A rare case report of Extraskeletal Ewing’s sarcoma of the trachea

Clinical History:

A 14-year-old female presented with complaints of breathlessness upon exertion for 6 months, and aggravated since 1 week. Chest x-ray taken appeared unremarkable. Contrast enhanced CT was performed for further Evaluation.

Imaging Findings:

Chest X-ray PA view-trachea appeared central. No significant abnormality was noted.

CECT thorax showed a well defined enhancing polypoidal soft tissue density lesion in the left anterolateral wall of the trachea at the T2 vertebral Level, with significant luminal narrowing and mediastinal extension.

Bronchoscopy showed a polypoidal lesion in the lateral wall of the trachea projecting into the tracheal lumen with luminal narrowing.

Microscopy showed diffuse proliferation of round cells within a hyalinized stroma, which had high-grade nuclei, occasional nucleoli, and minimal cytoplasm.

Discussion:

Primary endotracheal tumours are uncommon in childhood. The most common malignant endotracheal lesions are carcinoid tumour and mucoepidermoid carcinoma. Inflammatory pseudotumour and endotracheal hamartoma are common benign pathologies. Most of the malignant lesions are found in adolescents while benign lesions like hemangiomas are commonly seen in infants and children [1].

These tumours most frequently present as wheezing, stridor and recurrent pneumonia in childhood. Imaging should be considered to rule out an obstructive process when there is no response to standard treatment with antibiotics and bronchodilators. Chest radiograph is usually normal. Contrast enhanced CT is the imaging modality of choice [1].

Extraskeletal Ewing’s sarcoma (EES) of the trachea is extremely rare with only 2 published cases. EES belongs to the Ewing’s sarcoma family of tumours which also include osseous Ewing’s sarcoma, Askin’s tumour and PNET.

They are neuroectodermal in origin and share the cytogenetic marker, translocation of chromosomes t (11;22)(q24;q12). The most common locations of EES are paravertebral region, lower extremities, chest wall, retroperitoneum, pelvis and hip [2].

The criteria proposed for the diagnosis of EES are absence of osseous involvement at MRI, no increased uptake in
bone or periosteum on scintigraphy, and small blue round cells on histology [2]. Immunohistochemical and histochemical staining positive for glycogen (PAS), NSE, S-100 and MIC-2 marker [3].

The imaging features of EES on CT as an enhancing soft tissue mass of muscle attenuation are nonspecific. Necrosis and haemorrhage may correspond to low attenuation areas. On MRI EES is heterogeneous, iso- to hypersignal on T2. Features suggestive of haemorrhage, fluid levels, serpentine flow voids and a pseudocapsule may be seen [2]. Tracheal EES manifests as an enhancing endoluminal mass with significant stenosis and mediastinal infiltration. Proper airway intervention at the time of diagnosis is important prior to definitive treatment [4].

The role of imaging is for staging, which includes a CT of the chest, Tc99 bone scan and FDG PET scan for evidence of metastatic disease. At FDG PET, this tumour can demonstrate avid uptake depending on metabolic activity [4]. Often the diagnosis of primary tracheal EES is made after bronchoscopy and biopsy.

The treatment of EES involves combined systemic chemotherapy and local therapy by surgical resection, radiation, or both.

In conclusion, endotracheal tumours in the adolescent population is more likely to be malignant. Although the imaging appearance of primary EES of the trachea is nonspecific it must be considered as a possibility in case of a rapidly growing endotracheal soft tissue mass.

**Differential Diagnosis List:** Primary Extraskeletal Ewing's sarcoma of the trachea, Mucoepidermoid carcinoma, carcinoid, inflammatory pseudotumour

**Final Diagnosis:** Primary Extraskeletal Ewing's sarcoma of the trachea

**References:**

Brianne Barnett Roby, MD; Dennis Drehner, MD; James D. Sidman, MD. (2011) Pediatric Tracheal and Endobronchial Tumors An Institutional Experience. Arch Otolaryngol Head Neck Surg137(9):925-92
Description: Plain CT Thorax shows a well defined polypoidal soft tissue density lesion in the left anterolateral wall of the trachea at T2 vertebral level with significant luminal narrowing and mediastinal extension. Origin: Department of Radiodiagnosis, Government medical college, Calicut, Kerala

Description: The lesion shows significant post contrast enhancement. Origin: Department of Radiodiagnosis, Government medical college, Calicut, Kerala
Description: CT Thorax lung window shows no abnormalities in lung parenchyma. Origin: Department of Radiodiagnosis, Government medical college, Calicut, Kerala
**Figure 2**

*Description:* Diffuse proliferation of round cells within a hyalinized stroma, which has high-grade nuclei, occasional nucleoli, and minimal cytoplasm.

*Origin:* Department of pathology, Government medical college, Calicut, Kerala
Description: Chest xray showed no significant abnormality
Origin: Department of radiodiagnosis, government medical college, Calicut