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Inflammatory Myofibroblastic Tumour of Larynx – An Unusual Cause of Airway Obstruction

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Abstract: *Inflammatory myofibroblastic tumours (IMT) or plasma cell granuloma of the larynx is a rare lesion of larynx. There is a wide variability in the clinical manifestations, histologic features, and prognosis of IMTs depending on their exact site of occurrence and age group affected. We here present a case of huge laryngeal IMT, its diagnosis, management & brief review of literature. Our case illustrates the challenges raised by large laryngeal IMTs, one of the largest ever reported*

Keywords*: Inflammatory Myofibroblastic tumour, Plasma cell Granuloma, Inflammatory pseudotumour, larynx, laryngofissure.*

I. Introduction

Inflammatory myofibroblastic tumours (IMT) or plasma cell granuloma of the larynx is a rare lesion of larynx, first described by Keen et al [1] in 1986. Wenig et al [2] proposed the term IMT Many issues still remain relative to the entire group of inflammatory myofibroblastic (pseudo) tumours, not the least of which is whether these are reactive lesions or neoplastic proliferations. There is a wide variability in the clinical manifestations, histologic features, and prognosis of IMTs depending on their exact site of occurrence and age group affected. We here present a case of huge laryngeal IMT, its diagnosis, management & brief review of literature. Our case illustrates the challenges raised by large laryngeal IMTs, one of the largest ever reported.

II. Case report

A 19 year old girl presented to otorhinolaryngolology department of Gauhati Medical College and Hospital with left sided neck swelling for 4 years and occasional mild difficulty in swallowing for 2 years. There was no history of smoking, trauma or previous neck surgeries. On examining, a smooth, firm, non tender swelling of size 5x3x2 cm over upper aspect of left side of neck which is mobile in nature but not in relation to deglutition and breathing. Fibreoptic laryngoscopy showed a smooth mass arising from left aryepiglottic fold (AEF) with a broad pedicle (Figure 1).

MRI revealed a smooth hyperintense polypoidal lesion involving the left aryepiglottic fold plunging into the larynx causing significant narrowing of the same, with caudal extension upto the level of glottis on T2 (Figure 2). After e-phonation, the lesion appeared to change its position and is seen extending along the lateral aspect of the aryepiglottic fold reaching upto the apex of pyriform sinus (Figure 2). On post contrast study, the lesion showed delayed but intense enhancement with few non enhancing areas (Figure 2). There was no diffusion restriction. MR angiography didn't show any connection with great vessels. The lesion measured approximately 2.98 x 2.3 x 4.8 cm. No neck node was identified.

Multiple biopsies were taken to look for a malignancy (lymphoma or epidermoid cancer), rare tumour (plasmacytoma, myxoma), amyloidosis, or an IMT. Specimens for bacteriological studies were collected. Results were inconclusive.

An elective tracheostomy was done preoperatively and under general anaesthesia, a larygofissure approach was performed to remove the mass which was found attached to the left aryepiglotic fold on its medial aspect and left ventricle with a broad base pedicle. The entire mass was removed in piece meal and sent for histopathological examination.

The histologic examination of the surgical specimens demonstrated multifocal infiltrations of lymphocytes, histiocytes, plasma cells, and eosinophils. Between the inflammatory cells, spindle cells with eosinophilic cytoplasm and oval to elongated nuclei were seen which was suggestive of inflammatory myofibroblastic tumour (Figure 3). Post-operative period went uneventful. She was discharged & kept under follow up (Figure 4).

I. Figures

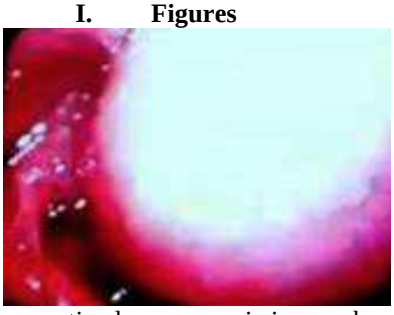

Figure 1. Preoperative laryngoscopic image showing Smooth globular mass in the left AEF region obstructing Laryngeal inlet

