A 53-year-old Caucasian male was evaluated for cough and exertional dyspnea. His past medical history was significant for heavy cigarette smoking and recurrent pneumonias. 

CT scan of the chest performed in January 2007 revealed multiple, sub-centimeter, calcified and non-calcified pulmonary nodules those were stable when compared to a scan done in May of 2005. However, a new 6 mm non-calcified nodule was noted in the right lower lobe (Figure 1(A)). In June 2007, a repeat CT scan of the chest demonstrated enlargement of the nodule to a diameter of 2.5 cm (Figure 1(B)). Three weeks later, PET-CT showed high uptake within the mass (standardized uptake value maximum of 15.6) with a new subjacent nodule measuring 1.2×2.0 cm and no evidence of distal metastases (Figures 1(C) and (D)).

Because of the patient’s good performance status, the size of the tumor, and the lack of distant metastases, it was decided to proceed with surgical resection with curative intent.

During surgery, the mass was located in the superior segment of the right lower lobe. Initial pathologic examination on frozen sections revealed poorly differentiated non-small cell carcinoma. Microscopic examination of the tumor revealed solid sheets of cells divided into irregular masses by fibrous strands and the majority of the cells were large and pleomorphic containing conspicuous nucleoli. Complete lymph node dissection was performed, and all but one subcarinal lymph node were free of tumor. Examination by electron microscopy showed multiple small foci of glycogen joined by rare rudimentary cell junctions. A few dense-cored (neurosecretory) granules were identified. Immunohistochemical staining showed the tumor expressed vimentin, CD99 (HBA71), MYC, neurofilament, synaptophysin,
CD10, CDR45 and KI-67, but not the following surface markers: MYF, Pancytokeratin, cytokeratin 7, cytokeratin 20, chromogranin, S-100, PSA and RCA (Figure 2). A diagnosis of primitive neuroectodermal tumor (PNET) of the lung was confirmed.

Discussion

PNETs are exceedingly rare in the adult population. In the chest, PNET primarily involves the chest wall or the lung as a distant metastasis of an extra-thoracic soft tissue sarcoma. PNET rarely presents as a primary pulmonary neoplasm with very few documented cases of primary lung PNET in the literature.

PNETs are part of a spectrum of neoplastic diseases known as the Ewing’s sarcoma family of tumors, which also includes atypical Ewing’s sarcoma, Askin’s tumor (malignant small cell tumor of the thoracopulmonary region), Neuroblastoma and paravertebral small cell tumor. In this particular case, the tumor was confined to the posterior aspect of the right lower lobe with no chest wall involvement, suggesting a primary lung tumor as opposed to the more common sarcoma of the chest wall.

Over the last decade, diagnosis has greatly improved with the introduction of an array immunohistochemical markers. The majority of Ewing’s sarcomas (84%) have a stereotypical immunophenotype with the expression of CD99 in all cases, FLI1 in 90% of cases and no expression of cytokeratins or desmin. In cases where morphology is inconclusive, cytogenetic analysis has rapidly become the standard for confirming the diagnosis. Most PNETs are characterized by a reciprocal translocation of the long arms of chromosomes 11 and 22, which creates a chimeric gene product of unknown function.

Because of their immunohistochemical and neurosecretory phenotype, PNETs are thought to originate from the neural crest, similar to neuroblastomas. However, PNETs can arise in organs not directly related to the neural crest (i.e. kidney) suggesting other histogenic possibilities.

Mainstay of treatment for PNET is doxorubicin-based chemotherapy. For patients with high-risk tumors (primary...
larger than 5 cm, local recurrence or of high grade), overall survival is improved with regimens including anthracycline and ifosfamide with growth factor support with survival benefit noted only for extremity sarcomas.

The decision of surgical resection was based on the pre-operative clinical and radiographic staging (I-B). Surgery revealed a positive lymph node consequently changing the stage to II-B. The patient was given adjuvant chemotherapy with one cycle of cyclophosphamide, adriamycin and vincristine followed by two cycles of ifosfamide, etoposide and mesna. The patient is currently doing well with no evidence of recurrence after nine months of treatment.

Conflict of interest statement

This manuscript has been approved by all of the authors. None of the authors have any conflict of interest to declare in relation to this work.

References