A case of mediastinal lymphadenopathy, with plot twists.

A Mediastinal Mystery

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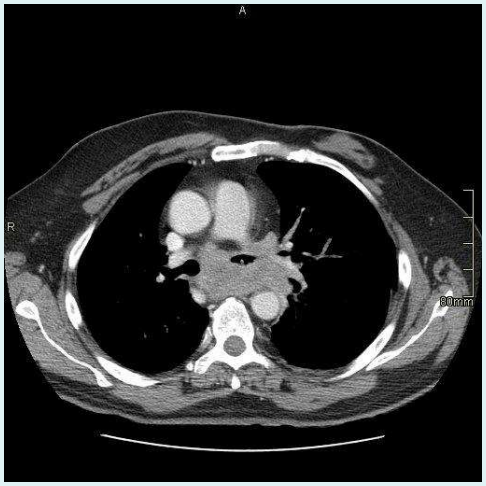
A 55 year old man underwent computed tomography (CT) of the chest and abdomen to investigate new haemoptysis. He had a past medical history of what was presumed to be squamous cell carcinoma of the larynx six years previously, undergoing chemoradiotherapy and laryngectomy.

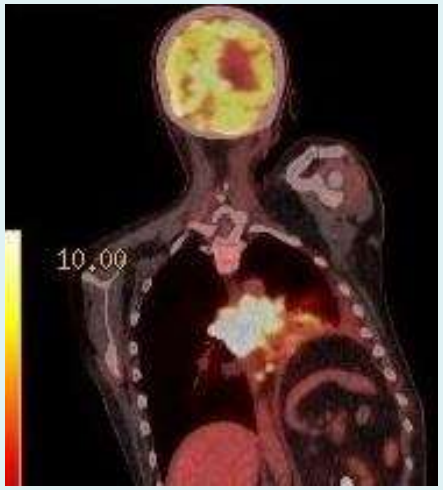


Axial arterial phase contrast-enhanced CT thorax

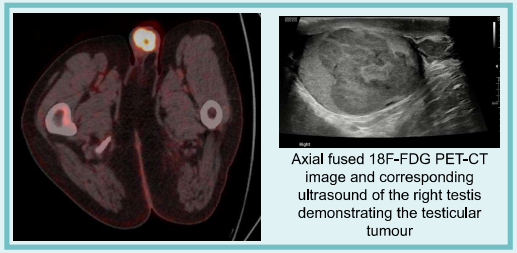
The CT showed extensive enlarged left hilar, aortic and mediastinal nodes, with infiltration into the left main bronchus, thought to most likely represent **metastatic lung cancer.**

The patient underwent an endobronchial ultrasound (EBUS) examination and biopsy, with histopathological testing and initial immunohistochemistry suggesting a low-grade neuroendocrine neoplasm.

18 F-fluorodeoxyglucose (FDG) positron-emitting tomography (PET)-CT was performed. This showed that the mediastinal lymphadenopathy was intensely avid, with interval progression, as well as an involved right supraclavicular node. No definite primary lung lesion was identified, although there was new collapse and consolidation in the left lower lobe and lingula, with a left pleural effusion.



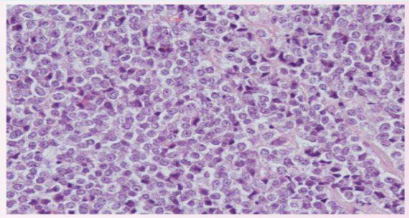
Coronal fused 18F-FDG PET-CT image of the thorax



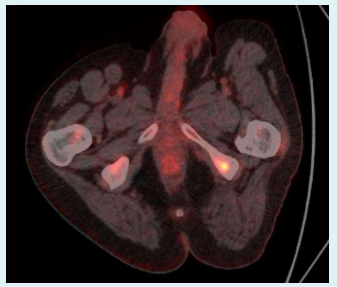
Axial fused 18F-FDG PET-CT image and corresponding ultrasound of the right testis demonstrating the testicular tumour

Intense FDG uptake with central photopenia was noted in the right testis and subsequent ultrasound demonstrated a right testicular tumour. **Metastatic testicular cancer** was considered as a potential unifying diagnosis, but testing for the relevant tumour markers proved to be negative.

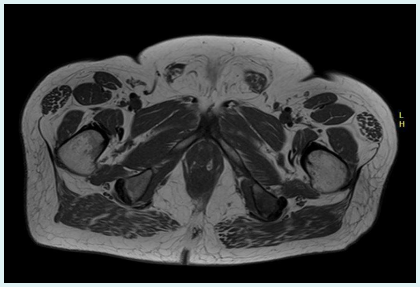
Focal increased FDG uptake was identified in the left ischium, and magnetic resonance (MR) imaging confirmed appearances suspicious for metastasis, with further bone lesions elsewhere. The testicular tumour was subsequently also thought to represent a metastasis.



Representative hematoxylin and eosin stain of Ewing sarcoma showing poorly differentiated tissue consisting of small, round, blue cells with prominent nuclei and minimal cytoplasm[1]



Axial fused 18F-FDG PET-CT image and axial T1-weighted MR sequence demonstrating the left ischial metastasis



Assessment of the initial samples obtained by EBUS continued, with cytogenetic testing surprisingly subsequently revealing **Ewing sarcoma**. This was discussed at the regional sarcoma multidisciplinary team meeting where the previous histopathology samples of the laryngeal malignancy were reviewed and, in hindsight, were also suspicious for Ewing sarcoma.

Sadly the patient had been steadily deteriorating and died before any treatment could be started.

Ewing sarcoma is a rare aggressive small round blue cell tumour, derived from the neuroectoderm, which predominantly arises in bone but can also originate in soft tissue[1]. It mainly affects the young and is rare in those aged over 30 years.

Laryngeal Ewing sarcoma is even rarer, accounting for 2-3% of extraosseous cases with only case reports described in the literature[2].

Primary Ewing sarcoma of the genitourinary tract has been described, including of the testis, but testicular metastases are exceptionally rare with only a single case reported as of 2024[3].

The 5-year survival for metastatic Ewing sarcoma at presentation is 30%[1].

1. Riggi N, Suvà ML, Stamenkovic I. Ewing's Sarcoma. N Engl J Med. 2021 Jan 14;384(2):154-164., 2. Poojari M, Madabhavi I, Desai S. Extraskeletal Ewing's sarcoma of supraglottis: A rare case report. Oral Oncol. 2024 May;152:106789., 3. Aboulaich, Y., Kharbach, Y. & Khallouk, A. Unusual testicular metastatic localization of Ewing's sarcoma: case report. Afr J Urol 30, 38 (2024).