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CASE REPORT

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MYXOFIBROSARCOMA of the Subglottis - A Rare Case Report

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Introduction

Larynx is an essential midline structure situated at the meeting point of digestive and respiratory passage in the neck. As it is a composite Osseo-fibro cartilaginous structure, it comprises of various cell types. Laryngeal cancers are an important cause of morbidity and mortality, with squamous cell carcinoma being the most common histological type [1]. Sarcomas arising in the larynx are uncommon, representing less than 1% of malignant laryngeal neoplasms, out of which the commonest are chondrosarcomas [2-6]. Myofibroblastic sarcoma, and myxofibrosarcoma have an incidence of less than 2% of all malignancies in the larynx. It accounts for approximately 5% of all sarcomas of the head and neck [7]. Myxofibrosarcoma (MFS) is a soft tissue sarcoma located mainly in the subcutaneous soft tissues of extremities. It is a fibroblast-derived sarcoma, accounting for roughly 5–10% of all soft tissue malignant tumours. About 77% of MFS cases occur in the extremities, commonly in the upper extremities. Reports of MFS in the trunk (12%), retroperitoneum or mediastinum (8%), abdominal wall and heart are also there in the literature [8]. MFS of the head and neck is quite rare [8]. As to our best knowledge, we are reporting the first case of myxofibrosarcoma arising from the subglottis region of the larynx.

Case Report

A 57-year-old male presented to ENT clinic with complaints of blood-tinged sputum since one month and noisy breathing since 15 days. Patient gave history of smoking for the past 20 years. Rest of the history was insignificant. On examination, patient had biphasic stridor. Laryngoscopy showed a proliferative growth with streaks of blood in the sub glottis with normal movements of vocal cords on phonation and respiration. Rest of the examination was normal.

The patient was taken up for emergency tracheostomy and direct laryngoscopy and biopsy. Under direct vision, a friable growth was noted in the sub glottis arising from the right lateral wall. Sufficient tissue was taken and sent for histopathological examination (HPE).