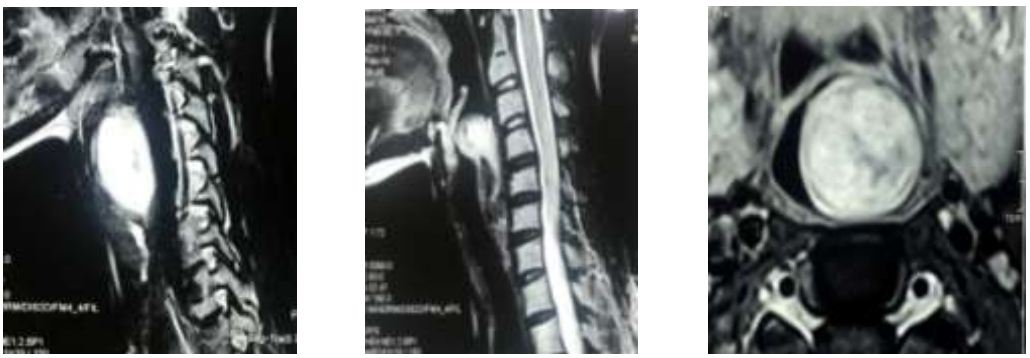

Figure 2: MRI of the lesion showing smooth hyperintense polypoidal lesion involving the left aryepiglottic fold plunging into the larynx causing significant narrowing of the same, with caudal extension upto the level of glottis which appeared to change its position on e-phonation and is seen extending along the lateral aspect of the aryepiglottic fold reaching upto the apex of pyriform sinus

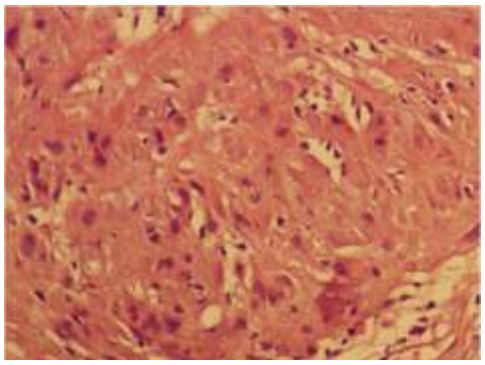

Figure 3: Histopathological staining of the tumour. Inflammatory myofibroblastic tumour consisting of foci of numerous plasma cells and lymphocyte which appear to be infiltrating between collagen fibers

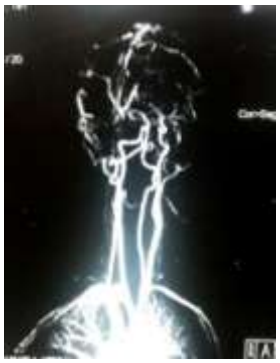

Figure4: MRA showing no major vascular connections

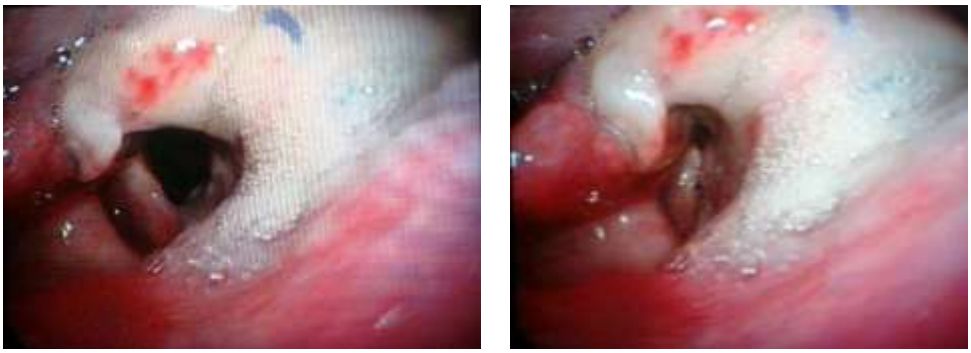

Figure 4: Post operative follow-up flexible laryngoscopic image at 3 months.

III. Discussion

Inflammatory myofibroblastic tumour is a neoplasm that usually involves the lungs and broncopulmonary tree & also rarely in other sites, like orbit, spleen, genitourinary tract, mesentery, cardio esophageal junction, breast, central nervous system and larynx [3]. Up to date, 31 cases of laryngeal IMT have been reported in literature (Table1) [3].

The origin and pathogenesis of this tumour is still unclear. Several theories have been proposed, including infectious, reactive (post-surgical or foreign body) and immunologic factors. The etiology is unknown; trauma may be the initiating factor but in majority of cases these lesions appear to arise spontaneously. [2]

Table 1. Reported laryngeal IMT cases in literature [3]

