

## Epithelioid Haemangioendothelioma of Hypopharynx- A Case Report

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**Abstract:** Epithelioid hemangioendothelioma is a rare vascular neoplasm of borderline or low-grade malignant potential uncommonly seen in head and neck region and has not been reported in hypopharynx. We present here a case of epithelioid hemangioendothelioma arising from the hypopharynx in a young female patient who presented with progressive dysphagia for one year and difficulty in breathing for one month. This is first reported case in published English literature to the best of our knowledge. Recognition of this borderline entity is necessary because of its potential formal malignant transformation and recurrence. Presently, wide excision and regular clinical follow-up is an ideal treatment protocol. Role of other therapeutic modalities such as chemotherapy and/ or radiotherapy is not yet well established.

**Keywords:** Epithelioid hemangioendothelioma, vascular neoplasm, hypopharynx

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### I. Introduction

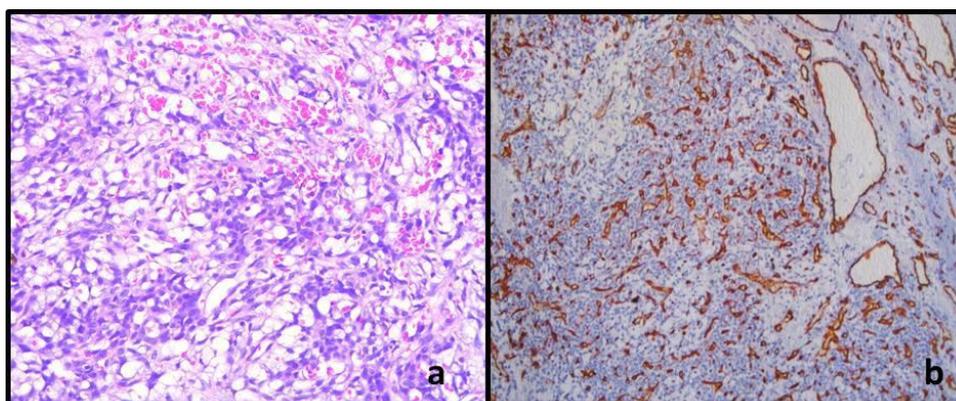
Epithelioid hemangioendothelioma is a rare vascular tumour with low grade malignant potential commonly seen in the soft tissues of the extremities, cases have been reported in lung, liver, bone and skin and few cases in head and neck region. [1,2] This lesion is usually asymptomatic, can occur in any age group and has female predilection with a female-to-male ratio of 2.5:1. We present here a case of epithelioid hemangioendothelioma of the hypopharynx. This is the first reported case of an epithelioid hemangioendothelioma occurring in the hypopharynx.

### II. Case Report

A 25 year-old female patient presented with complaint of progressive difficulty in swallowing for one year. She also had difficulty in breathing during exertion for one month. There was no history of throat pain, cough, fever, stridor, hoarseness of voice, hemoptysis, loss of appetite, weight loss, associated comorbidities or substance abuse. Oral cavity and oropharyngeal examination were normal. Hopkins's laryngoscopy – revealed a globular mass projecting into the supraglottis from the lateral wall of the right pyriform fossa (Fig 1a). Right aryepiglottic fold and false cord were obscured by the mass. Both vocal cords were normal. Epiglottis, pharyngoepiglottic folds, vallecula and base of tongue were normal. There was no palpable mass in the neck. Rest of the ear, nose and systemic examination revealed no abnormality. Routine blood, urine, chest X-ray, ECG investigations were within normal limits. Patient underwent microlaryngoscopy and excision biopsy under general anaesthesia. Post-op period was uneventful. Patient was started on oral feeds on the same post-operative day. Follow up laryngoscopy on tenth post-operative day revealed some slough in the right pyriform fossa. Grossly, the mass was greyish- white and measured 2.5 x 2 cm. (Fig 1b). H&E stained sections showed round to polygonal tumor cells arranged in cords and small nests having prominent cytoplasmic vacuolisation. Intraluminal erythrocytes were noted in many of the vacuoles, reminiscent of primitive vascular channels which were confirmed by CD 34 immunostain. There was no evidence of nuclear atypia, abnormal mitosis or areas of necrosis. Based on the microscopic findings of H&E and immunostaining, the lesion was diagnosed as epithelioid haemangio-endothelioma of pyriform fossa (Fig 2 a, b). Patient has been follow up for 1 year without any evidence of loco-regional recurrence or dysphagia (Fig 1c).



**Fig. 1:** a) Hopkins Telescopic image Pre-operative; b) Excised Specimen; c) Hopkins Telescopic image one year post-operative.



**Fig 2:** Histopathological image – a) H&E staining showing numerous proliferating capillaries lined by plumped endothelium arranged in lobules and horn-like pattern; (400 X magnification, hematoxylin & eosin). b) Immunohistochemical staining showing CD34 positive pericytic cells.

### III. Discussion

Epithelioid hemangioendothelioma is a rare neoplasm of vascular origin initially described by Weiss and Enzinger in 1982. [3] Vascular tumours composed of histiocytoid or epithelioid endothelial cells are divided into epithelioid hemangioma, epithelioid hemangioendothelioma and epithelioid angiosarcoma. However, owing to the overlapping histologic features they are also considered as a continuous spectrum of lesions, where epithelioid hemangioendothelioma represents a borderline or low-grade malignant variant. [4] It is commonly seen in liver, spleen, bone, skin, heart, soft tissues and vascular system. [5] In head and neck region it is very rarely seen. Cases have been reported in oral cavity, thyroid gland, submandibular area, neck, scalp, larynx, parapharyngeal space, parotid gland and mandible. Most common site in head neck region is at or below the level of the mandible and submandibular region. [6,7,8] It can occur in any age group and is commoner in females. [9] Histopathological findings of epithelioid hemangioendothelioma are round or spindle-shaped epithelioid cells with pale cytoplasm. [6] Tumor cells have vacuolisations containing red blood cells. On Immunohistochemistry they are positive for endothelial cell markers (CD31, CD34 and factor VII-related antigen). [6,10] Due to noticeable malignant potential wide local excision and regular follow up is the preferred management option. [7] Our patient was a young female patient with lesion in hypopharynx, showing the typical histopathological picture with CD34 positivity on immunohistochemistry. She underwent wide excision with monthly follow-up for one year without any evidence of recurrence. This is the first reported case of epithelioid hemangioendothelioma arising from hypopharynx.

### IV. Conclusion

Epithelioid hemangioendothelioma (EH) is a rare tumour of vascular origin which usually occurs in soft tissues. Presence of this tumour in the hypopharynx may be misdiagnosed because of their unusual presentation. Wide local excision with close clinical follow-up appears to be the treatment of choice for these tumours because of their risk of recurrence. Role of chemotherapy/radiotherapy are still controversial and needs further study.

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