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Breath Of Challenge - Pulmonary Synovial Sarcoma

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Primary pulmonary synovial sarcoma is a rare malignant tumour of mesenchymal origin. It accounts for less than 0.5% of all lung neoplasms. It is highly aggressive and arises most commonly from the lung parenchyma. Histologically the monophasic subtype is most common but is frequently mistaken as biphasic due to the resemblance between entrapped pneumocytes and the epithelial component of biphasic tumours. The gold standard for diagnosis remains the presence of chromosomal translocation, which produces the SS18-SSX fusion gene and is characteristic of more than 90% of synovial sarcomas (SS). The mainstay of treatment involves complete surgical resection along with adjuvant chemo-radiotherapy. The present case report describes a 64-year-old who presented with persistent upper back pain and progressive breathing difficulties for one year. PET-CT revealed a large heterogeneous soft-tissue mass in the left upper and middle lobes of the lung with areas of necrosis (9.1 × 13.6 × 13.5 cm). Biopsy demonstrated a spindle cell neoplasm. Immunohistochemistry was positive for cytokeratin 7 and 19, Bcl-2, EMA, and SMA, while negative for CD-34, Desmin, and vascular tumour markers. Molecular analysis showed EGFR and TP53 variants were positive, whereas ALK rearrangements were negative. In advanced disease, pazopanib has demonstrated improved median progression-free survival, offering an alternative targeted therapy. Future directions involve SS18-SSX-derived peptide vaccines, therapies against vascular endothelial growth factor, and epidermal growth factor receptor blockade. The present case will help clinicians consider pulmonary synovial sarcoma as the differential diagnosis of patients with a sizable mass in the mediastinum.

Keywords: Pulmonary synovial sarcoma, Spindle cell neoplasm, Immunohistochemistry, Pazopanib

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