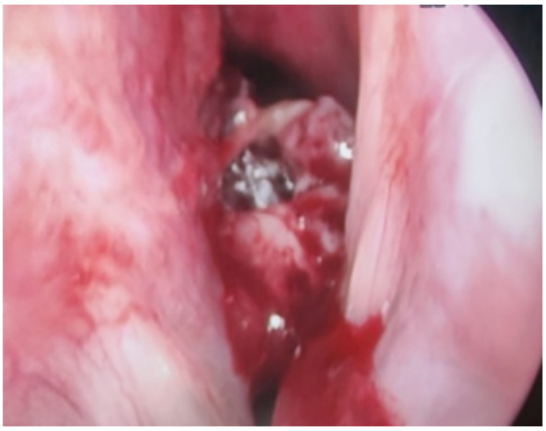

Figure 1: Video laryngoscopic view of the lesion

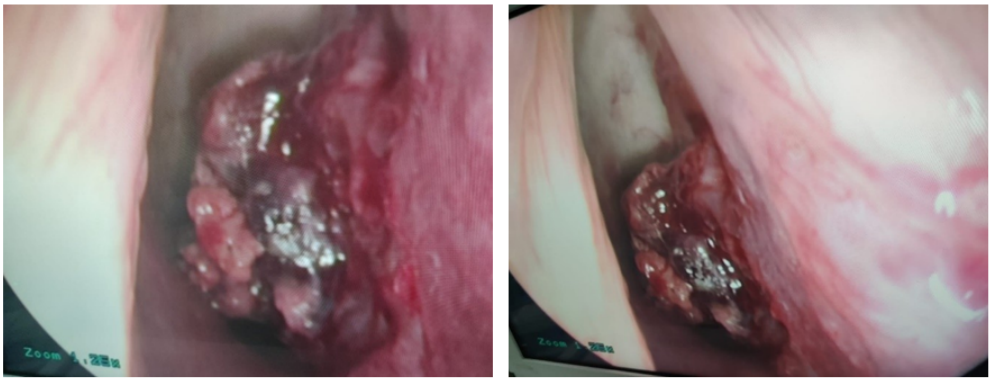

Figure 2: Direct laryngoscopic view of the lesion

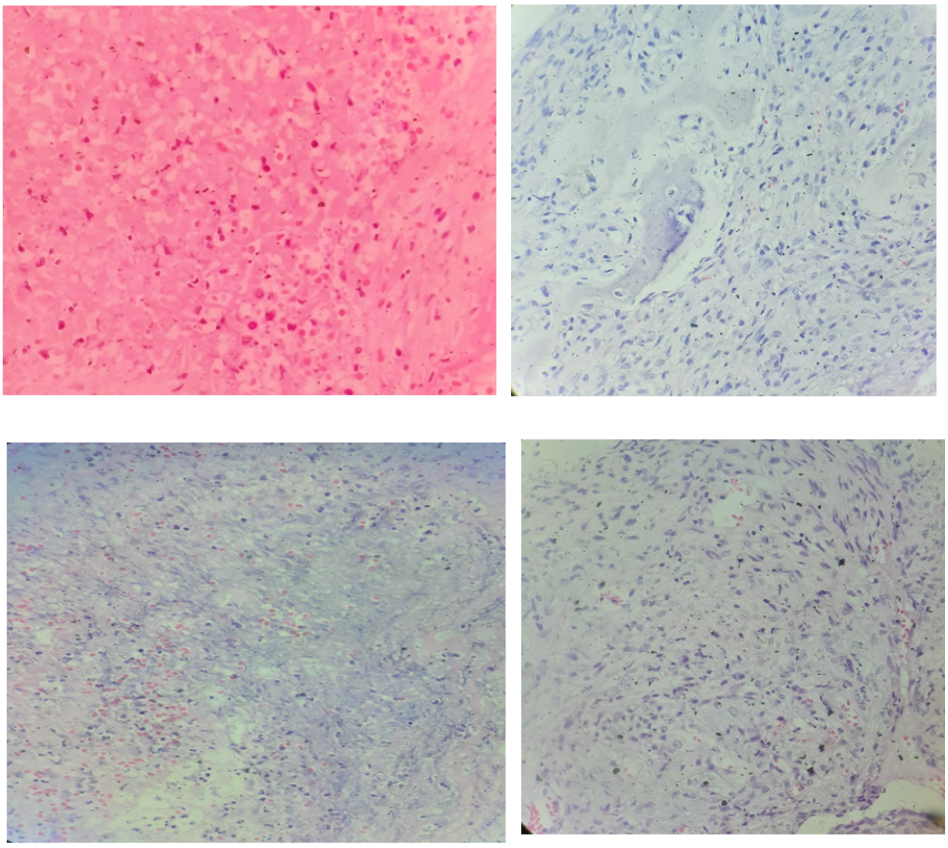

Figure 3: Histopathological examination of the lesion

HPE showed myxofibrosarcoma with areas of necrosis and was reported as LOW- GRADE MYXOFIBROSARCOMA SUBGLOTIS.

On CECT, a heterogeneously enhancing lesion epicentered at right subglottic region was noted with extensions, anteriorly to the preepiglottic fat pad and the lamina of thyroid cartilage with loss of fat planes. Posteriorly the lesion showed loss of fat planes with cervical oesophagus. Laterally, it was eroding into the cricoid cartilage on the right side. Medially it was crossing midline and causing severe subglottic luminal narrowing at level of C5 vertebra. Superiorly, it was extending up to arytenoid cartilage (up to level of inferior end plate of C4 vertebra) with erosions of arytenoid cartilage and suspicious extension to the true vocal cord. No extension to false vocal cords was noted. Inferiorly, it was extending up to superior end plate of C6 vertebra.

