Case Report
An Incidentaloma: Primitive Neuroectodermal Tumor of the Thymus

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A 57-year-old man presented for routine EGD as follow-up for his Barrett’s esophagus, and upon complaining of palpitations, was referred to the Emergency Department. An EKG revealed new-onset atrial fibrillation. On subsequent workup, a nuclear stress test was performed, but the study was deemed limited given the marked amount of radioactive uptake in the upper thorax. Suspicious for a mass lesion, a CT of the chest was performed. A heterogeneously enhancing anterior mediastinal mass was found, with differential diagnosis including thymic carcinoma, germ cell tumor, or lipoma. An ultrasound-guided biopsy was performed, which revealed a trabecular neoplasm highly suggestive of neuroendocrine tumor. PET tumor imaging showed the anterior mediastinal mass to have intense FDG activity, which in correlation with the pathology report, favored thymic carcinoma. There was no evidence of lymph node or distant metastases. The surgical service was consulted for tumor debulking, and a sternotomy with mediastinal exploration was planned. The tumor was very vascular. It extended into the right pleural space, and even deep into the left pleural cavity, going up to the thoracic inlet. The posteroinferior aspect of the tumor was overlying the great vessels. The tumor was infiltrating the innominate vein and left phrenic nerve. Due to tumor extent and infiltration into surrounding structures, extensive surgical resection or thymectomy was abandoned. Surgical pathology showed a malignant neuroendocrine tumor histologically and immunophenotypically consistent with a peripheral primitive neuroectodermal tumor.

Pathologically, the ESFTs are classically described as small round blue cell tumors of neural crest derivation with differentiation along a neuroendocrine lineage [2].

Pathology reports for our patient’s tumor revealed the following immunochemistry: positive for CD99, cytokeratin, CD57, and NSE; negative for TTF-1, CK7/CK20, CD56, syn-
aptophysin, chromogranin, CD117, PLAP, S100, CD30, AFP, and CD1a. Histology revealed a poorly differentiated trabecular neoplasm highly suggestive of a peripheral neuroectodermal tumor (Figure 1).

Radiographically, PNET imaging is nonspecific, with use (of CT or MRI) aiding primarily in defining tumor scope, treatment strategy, and the evaluation of therapeutic outcomes [3]. The appearance of tumor heterogeneity on imaging represents areas of hemorrhage and necrosis, as was the case in our patient's large lesion (Figure 2) [4].

Initial presentation is the only predictor for long-term survival, with primary tumor-only presentation having a 5-year survival rate of 60%, compared to 33% for primary plus metastatic disease [2, 5]. Overall survival rates in adults are in line with pediatric study outcomes [6].

Current treatment recommendations advocate complete surgical resection whenever possible, radiation therapy, and adjuvant versus neoadjuvant chemotherapy. In the pediatric population, treatment for PNET typically lasts 6–9 months and consists of alternating courses of 2 chemotherapeutic regimens: (1) vincristine, doxorubicin, and cyclophosphamide and (2) ifosfamide and etoposide. Treatment is similar for adults [7].

Our patient was scheduled for concurrent chemoradia
tion with curative intent. Outcome was optimistic, and an uncomplicated treatment course was carried out.

Conflict of Interests
All authors confirm that there is no conflict of interests to declare.

Authorship
All authors had full access to the data and a role in writing the paper.

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