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CASE REPORTS

CRICOTRACHEAL CYLINDROMAS- RARE CASES AND REVIEW OF LITERATURE

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ABSTRACT

Adenoid cystic carcinoma of laryngotracheal areas are rare. They have no causal relation with smoking unlike the commoner squamous cell carcinoma of trachea and larynx. It is a low grade malignancy mainly affecting individuals of 4th-5th decade of life. They presents with breathlessness, cough and ocassionally hemoptysis and are misdiagnosed as asthma or reactive airway disease. The prolonged clinical course with late onset locoregional and or distant metastasis tends to be characteristic. Though they mandate surgical resection, adjacent critical structures hinder complete resection. Hence adjuvant radiation therapy is suggested. However, radiation therapy when offered as primary treatment for advanced disease, also show fairly comparable survival benefits, as in the two cases reported.

INTRODUCTION

Tracheal tumors are rare. It accounts for 0.2% of respiratory malignancies, with squamous cell carcinoma being commonest .Adenoid cystic carcinoma contribute to 10% of them (Gaissert, 2006). ACC, also known as Cylindroma, was first described by Bilroth in 1859 (Billroth, 1859) Tracheal ACC has neither sex predeliction nor is it associated with smoking. It is a low grade malignancy mainly affecting tissues of salivary gland origin, mostly in 4th-5th decade of life (Chappidi *et al*., 2004). Tracheal ACC presents with breathlessness, cough and occasionally hemoptysis. They get treated as asthma or reactive airway disease (Yang *et al*., 2005). It has a prolonged clinical course with late onset locoregional and or distant metastasis The treatment is often surgical resection. But adjacent critical structures hinder complete resection where, adjuvant treatment with radiation has been suggested. However, situations where radiation is offered as primary treatment in view of resection difficulty, due to proximity of critical structures, also show fairly comparable survival benefits (Kaminski *et al*., 2003; Bittner *et al*., 2008) whether due to radiosensitivity, or slow course of disease progression or low reported case numbers, still requires clarification, as in the two cases reported.

Case reports

Case -1

24 year old male with no comorbidities or habit of smoking, was evaluated for breathlessness of 3 months duration .He was diagnosed with posterior tracheal wall tumour on bronchoscopy.

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In view of critical airway, he underwent tracheostomy. CT neck revealed lesion measuring (Table/Fig 1) 30 X 20 mm involving the posterior wall of the trachea extending from the lower border of cricoid cartilage to C7 level inferiorly with cricoid cartilage infiltration.No nodal or pulmonary metastasis was noted. Biopsy from the same was reported as adenoid cystic carcinom.He underwent total laryngectomy with resection of upper four tracheal rings and thyroid isthumectomy in January 2014. Final histology (Table/Fig 2) confirmed the diagnosis of adenoid cystic carcinoma with cribriform pattern and was reported to have adequate margins. He received adjuvant post op radiation therapy and was on followup since then. 6 months later, he underwent Stomaplasty in view of stomal stenosis and secondary Tracheoesophageal Prosthesis insertion. He is on regular follow up till date with no evidence of locoregional recurrence or distant metastasis (DFI >2.5years), except for two more instances of stomal stenosis with keloid formation (Table/Fig 3) and revision stomaplasty (Table/Fig 4), recently done in May 2016 followed by weekly intralesional triamcinolone injection for a month.

Case -2

A 58 year old hypertensive gentleman, who underwent tracheostomy in October 2012 for stridor was diagnosed to have upper tracheal growth on CT evaluation (Table/Fig5). It revealed tracheal lesion infiltrating esophagus that measured about 5cm craniocaudally from C6 vertebral level to C7 vertebral level. Biopsy from the same was reported as adenoid cystic carcinoma. In view of esophageal infiltration, though surgical treatment was suggested, patient was not willing for extensive resection and opted for alternative line of treatment. He received Radical RT (66Gy / 33F) that was completed in Jan 2013.

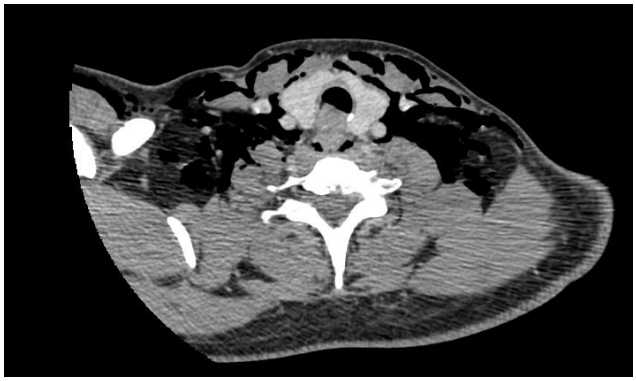

Figure 1. 30 X 20 mm subglottic lesion involving the posterior wall of the trachea extending from the lower border of cricoid cartilage to C7 level inferiorly with cricoid cartilage infiltration