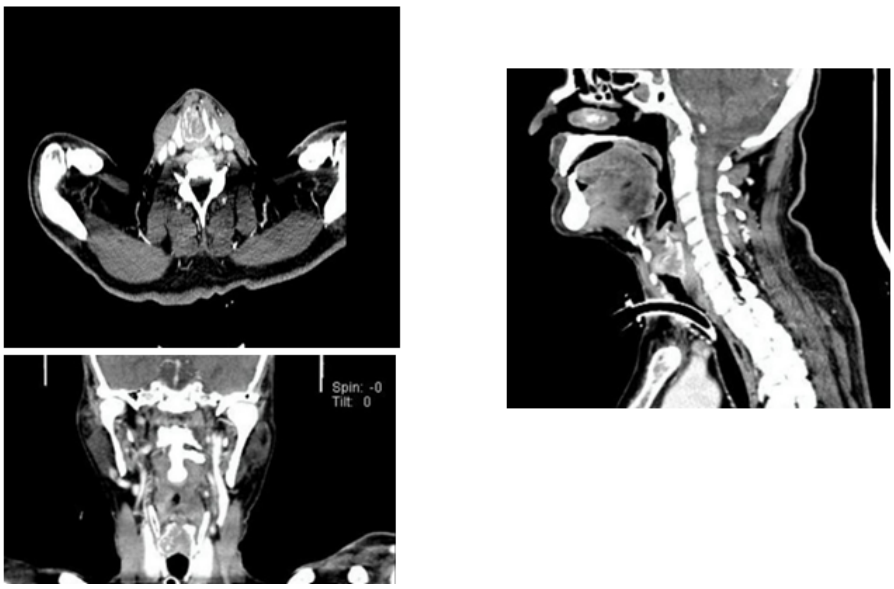

Figure 4: CECT scan of the patient

TNM staging was T4aN0M0. As the tumour was very extensive involving the structures as described in the CT and as the primary modality of treatment in MFS is surgery, patient was subjected to total laryngectomy. Patient was not given any adjunct radiotherapy due to the tumour being low grade on HPE. Patient was lost to follow up after 6 weeks of surgery.

Discussion

Primary sarcomas in larynx are rare and are associated with diagnostic and treatment challenges. Among laryngeal sarcomas, chondrosarcoma is the most common, but other types such as leiomyosarcoma, rhabdomyosarcoma, osteosarcoma, liposarcoma, and synovial sarcoma, have also been described [9]. Sarcomas of the larynx may develop in any portion of the organ. Associated symptoms depend on the size and location of the tumour, with the most common being hoarseness and dyspnea [1]. Though previously, few sarcomas have been reported to have arisen from the subglottis, we are reporting the first case of myxofibrosarcoma arising from this region [10,11]. The term myxofibrosarcoma (MFS) is proposed as a synonym for myxoid MFH and covers a spectrum of malignant fibroblastic lesions, which have cellular distribution, pleomorphism of the nucleus, and mitotic activity that ranges from a less cellular lesion with minimal cytologic atypia to a greater cellular lesion with pronounced atypical features [12,13]. Myxofibrosarcoma (MFS) is commonly seen in elderly patients, more so in 6th to 8th decade of life [14]. MFS can be either low grade tumour with low metastatic potential or a high-grade tumour with high metastatic potential of 20-35%. Metastasis is more common in lungs and bones [15]. Surgical excision is taken as the mainstay of therapy. Radiation therapy continues to play an important role when surgery is unfeasible or as an adjunctive either preceding or following surgery to treat aggressive tumours or positive margins [16]. In a randomized controlled trial (RCT) involving soft tissue sarcomas of multiple subtypes, radiotherapy usage was

found to reduce the recurrence rate of the sarcomas [17,18] Till date, there are no RCTs specifically evaluating MFS treated by radiotherapy. In MFS patients with distant metastasis, chemotherapy is commonly used. The combination of anthracycline and isocyclophosphamide have been reported to increase the survival rate in MFS patients [19]. Though the treatment regime is similar in treatment of all laryngeal sarcomas, we are citing this case to throw light on the possible existence of such a rare malignancy arising from an uncommon site and for epidemiological purposes. The local recurrence rate for MFS is not based on the histopathological grade. Generally MFS are low grade lesions and prognosis of this subtype is generally good. When it comes to recurrence, both types of MFS have similar rates. Low-grade MFS, however, can change into a high-grade MFS after recurrence [20]. The local recurrence rate for MFS is of about 16–57%. Multiple recurrence is also noted and estimated to be between 25 and 52%. It has a 5-year overall survival rate between 61 and 77%. [21] Patients with MFS have to be followed long time for tumour recurrence. Recurrence has been reported to occur as early as 2 months post surgery and as far out as eight years postoperatively. 27 months is the reported median recurrence time [21]. Unfortunately, we lost our patient to follow up.

Conclusion

This is the first case of myxofibrosarcoma arising from the subglottis being reported in the literature. Sarcomas in the larynx are rare and are of various subtypes. It is important to be aware of the various clinical presentations, different possible histopathological diagnosis of laryngeal masses including the one reported by us so that appropriate diagnosis is made and treatment can be initiated at the earliest.

References

1. Covello R, Licci S, Pichi B, Spriano G, Vidiri A et al. (2011) Low- grade myofibroblastic sarcoma of the larynx. Int J Surg Pathol, 19: 822-6.

