CRICOID CHONDROSARCOMA PRESENTING AS BREATHY DYSPHONIA

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This is a case of chondrosarcoma of the cricoid cartilage in a 70 years old man who presented to us with breathy dysphonia of 3 years. Strobovideolaryngoscopy revealed the glottic insufficiency. A smooth subepithelial mass with prominent vessels superficially was noted just underneath the posterior glottis. On C.T scan, a well-demarcated lesion extending beyond the postcricoid area causing destruction of the posterior lamina of the cricoid cartilage was seen. The patient underwent temporary tracheostomy and excision of the mass was done through laryngofissure approach, preserving most of the inner perichondrium. The postoperative period was uneventful & the biopsy report revealed chondrosarcoma.

Keywords: Chondrosarcoma, Strobovideolaryngoscopy, Laryngofissure.

INTRODUCTION:

Chondrosarcoma of the larynx is a rare tumor of the upper respiratory tract that originates from cartilaginous tissue and represents the most common sarcoma of this site. The incidence of cartilaginous tumors of the larynx ranges from 0.02% to 0.07% of all head and neck tumors.[1,2,3] This tumor occur more frequently in the sixth and seventh decades of life and has a male predominance.[1,2,3] The etiology is unknown but the most common hypothesis is a primary disordered ossification of the cartilages.[4] Other theories presume a status of chronic inflammation or ischaemic changes in a preexisting chondroma [3,4,5] and the most frequent site is the cricoid cartilage followed by thyroid cartilage. Normally, chondrosarcoma has a low grade of malignancy and a slow growth rate.[5] The mainstay of treatment is complete surgical excision.

CASE REPORT:

A 70 year old retired serviceman presented to us with a 3 years history of breathy dysphonia which was slowly progressive in nature. On further questioning, his voice quality had steadily deteriorated ever since following an upper respiratory tract infection 3 years back. He does not give history of dysphagia, dyspnea or coughing while drinking water. He had been treated previously at different centers for GERD, but the treatment did not alleviate his symptoms.

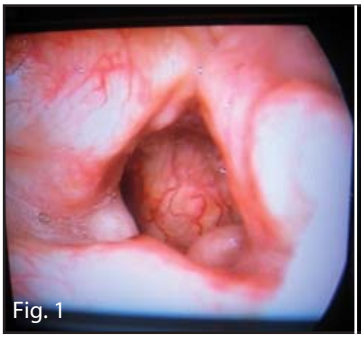


Fig. 1

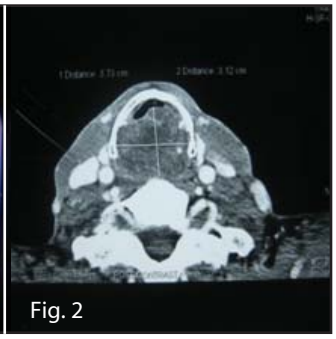
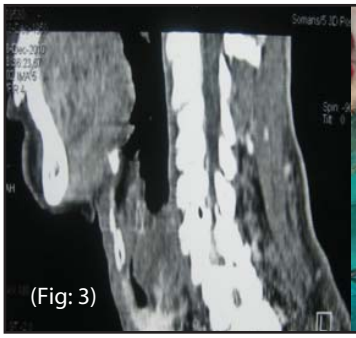
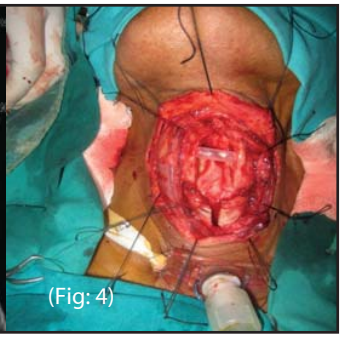


Fig. 2

Flexible nasostroboscopy & peroral strobovideolaryngoscopy (fig.1) revealed the glottic insufficiency with intact neuromuscular function on electoglottogram. On closer inspection, a smooth subepithelial mass with prominent vessels superficially was noted just underneath the posterior glottis with broad attachment preventing the vocal cords to adduct completely.