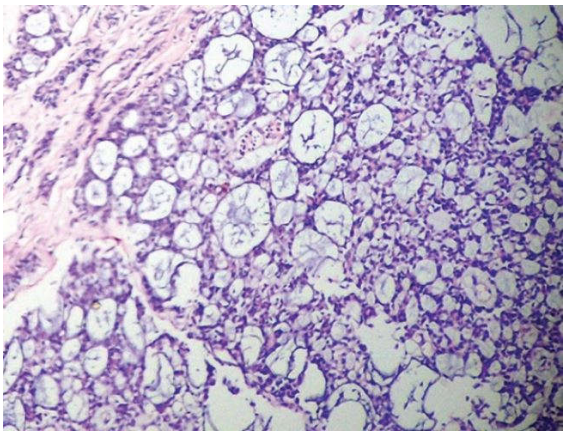

Fig. 2. Histopathology showing adenoid cystic carcinoma with cribriform pattern

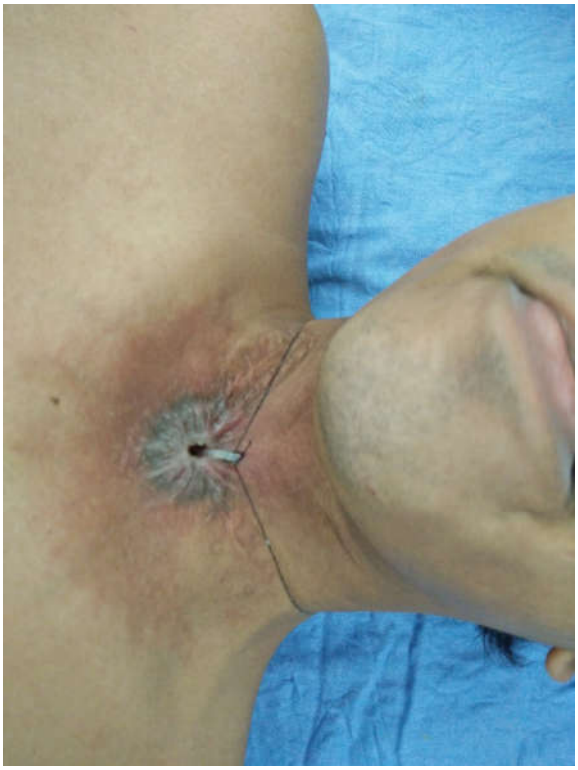

Fig. 3. Stomal stenosis with keloid formation

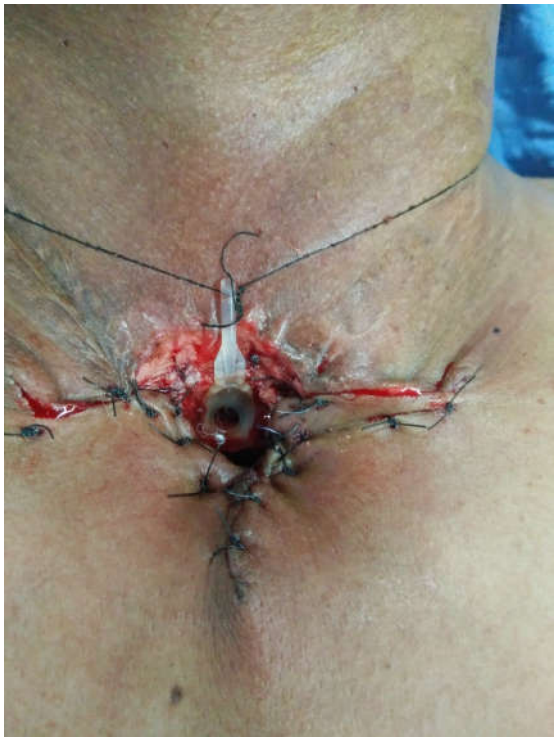

Fig. 4. After revision stomaplasty

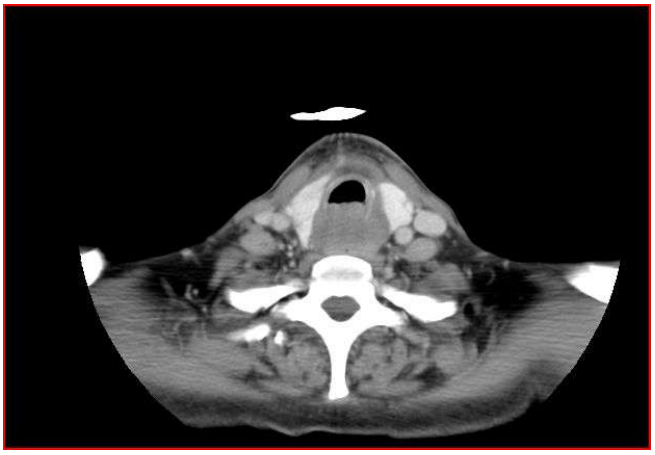

Fig. 5. A tracheal lesion infiltrating esophagus that measured about 5cm craniocaudally from C6 vertebral level to C7 vertebral level