Supraglottic			
1.	Hanna et al	7/M	Right laryngeal ventricle.
2	Rodrigues et al	30m/M	Right aryepiglottic fold.
3.	Suh et al	52/F	Right false cord.
Glottic			
1	Wenig et al	19/M	Right anterior vocal cord, 1cm.
2	Wenig et al	65/M	Left true vocal cord and anterior commisure, 3cm.
3	Wenig et al	64/M	Right true vocal cord 0.7x0.5x0.3cm.
4	Wenig et al	26/F	Right true vocal cord 1x0.5cm.
5	Wenig et al	67/M	Right anterior true vocal cord, 0.4cm.
6	Wenig et al	22/F	Vocal cord.
7	Corsi et al	57/M	Anterior commisure, 0.8cm, recurrent mass 0.6cm.
8	Kendall et al	51/M	Right vocal cord, 0.4cm.
9	Matsumoto et al	53/M	Anterior lift vocal cord, 0.5cm.
10	Martinez et al	72/M	Left true vocal cord, 1.5cm.
11	Ereno et al	74/M	Left vocal cord, 3.5cm.
12	Guilemany et al	62/M	Right vocal cord.
13	Volker et a	32/F	Right vocal cord polyp, 0.8cm.
14	Belleza et al	23/M	Right vocal cord, 0.5cm.
15	Zitsch et al	57/F	Left true vocal cord.
16	Idrees et al	56/M	Anterior commisure.
17	Idrees et al	28/M	Right vocal cord.
18	Kumar et al	10/M	Right vocal cord.
19	Humaid et al	54/M	Left vocal cord.
Subglottic			
1	Wenig et al	54/F	Subglottic 1.5cm.
2	Munoz et al	5/M	Right lateral wall of subglottic, 1.8x0.5cm.
3	Alaani et al	49/F	Right subglottic
4	Zitsch et al	33/F	Left subglottic, 1.3x1.2cm
5	Zapatero et al	6/M	Cricotracheal , 1x1.5cm.
6	Keen et al	11/F	Subglottic
7	Humaid et al	38/M	Left lateral wall of subglottic.
Others			
1	Wenig et al	69/M	Pyriform sinus
2	Das-purkayastha et al	19m/M	Immediate upper trachea

IMT should be considered when histological findings from an inflammatory mass are inconclusive. IMTs can mimic malignancies and therefore require closely spaced follow-up evaluations. The diagnosis may be difficult. The time interval between symptom onset and treatment initiation is long as the disease behave indolently and manifestation depend upon site of occurrence. [2, 3]

Gross examination shows a fleshy, well-demarcated, whitish tumour that is firm to palpation [4, 5]. The classical histopathological feature includes spindle or satellite cells distributed in chronic inflammatory back ground [4, 5]. Differentiated myofibroblasts proliferate into a mass, which also contains a chronic lymphoplasmacytic inflammatory infiltrate [4, 5]. The mucosa show non-specific ulceration with granulation tissue. The submucosa show plump to ovoid spindle cells with focally collagen stroma. The findings do not suggest malignancy: neither necrosis nor atypical mitoses are seen, the mitotic index is low, and the plasma cell infiltrate is polyclonal. [2, 3]

Radiological imaging can help in diagnosis, can determine the extent of the lesion and its relationship with adjacent tissues and thus facilitates the prediction of surgical respectability. T1 weighted MRI images shows isointense lesions. T2 weighted images shows mixed isointense to hyperintense lesions. They usually shows mild to moderate contrast enhancement on CECT and MRI.

The histological diagnosis of IMT rests on immunohistochemical studies: antibodies to vimentin and actin bind strongly to myofibroblasts & stains for cytokeratins or protein S100 are usually negative [2, 3, 4].

Support for a neoplastic mechanism has come from studies showing chromosomal abnormalities responsible for abnormal ALK protein expression [3]

The main differential diagnoses are epidermoid carcinomas with spindle-shaped cells, malignant mesenchymatous tumours and lymphomas. True IMTs must be distinguished from inflammatory pseudo-tumours developing in response to post surgical healing processes, injury, or infection. Clinical course with multiple recurrences may suggest neoplastic process. [3]

The treatment rests on long-term high-dose corticosteroid therapy combined with conservative open or endoscopic surgery [2,3]. Local recurrences have been reported [2, 3], but the potential for malignant transformation remains debated. The overall prognosis is favourable. Complete surgical excision, eighter by endoscopic or open procedures, is the mainstay of treatment but may face obstacles related to the location of the tumour. The main risk is lies in balancing between excessive aggressive treatment of this benign growth and inadequate removal owing to its high recurrence rate. Failure of surgery, persistent growth of the mass despite corticosteroid therapy may require open partial or even total laryngectomy. [2, 3]

IV. Conclusion

The laryngeal IMT are rare localized lesions which behave indolently producing regional symptoms. It primarily affect adults with predilection to the glottis, & produce symptoms depending on their exact site of occurrence, size and age group affected. IMTs are amenable to conservative surgical resection. Key to successful management is the delicate balance between excessive aggressive treatment of this benign growth and inadequate removal.

Conflict of interest – none
Informed consent obtained
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Compliance with ethical standards.

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