Case Report

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Epitheloid hemangio-endothelioma of right aryepiglottic fold: a rare case report with review of literature

Alpesh D. Fefar*, Paresh J. Khavdu, Mukesh N. Dodia, Sejal N. Mistry, Manish R. Mehta

Department of Otorhinolaryngology (ORL), P. D. U. Medical College, Rajkot, Gujarat, India

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*Correspondence: Dr. Alpesh D. Fefar,

E-mail: alpay.dr@gmail.com

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ABSTRACT

Epitheloid hemangio-endothelioma is a very rare tumour of intermittent malignancy of vascular origin, having a tendency to recur with rare incidence of metastasis. The tumour is intermediate between haemangioma and angiosarcoma, mainly affecting liver, lung as well as bones, skin, penis, ovary, scalp, or any part of the body. Internet search was made with the key words epitheloid hemangio-endothelioma and epitheloid hemangio-endothelioma of Larynx, since now only single case has been reported from larynx involving subglottis. Hence we report this rare entity with involvement of the larynx (Sub site: Rt. Aryepiglottic fold) describing clinical and histopathological characteristic. This is perhaps the first case of epitheloid hemangio-endothelioma involving aryepiglottic fold.

Keywords: Epitheloid hemangio-endothelioma, Aryepiglottic fold, Larynx

INTRODUCTION

Epitheloid hemangio-endothelioma is a very rare tumour of intermittent malignancy of vascular origin, having a tendency to recur with rare incidence of metastasis. The tumour is intermediate between haemangioma and angiosarcoma, mainly affecting liver, lung as well as bones, skin, penis, ovary, scalp, or any part of the body. Internet search was made with the key words epitheloid hemangio-endothelioma and epitheloid hemangio-endothelioma of larynx, since now only single case has been reported from larynx involving subglottis. Hence we report this rare entity with involvement of the larynx (Rt. aryepiglottic fold) describing clinical and histopathological characteristic with review of literature. This is the first case of epitheloid hemangio-endothelioma involving aryepiglottic fold.

CASE REPORT

A male patient 35 years old presented in our department with chief complains of difficulty in swallowing to solids

increasing progressively, dyspnoea, hoarseness of voice for 5 months. He underwent laryngoscopy which is suggestive of right pyriform fossa mass, followed by Contrast CT scan of neck which confirmed right pyriform fossa soft tissue density lesion of 24 x 21 x 27 mm with involvement of right aryepiglottic fold. No cervical noted. lymphadenopathy Routine haematological investigations were within normal limits. Patient was planned for micro laryngeal LASER pedunculated greyish white solid mass obscuring right pyriform fossa was seen which was arising from right aryepiglottic fold, excised with diode laser, sent for histopathological examination. The histopathology report states presence of histiocytoid, epitheloid, and spindle tumour cells arranged predominantly perivascularly having vacuolated cytoplasm at places with focal myxoid area and moderate atypia.

Final diagnosis of epitheloid hemangio-endothelioma was made. At 9 months follow up patient is well without any complains.

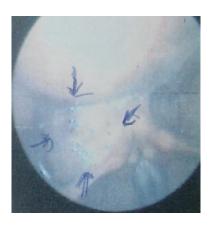


Figure 1: Endoscopy picture. (Arrows showing mass in Rt. pyriform fossa).

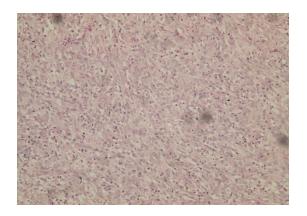


Figure 2: Histo-pathological features of epitheloid hemangio-endothelioma, H & E staining.

DISCUSSION

Epitheloid hemangio-endothelioma is a very rare tumour of intermittent malignancy of vascular origin, have a tendency to recur with rare incidence of metastasis. EHE is a distinct entity that was first described by Weiss and Enzinger in 1982² and studied in detail by Ishak et al. in 1984 who described 32 cases.³ However, a decade and a half later in 1999, Makhlouf et al. reported the largest series of 137 cases.⁴ EHE is a rare vascular tumor that is intermediate in morphological features between hemangioma and conventional angiosarcoma. EHE has been reported in the liver, lung, gastrointestinal tract, head, neck, central nervous system, heart, bone, and skin.⁵⁻¹³ Liver and lung lesions are more common in females, whereas tumors of the bone and soft tissues have an equal sex distribution.¹⁴ Epitheloid hemangioendothelioma rarely affects head and neck, larynx still rare. Single case had been reported in the literature of epitheloid hemangio-endothelioma of larynx. Immunohisochemical analysis reveals that epitheloid hemangioendothelioma expressed platelet endothelial cell adhesion molecule 1 transmembrane glycoprotein (CD31), cytokeratin CD34, factor VIII-related antigen, vimentin, and occasionally AE1/AE3. Tumor cells are negative for epithelial membrane antigen and carcinoembryonic antigen. Treatment is wide surgical excision.

CONCLUSION

This case report is unique as it documents the clinical and pathological features, surgical and postoperative treatment, and long-term follow-up required for a patient with epitheloid hemangio-endothelioma. Although it is very rare tumour, it should be kept in differential diagnosis as treatment options vary.

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