

CASE REPORT

Pleomorphic Sarcoma of Larynx

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ABSTRACT

Background: Malignant fibrous histiocytoma (MFH) is a malignant type of sarcoma with uncertain origin and it may arise from bone and soft tissue. In 3% cases it involves head and neck and it is rare in larynx.

Case report: We present a rare case of a 55-year-old male diagnosed as pleomorphic sarcoma (PS) of larynx.

Conclusion: Pleomorphic sarcoma, commonest subtype of MFH, should be kept in mind as a differential diagnosis in aggressive tumors of larynx.

Clinical significance: Pleomorphic sarcoma of larynx may be diagnosed as rare tumor of larynx and may need aggressive treatment.

Keywords: Giant cells, Larynx, Malignant fibrous histiocytoma, Pleomorphic sarcoma.

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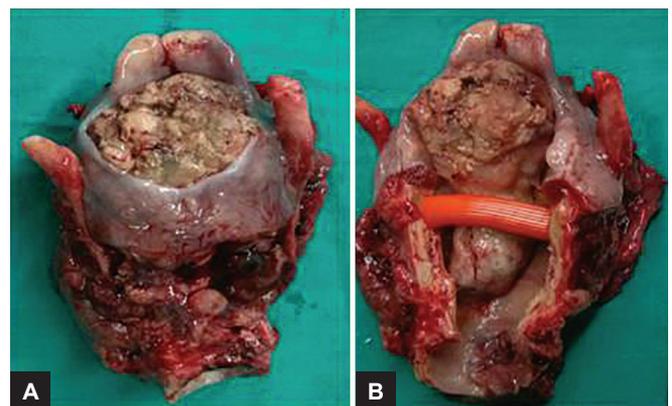
BACKGROUND

Sarcoma is a cancer that arises from mesenchymal cells. Malignant fibrous histiocytoma is a malignant type of sarcoma with uncertain origin and it may arise from bone and soft tissue. In 3% of cases, it involves the head and neck.¹ Nose, paranasal sinuses, nasopharynx are the common site in head and neck and it is rare in larynx.² There are five subtypes of MFH, and PS is the most common subtype.³

We present here a rare case of PS involving larynx.

CASE REPORT

A 55-year-old male patient presented with chief complaint of hoarseness for 7 months. He also had history of progressively increasing respiratory problem for which tracheostomy was done 1 month back. There was no history of dysphagia, neck swelling, pain, and bleeding from mouth. Indirect laryngoscopy showed ulceroproliferative mass filling endolarynx completely with bilateral immobile vocal cords. The hypopharynx and neck were normal on examination. Direct laryngoscopy showed involvement of bilateral aryepiglottic, false vocal cords, true vocal cords, arytenoids with subglottic extension, and biopsy from lesion was squamous cell carcinoma. Contrast-enhanced computed tomography of neck showed soft tissue density completely filling endolarynx in the region of supraglottis, glottis, and subglottis. After informed consent, patient was planned for wide-field laryngectomy and tumor was found filling endolarynx completely (Fig. 1A). After cutting cricoid cartilage, tumor was found adhered at anterior part of larynx in the region of supraglottis, glottis, and subglottis (3 cm extension) (Fig. 1B). Postoperative period was uneventful, and oral trial was given on 9th postoperative day. The final histopathology report showed presence of anaplastic round to oval to spindle cells in interlacing bundles, fascicles, and whorls with high degree of anaplasia with bizarre giant cells suggestive of PS (Fig. 2). The patient underwent postoperative radiotherapy for total dose of 60 Gy in 30 fractions over 6 weeks. Now patient is disease-free with regular follow-up from last 6 months.



Figs 1A and B: Tumor filling whole of endolarynx was found attached with anterior part of supraglottis, glottis and subglottis (3 cm extension)

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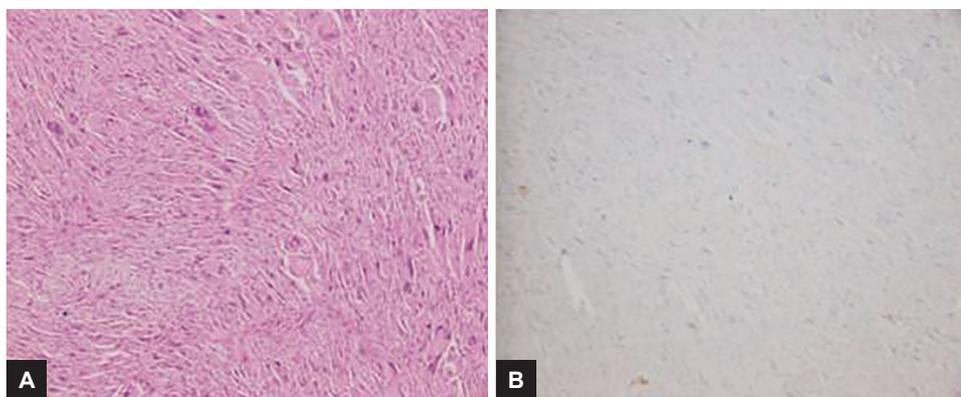
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Figs 2A and B: (A) Hematoxylin and eosin stain, 20×: tumor is composed of malignant spindle cells arranged in whorls and interlacing fascicles with tumor giant cell; and (B) cytokeratin negative

DISCUSSION

Malignant fibrous histiocytoma term was coined by Kauffman and Stout in 1961.⁴ Pleomorphic sarcoma is the most common type of sarcoma in late adult age, i.e., approximately 50 to 70 years of age, although it can appear at any age. It is more common in lower and upper extremities with slight male preponderance. Pleomorphic sarcoma is also reported in retroperitoneal region, lungs, liver, kidneys, bladder, scrotum, vas deferens, heart, aorta, stomach, small intestine, orbit, central nervous system, paraspinal area, dura mater, facial sinuses, nasal cavity, oral cavity, nasopharynx, and soft tissues of the neck.⁵⁻⁷ There are few case reports in literature where PS involved larynx.^{8,9}

Glottis is more common site in males and subglottis is more common in females for primary MFH, whereas in our case it was transglottic.

The clinical symptoms for laryngeal PS are dyspnea, hoarseness, stridor, dysphagia, hemoptysis, and diagnosis established after removal,^{10,11} as in our case. On indirect laryngoscopy, one can find pedunculated or polypoidal mass, whereas ulceration is common finding for laryngeal carcinoma similar to our case. Magnetic resonance imaging is the investigation of choice due to its superior soft tissue delineation and cartilage invasion by tumor. For definitive diagnosis, biopsy is required; and to establish subtype of sarcoma, immunohistochemistry is required.¹²

An *en bloc* resection is the treatment of choice.^{13,14} Small lesions can be managed by excision under direct laryngoscopy, but for large lesions wide-field laryngectomy is required. The indications for adjuvant radiotherapy are high-grade tumor, positive surgical margins, large tumor size (>5 cm), and recurrent disease.¹⁵ The differentiation of tumor, size, vascular invasion, and metastasis are the prognostic factors for survival and the 5-year survival is approximately 60%.¹⁶

CONCLUSION

Pleomorphic sarcoma is a rare tumor of larynx and is aggressive in nature. This differential diagnosis should be kept in mind while assessing carcinoma of larynx.

CLINICAL SIGNIFICANCE

Pleomorphic sarcoma of larynx may be diagnosed as rare tumor of larynx and may need aggressive treatment.

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