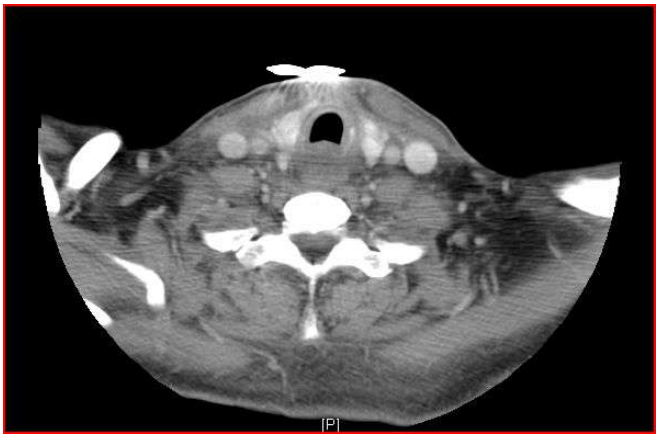

Fig 6- CT after 2 months of radical radiotherapy revealing complete remission of the lesion radiologically

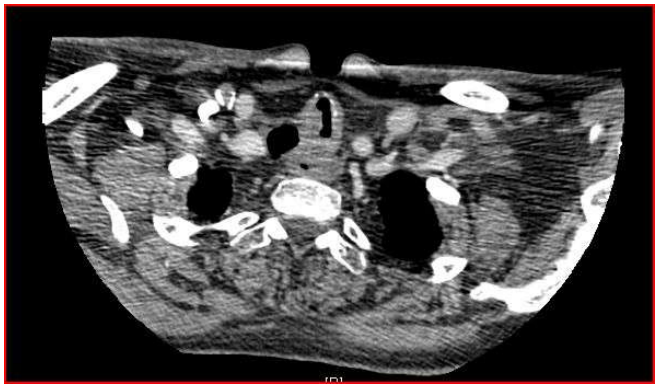

Fig. 7. An irregular heterogeneously enhancing soft tissue thickening in the posterior wall of the trachea from just below the lower border of cricoid cartilage to a distance of approximately 3 cm and causing gross narrowing of the tracheal lumen with esophagus and thyroid infiltration

First follow up CT (Table/Fig 6) after 2 months revealed no lesion in subglottis and was able to tolerate orally normal diet. One month later after confirming adequacy of airway with flexible laryngoscopy, he was decannulated and had been on close follow up with DFI of 3 years until recently when he developed noisy breathing. CT on May 2016 (Table/Fig-7) revealed an irregular heterogeneously enhancing soft tissue thickening in the posterior wall of the trachea from just below the lower border of cricoid cartilage to a distance of approximately 3 cm and causing gross narrowing of the tracheal lumen with esophagus and thyroid infiltration. As the patient is still not keen on surgery he being planned for chemotherapy with implied consent of guarded response.

DISCUSSION

Adenoid cystic carcinomas (ACC) are common in salivary glands. However its occurrence in cricotracheal area is rare and accounts for about only 10% of all tracheal tumours. ACC, also known as cylidroma, was first described by Bilroth. It is a low grade malignancy affecting patients of 4th-5th decade. No causal relation with smoking has been established. They present with noisy breathing, dyspnea, cough or hemoptysis, and often get treated as for reactive airway diseases. They are non encapsulated tumours that spread directly, submucosally, perineurally or hematogenously with lymphatic spread being uncommon. Lung being most common followed by brain, bone and liver metastasis, have been reported. CT identifies the extratracheal extension aspect and MRI reveals submucosal infiltration and extent of mediastinal invasion to assess resectability.

Though ideal treatment is surgical resection ,it is not feasible at times due to the proximity of critical structures, warranting adjuvant radiation or chemoradiation as the primary modality.The constrain of not able to resect >6cm trachea poses high chance of close resection margins, making them prone to recur. Infiltration of adjacent critical structures, makes them unresectable leading to 5-and 10 year survival rates of 33% and 10% respectively against 52% and 30% when resectable (Gaissert *et al*., 2004). Adjuvant radiation treatment claims a 5 and 10 year survival of upto 80 and 57% respectively (Maziak *et al*., 1996; Regnard *et al*., 1996; Perelman *et al*., 1996) though its role is still uncertain (Regnard *et al*., 1996).

The overall survival of patients in margin clear resections vary (Maziak *et al*., 1996; Honings *et al*., 2010). Though 13-26% nodal positivity is reported at resection (Maziak *et al*., 1996; Honings *et al*., 2010), there is no survival benefit noted with node negativity (Gaissert *et al*., 2004). Moreover, late locoregional failure and or distant metastasis probably warrants more focus on the impact of perineural invasion feature of ACC.

Conclusion

Cricotracheal adenoid cystic carcinomas are rare tumours. Surgical resection remains the main stay of treatment with adjuvant RT as in our case-1, that provides the best survival rates. However, radiation or chemoradiation is offered for advanced and unresectable tumours. These cases are presented due to their rarity after analyzing the available literature on this uncommon site of a known malignancy.

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