Case Report

# Epithelioid hemangioendothelioma of hypopharynx: A rare presentation

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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 the hypopharynx. We present here a case of epithelioid hemangioendothelioma arising from the hypopharynx in a young female patient who presented with progressive dysphagia for 1-year and difficulty in breathing for 1-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 for malignant transformation and recurrence. A wide excision and regular clinical follow-up would be an appropriate treatment protocol. The role of other therapeutic modalities such as chemotherapy and/or radiotherapy is not yet well established.

**Key words:** Epithelioid hemangioendothelioma, hypopharynx, vascular neoplasm

## INTRODUCTION

Epithelioid hemangioendothelioma is a rare vascular tumor 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 epithelioid hemangioendothelioma in hypopharynx.

## CASE REPORT

A 25-year-old female patient presented with a complaint of progressive difficulty in swallowing since 1-year. History revealed the difficulty in breathing during exertion since 1-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. On examination, no abnormality was detected in relation to the oral cavity and oropharynx. Hopkins's laryngoscopy revealed a globular mass projecting into the supraglottis from the lateral wall of the right pyriform fossa [Figure 1a]. Right aryepiglottic fold and false cord were obscured by the mass. Both vocal cords were normal. The epiglottis, pharyngoepiglottic folds, vallecula, and base of tongue were also normal. There was no palpable mass in the neck. The rest of the ear, nose, and systemic examination revealed no abnormality. Routine blood, urine, chest X-ray, and electrocardiogram investigations were within normal limits. The patient underwent microlaryngoscopy and excision biopsy under general anesthesia. The mass was grayish-white and measured 2.5 cm × 2 cm [Figure 1b]. Postoperative

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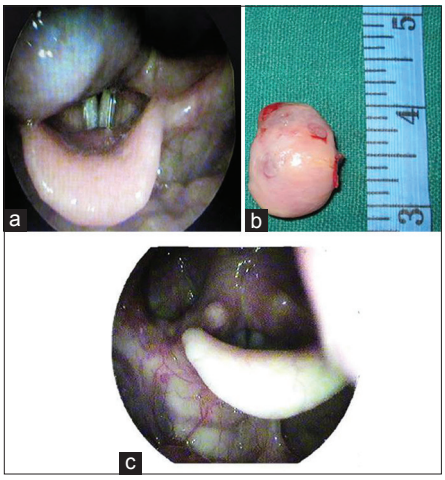


**Figure 1:** (a) Hopkins telescopic image preoperative; (b) excised specimen; (c) Hopkins telescopic image 1-year postoperative

period was uneventful. Patient was started on oral feeds on the same postoperative day. Follow-up laryngoscopy on the 10th postoperative day revealed some slough in the right pyriform fossa. Hematoxylin and eosin (H and E) stained sections showed round to polygonal tumor cells arranged in cords and small nests having prominent cytoplasmic vacuolization. Intraluminal erythrocytes were noted in many of the vacuoles, reminiscent of primitive vascular channels that were confirmed by CD34 immunostain. There was no evidence of nuclear atypia, abnormal mitosis or areas of necrosis. Based on the microscopic findings of H and E and immunostaining, the lesion was diagnosed as epithelioid hemangioendothelioma of pyriform fossa [Figure 2a and b]. Patient has been on follow-up since 1-year without any evidence of loco-regional recurrence or dysphagia [Figure 1c].

## DISCUSSION

Epithelioid hemangioendothelioma is a rare neoplasm of vascular origin initially described by Weiss and Enzinger in 1982.[3] Vascular tumors composed of histocytoid 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] It is very rarely encountered in the head and neck region. Cases have been reported in the oral cavity, thyroid gland, submandibular area, neck, scalp, larynx, parapharyngeal space, parotid gland, and mandible. The most common site in the head neck region is at

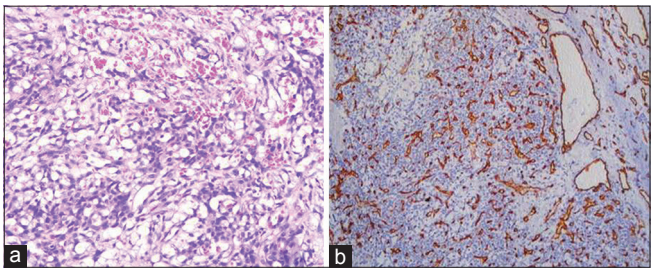


**Figure 2:** Histopathological image (a) H and E staining showing numerous proliferating capillaries lined by plump endothelium arranged in lobules and horn-like pattern; (H and E, ×400). (b) Immunohistochemical staining showing CD34 positive pericytic cells

or below the level of the mandible and submandibular region.[6-8] It can occur in any age group and is common in females.[9] Our was a young female patient with a lesion in the hypopharynx. This is the first reported case of epithelioid hemangioendothelioma to arise from hypopharynx.

Histopathological findings of epithelioid hemangioendothelioma suggest round or spindle-shaped epithelioid cells with pale cytoplasm.[6] Tumor cells show vacuolizations containing red blood cells. On immunohistochemistry, they are positive for endothelial cell markers (CD31, CD34 and factor VII-related antigen).[6,10] The present case also shows a typical histopathological picture with CD34 positivity on immunohistochemistry. Due to noticeable malignant potential, wide local excision and regular follow-up are the preferred management option.

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### Conflicts of interest
There are no conflicts of interest.

## REFERENCES

1. van Haelst UJ, Pruszczynski M, ten Cate LN, Mravunac M. Ultrastructural and immunohistochemical study of epithelioid hemangioendothelioma of bone: Coexpression of epithelial and endothelial markers. Ultrastruct Pathol 1990;14:141-9.
2. Tsuneyoshi M, Dorfman HD, Bauer TW. Epithelioid hemangioendothelioma of bone. A clinicopathologic, ultrastructural, and immunohistochemical study. Am J Surg Pathol 1986;10:754-64.
3. Weiss SW, Enzinger FM. Epithelioid hemangioendothelioma: A vascular tumor often mistaken for a carcinoma. Cancer 1982;50:970-81.
4. Tsang WY, Chan JK. The family of epithelioid vascular tumors. Histol Histopathol 1993;8:187-212.
5. McKenzie ML. Epithelioid hemangioendothelioma of the wrist. Plast Reconstr Surg 1985;76:781-3.
6. Ellis GL, Kratochvil FJ 3rd. Epithelioid hemangioendothelioma of the head and neck: A clinicopathologic report of twelve cases. Oral Surg Oral Med Oral Pathol 1986;61:61-8.

7. Lee JC, Lee BJ, Wang SG, Kim HW. Epithelioid haemangioendothelioma in the parapharyngeal space. J Laryngol Otol 2006;120:505-7.
8. Boscaino A, Errico ME, Orabona P, Tornillo L, Staibano S, Donofrio V, *et al.* Epithelioid hemangioendothelioma of the larynx. Tumori 1999;85:515-8.
9. Chi AC, Weathers DR, Folpe AL, Dunlap DT, Rasenberger K, Neville BW. Epithelioid hemangioendothelioma of the oral cavity: Report of two cases and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2005;100:717-24.
10. Mentzel T, Beham A, Calonje E, Katenkamp D, Fletcher CD. Epithelioid hemangioendothelioma of skin and soft tissues: Clinicopathologic and immunohistochemical study of 30 cases. Am J Surg Pathol 1997;21:363-74.