2. Fernández-Aceñero MJ, Larach F, Ortega-Fernández C (2009) Non-epithelial lesions of the larynx: review of the 10-year experience in a tertiary Spanish hospital. Acta Otolaryngol, 129: 108-12.

3. Karatayli-Ozgursoy S, Bishop JA, Hillel AT, Akst LM, Best SR. Non-epithelial tumors of the larynx: a single institution review. Am J Otolaryngol, 37: 279-85.

4. Thompson LD, Gannon FH (2002) Chondrosarcoma of the larynx: a clinicopathologic study of 111 cases with a review of the literature. Am J Surg Pathol, 26: 836-51.

5. Friedman AD, Burns JA, Lutch MJ, Zeitels SM. Submucosal neoplasms of the laryngeal introitus. J Laryngol Otol. 2012 Jul;126(7):706-13. doi: 10.1017/S0022215112000928. Epub 2012 May 25. PMID: 22624973

6. AbdullGaffar B, Keloth T Laryngeal sarcomas (2018) A case series of 5 cases. Ann Diagn Pathol, 37: 35-41.

7. Cao X, Liu J, Zheng Y, Li Q, Teng Y, Li Y (2012) Simultaneous squamous cell carcinoma with primary malignant fibrous histiocytoma of the larynx: a case report. Mol Med Rep, 5: 971-3.

8. Li Z, Liu X, Zhang Q, Zhang J, Huang M, Liu S (2020) Myxofibrosarcoma of the mandible: a case report and review of the literature. BMC Oral Health, 20: 113.

9. Imai Y, Sugawara Y, Okazaki M, Harii K (2000) Low grade myxofibrosarcoma in the orbit: A case report. Japanese Journal of Plastic Surgery, 43: 401-9.

10. Sridhar Reddy D, Shobhan Babu A, Lenin A (2007) Synovial sarcoma of larynx-a rare site. Indian J Otolaryngol Head Neck Surg, 59: 51-2.

11. Anithakumari AM, Somashekhar SP, Mondal SK (2015) Neurofibrosarcoma of the Subglottis-A Rare Case Report. Indian J Surg Oncol, 6: 113-5.

12. Angervall L, Kindblom LG, Merck C. Myxofibrosarcoma (1977) A study of 30 cases. Acta Pathol Microbiol Scand A, 85A:127-40.

13. Fletcher CDM, Unni KK, Mertens F (Eds) (2002) Pathology and Genetics of Tumours of Soft Tissue and Bone. Lyon: IARC Press.

14. Jo VY, Fletcher CD (2014) WHO classification of soft tissue tumours: an update based on the 2013 (4th) edition. Pathology, 46: 95-104.

15. Quimby A, Estelle A, Gopinath A, Fernandes R (2017) Myxofibrosarcoma in Head and Neck: Case Report of Unusually Aggressive Presentation. J Oral Maxillofac Surg, 75: 2709.e1-2709.e12.

16. Peng KA, Grogan T, Wang MB (2014) Head and neck sarcomas: analysis of the SEER database. Otolaryngol Head Neck Surg, 151: 627-33.

17. Yang JC, Chang AE, Baker AR, et al. (1998) Randomized prospective study of the benefit of adjuvant radiation therapy in the treatment of soft tissue sarcomas oftheextremity. JClinOncol, 16: 197-203.

18. Beane JD, Yang JC, White D, et al. (2014) Efficacy of adjuvant radiation therapy in the treatment of soft tissue sarcoma of the extremity: 20-year follow-up of a randomized prospective trial. Ann Surg Oncol, 21: 2484–9.

19. Colia V, Fiore M, Provenzano S, et al. (2017) Activity of anthracycline-and ifosfamide-based chemotherapy in a series of patients affected by advanced myxofibrosarcoma. Clin Sarcoma Res, 7: 1-16.

20. Huang HY, Lal P, Qin J, et al. (2004) Low-grade myxofibrosarcoma: a clinicopathologic analysis of 49 cases treated at a single institution with simultaneous assessment of the efficacy of 3-tier and 4-tier grading systems. Hum Pathol, 35: 612–21.

21. Sanfilippo R, Miceli R, Grosso F, et al. (2011) Myxofibrosarcoma: prognostic factors and survival in a series of patients treated at a single institution. Ann Surg Oncol, 18: 720-5.