Metastases from tracheal adenoid cystic carcinoma

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A 32-year-old female never smoker presented with dyspnoea and stridor. CT thorax showed a polypoidal mass obstructing the trachea. Bronchoscopy revealed a circumferential nodular tumour 4 cm from the vocal cords, removed using electrosurgical snare and thereby re-establishing trachea patency (figure 1A,B). Histology showed adenoid cystic carcinoma (ACC). She underwent tracheal resection; as the resection margins showed ACC with perineural and fascia invasion, she received adjuvant radiation therapy 72 Gy. Yearly CT chest was performed for surveillance and CT 8 years after surgery showed enlarged subcarinal lymphadenopathy, bilateral pulmonary nodules and a left thyroid nodule (figure 1C,D). Thyroid ultrasound fine-needle aspiration confirmed 1 cm hypoechoic nodule due to ACC. Endobronchial ultrasound-guided transbronchial needle aspiration of subcarinal lymph node revealed metastatic ACC (figure 2), but no recurrence in the trachea.

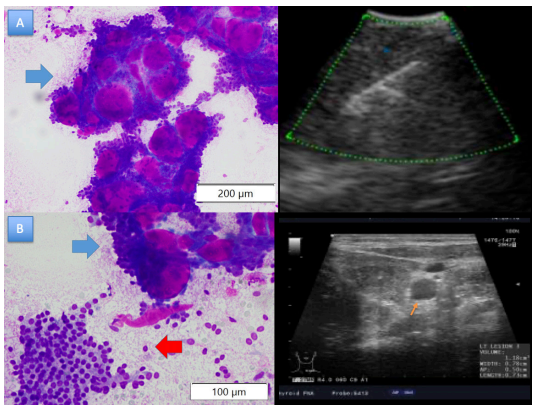

Figure 2 (A) EBUS-guided FNA of subcarinal lymph node (measuring 1.5 cm in widest dimension) showing tumour containing purple-staining globules characteristic of cribriform adenoid cystic carcinoma (blue arrow) (Hemacolor stain, original magnification x100). (B) FNA of thyroid nodule (measuring 1.0 cm in widest dimension) showing adenoid cystic carcinoma similar to figure 2A (blue arrow). Benign thyroid follicular cells are also seen, lower left (red arrow). (Hemacolor, original magnification x200). EBUS, endobronchial ultrasound; FNA, fine needle aspiration.

DISCUSSION

Primary tracheal tumours are rare, and 10%–15% are due to ACC, which comprises of tubular, cribiform and solid subtypes. All variants have the propensity for longitudinal, submucosal and perineural invasion.[1] Lymphatic and haematogenous metastases are even rarer at 18% and 7%, respectively.[1 2] Local recurrence and/or metastasis can develop up to 94 months after diagnosis,[2] thereby highlighting the indolent nature of ACC, and the need for prolonged surveillance. Our patient developed local invasion of thyroid, lymphatic and haematogenous metastases 8 years later. Surgery is first-line therapy for tracheal ACC, and promising results have been demonstrated using autologous composite tissue replacement when end-to-end anastomosis is impossible.[3]

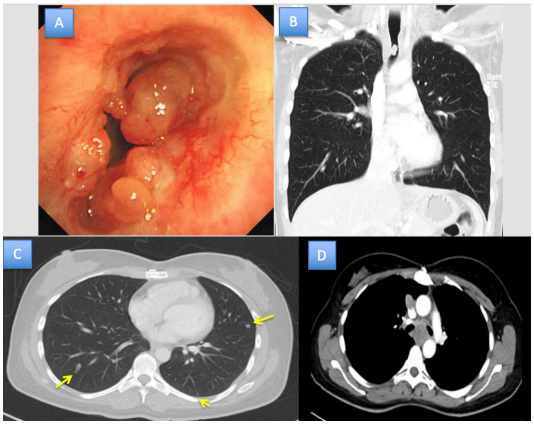

Figure 1 (A) Malignant polypoidal tracheal tumour with 80% obstruction (B) CT coronal view showed polypoidal lesion in mid-trachea (C,D): CT surveillance showed enlarged subcarinal lymph node and multiple pulmonary nodules seen (yellow arrows).

Radiotherapy is recommended in patients who have positive resection margins, perineural and lymphatic invasion, or are inoperable as ACC is radiosensitive.[1 2] Good local control is observed at radical doses in excess of 60 Gy. Radiotherapy with neutrons, photons or photon/neutron mixed beam can aid in dose escalation for disease control while minimising toxicity.[2]

ACC shows slow growth kinetics and poor response to single agent or combination chemotherapy.[4] Tumour expression of c-KIT and other growth factor receptors have led to clinical evaluation with imatinib (c-KIT inhibitor) and tyrosine kinases against epidermal growth factor receptor (EGFR), which have also yielded dismal results.[4]

Translocation between chromosomes 6q and 9p that juxtaposes the genes for MYB and nuclear factor 1/B (NF1B) transcription factors is specific to and found in 86% of ACC. MYB:NF1B gene fusion leads to over-expression of MYB oncoprotein, and deregulated MYB is central to tumour pathogenesis. Downstream effectors of MYB may herald future drug development against these novel targets.[4]

Five and 10-year survival rates of ACC are over 70% and 50%, respectively.[2] Since good local disease control is achieved through advanced surgical techniques and adjuvant radiotherapy in excess of 60 Gy, distant metastasis from hematogenous and lymphatic spread become more apparent

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consequent to longer survival. Surgery for solitary pulmonary metastasis should be considered if the local disease is under control. Bronchoscopy plays an important role not only for tissue diagnosis but also in re-establishing the airway necessary for surgery with, electrosurgical snare, argon plasma coagulation, laser or cryotherapy. In addition, bronchoscopy offers airway surveillance and palliation.[5] Our patient was offered radiotherapy and chemotherapy, but elected for expectant management of metastastic ACC and died 12 years after ACC was first diagnosed.

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