(Fig: 3)


(Fig: 4)

Contrast enhanced computed tomography (CECT) of the neck both axial & saggital cuts (fig. 2,3) demonstrated a well-demarcated lesion extending beyond the postcricoid area causing destruction of the posterior lamina of the cricoid cartilage measuring approximately 3.12 x 3.73 cm with some area of calcification. A clinical diagnosis of cricoid chondroma was made on the basis of the typical laryngoscopic and radiographic appearance. The size of the tumor was large enough to occlude the subglottic lumen & endotracheal intubation was not possible. Nevertheless, the patient underwent temporary tracheostomy and excision of the mass through laryngofissure approach (fig:4), preserving most of the inner perichondrium. He was decannulated 10 days following surgery & his postoperative period remained uneventful. The biopsy report confirmed the diagnosis of Chondrosarcoma. The patient is on regular follow up with a x-ray soft tissue neck (lateral view) & flexible laryngoscopy examination at each follow up. Till date he has no features of recurrence and his quality of voice has remained good.

Figure 5 shows 3 X-rays, 1st x-ray a preoperative one showing compromise of the airway at the level of subglottis while 2nd & 3rd x-rays are the postoperative x-rays showing a better air shadow suggesting a uncompromised airway.

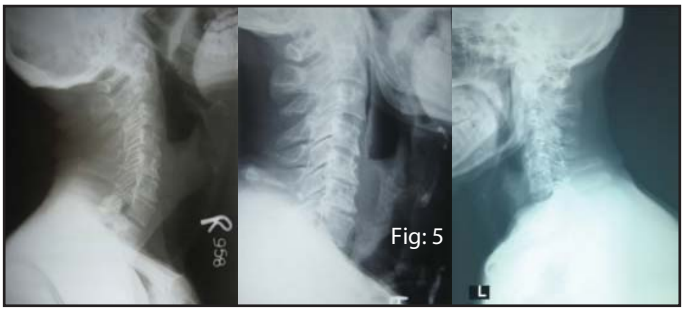


Fig: 5

DISCUSSION:

Laryngeal chondrosarcomas are rare, typically low-grade cartilaginous tumors that usually present with hoarseness and dyspnoea.[6,7] Most chondrosarcomas involve the posterior aspect of the cricoid cartilage.[6,7] Windfuhr suggested that mechanical stress might be an important causal factor since the most frequent sites (the posterior cricoid area and the infero-lateral wall of the thyroid) correspond with muscle insertion.[5] Their presence may lead to an expansion of the circumference of the laryngeal inlet, lateral displacement of the arytenoid cartilages, and consequent glottic insufficiency.[8] Computed tomography will usually show the presence of round lytic lesions with coarse calcification. The most typical CT image is of so called "popcorn" intratumoral calcification, found in some 80% of cases.[9,10] If a chondrosarcoma is diagnosed while it is still small, it may be amenable to conservative surgery; several different procedures have been recommended, depending on the location and extent of the tumor.[8,11].

The goal is excision with maximum preservation of laryngeal function. Surgery [4,11] is either on an external approach or by endoscopy. It must in all cases adhere to carcinological rules, ensuring complete excision. Grade 2 and 3 chondrosarcomas may be treated by partial surgery on condition that total excision with adequate safety margins is technically feasible. Chondrosarcoma is considered poorly sensitive to radiation therapy[13], which thus plays a very limited role. It may nevertheless be considered where surgery is contraindicated, if the lesion is judged not to be resectable or postoperatively in case of incomplete excision.[4,12] Most authors, however, agree that there is little evidence in favor of adjuvant postoperative radiation therapy following complete excision, even in grade 2 or 3 tumor.[4] Chemotherapy has no curative role in this indication.[13] Prognosis basically depends upon histologic grade and quality of exeresis. Overall five-year survivorship ranges from 79% to 90%, depending on the report.[11,12] Recurrence or metastasis is rare in case of complete surgery, occurring in 8% to 14% of cases.[8,11,14]

Chondrosarcoma classification by Evans et al.		
Grade 1	Well differentiated (low-grade)	Small, densely staining nuclei often with multiple nuclei within one lacune
Grade 2	Moderately differentiated (intermediate grade)	Increased cellularity, significant amount of cells having moderately sized nuclei but demonstrate a low mitotic rate of less than 2 mitoses per HPF (also includes myxoid chondrosarcoma)
Grade 3	Poorly differentiated (high grade)	More than 2 mitoses/HPF, nuclear size generally greater than seen in grade 2 (also includes dedifferentiated chondrosarcoma)

CONCLUSION:

Laryngeal chondrosarcoma is a rare tumor in which management is basically surgical. Prognosis is generally good, and basically dependent on histologic grade.

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