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LETTER TO THE EDITOR

Tracheolaryngeal Adenoid Cystic Carcinoma

The tumour that almost took her breath away

كيس ليمفاوي سرطاني في القصبة الهوائية والحنجرة

الورم الذي أوشك علي خطف أنفاسها

Dear Sir,

A 40-year-old female patient presented to the Southern Command Hospital, Pune, India, in August 2015 with a six-month history of breathlessness. She was experiencing three to four episodes of dyspnoea per week with each episode lasting a few minutes. The breathlessness was aggravated on exertion and relieved with rest. She had no known comorbidities and had previously been treated symptomatically for asthma with no significant improvement. An indirect laryngoscopy revealed a proliferative growth inferior to the postcricoid area which was partially filling the trachea without affecting the glottic closure [Figure 1A]. Contrast-enhanced computed tomography of the neck revealed a well-defined soft tissue lesion (2.5 x 2.2 x 2.2 cm) in the infraglottic cavity and upper trachea with no significant enhancement [Figure 1B].

An emergency tracheostomy was performed and a biopsy revealed tumour cells arranged in cribriform structures, tubules and clusters amidst a fibrous *stroma* [Figure 1C]; the cells had minimal eosinophilic cytoplasm, indistinct cell borders, oval hyperchromatic nuclei and inconspicuous nucleoli. On subsequent elective excision, all of the surgical margins were positive with the tumour abutting the tracheal cartilage [Figure 1D] and involving the inferior and posterior margins of the left lobe of the thyroid. The tumour showed similar histomorphology with *foci* of perineural invasion. A diagnosis of an adenoid cystic carcinoma (ADCC) was made at stage T4N0M0, indicating no regional lymph node or distant metastasis. As all of the surgical margins and the thyroid gland were seen to be involved during a frozen section procedure, the surgeons opted for palliative surgery with minimal local resection of the cricoid cartilage and first tracheal ring in addition to a total thyroidectomy. The defect was closed by end-to-end *anastomosis* with no attempt made at reconstruction. The patient subsequently received palliative radiotherapy (70 Gy) for four weeks. Due to the possibility of mucosal erosion and tracheal restenosis, the tracheostomy was continued for 12 weeks and the incision was closed at the end of the 14th week. The patient was stable with no clinical evidence of recurrence at a four-month follow-up.

ADCCs are malignant neoplasms of the laryngeal salivary glands characterised by a protracted and aggressive clinical course.[1] These carcinomas form less than 1% of all tracheolaryngeal malignancies and are known to reach large sizes; they usually occur among patients in their fifth and sixth decades of life and no gender predilection has been observed.[2] As with the current case, ADCC can present with symptoms of obstruction, such as breathlessness.[3]

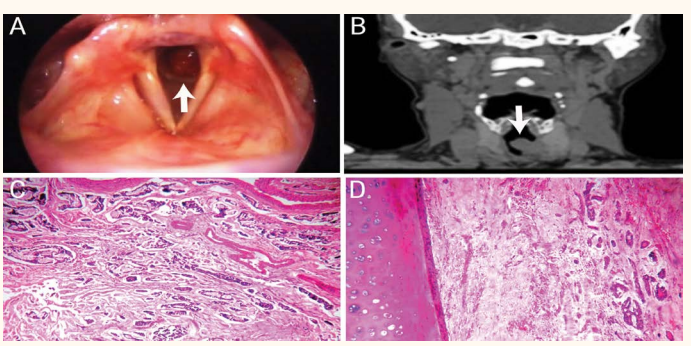

Figure 1A–D: A: Fibre-optic bronchoscopy image of a 40-year-old female with a tracheolaryngeal adenoid cystic carcinoma showing a proliferative growth (arrow) inferior to the postcricoid area, partially filling the trachea without hampering the glottic closure. **B:** Contrast-enhanced computed tomography of the neck revealing a well-defined soft tissue lesion (arrow) in the infraglottic area and upper trachea with no significant enhancement. **C:** Haematoxylin and eosin stain at x400 magnification showing tumour cells arranged in cribriform structures, tubules and clusters amidst a fibrous *stroma*. **D:** Haematoxylin and eosin stain at x400 magnification showing the tumour abutting the tracheal cartilage.

The differential diagnosis for ADCC includes polymorphous low-grade adenocarcinomas, basal cell adenomas, mixed tumours and basaloid squamous cell carcinomas.[4] A cribriform pattern of tumour cells is typical of ADCC; however, this pattern is rarely seen in basal cell adenomas, basaloid squamous cell carcinomas and mixed tumours. While distinction between polymorphous low-grade adenocarcinomas and ADCCs can be challenging, the Ki-67 index is 10 times higher for the latter.[4] In addition, smooth

muscle markers of myoepithelial differentiation are positive in cases of ADCC but negative for polymorphous low-grade adenocarcinomas. Pseudocysts are usually positive with periodic acid-Schiff and Alcian blue stains and contain basement membrane components.[4] The epithelial cells are usually positive for carcinoembryonic and epithelial membrane antigens while duct-lining cells are positive for cluster of differentiation 117 and tumour protein 63. Alterations in chromosomes 6q, 9p and 17p12–13 are the most frequent cytogenetic alterations reported.[4]

Although the histologic appearance of ADCC is well-described with certain features that may predict prognosis, surgical removal is often difficult given the infiltrative nature of these neoplasms with their propensity to invade into the adjacent tissues and extend along nerve segments.[5] This is further compounded by the deceptively circumscribed macroscopic appearance of the tumours.[3] These factors are challenging for surgeons attempting to remove the tumour with negative margins. Due to inadequate surgical excision, recurrence rates are high, ranging from 75–90%.[6] Radical surgery has not been shown to improve survival rates or decrease local recurrence when compared with a conservative surgical approach and postoperative radiation.[3] Optimal treatment therefore remains wide local excision with or without postoperative radiation.

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