpatient setting which may not be as controlled an environment as in the operating theatre. In case of doubt, a wide excision should be carried out and the specimen sent for histological examination. The patient might need to be followed up closely at least until the histology has been proven to be benign. IPEH needs to be reported due to its close resemblance to aggressive tumors such as angiosarcoma, both clinically and histologically. Microscopic examination by an experienced pathologist with a high index of suspicion and a thorough knowledge of the differentiating features of these various vascular tumors is necessary to ascertain the diagnosis. Immunochemistry could be valid in confirming the diagnosis with conviction.

**Financial Disclosure and Conflicts of Interest**

None of the authors have any financial disclosures or conflict of interest.

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