Ewing sarcoma, first described in 1921 (1), represents the second most common primary osseous malignancy in children and young adults, surpassed in incidence only by osteogenic sarcoma (2). Common locations include the axial skeleton, as well as the diaphyseal portion of long bones, most commonly those of the lower extremities (3). However, extraskeletal Ewing’s sarcoma, first described in 1969 (4), represents a less frequent but histologically similar entity that can originate within a wide array of extraosseous/soft-tissue locations, including the retroperitoneum, chest wall, or paravertebral space (5). Primary pulmonary involvement is rare; to our knowledge, only about twelve cases have been described in the literature.

Case report

A 23-year-old male presented at an outside institution with cough. A radiograph was recommended but not obtained due to lack of insurance. Two months later, the patient developed hemoptysis with interval worsening of his cough, as well as worsening dyspnea on exertion. Five months later, significant weight loss of 9 pounds over a 2-month period was also noted. During this time, he received several courses of antibiotics, with no significant improvement in clinical symptoms.

A chest radiograph was eventually obtained (Fig 1A). Partial opacification of the right hemithorax was noted, spanning the pleural and mediastinal surfaces. A noncontrast CT of the chest was subsequently performed (Fig. 1B). A large soft-tissue mass was noted within the right hemithorax, centered within the lung parenchyma and demonstrating diffuse, speckled internal calcifications. The mass insinuated within the mediastinum, with significant mass effect upon the right mainstem bronchus; it also extended to the pleural surface and chest wall, however without definite evidence for invasion.

CT-guided biopsy yielded an undifferentiated malignancy. Metastatic workup, including MRI of the brain, CT of the abdomen and pelvis, and a nuclear medicine bone scan demonstrated no evidence for distant metastases. The patient was then transferred to our facility for continued metastatic workup and management. PET/CT confirmed a metabolically active mass (SUV = 14.7) in the right hemithorax within the lung parenchyma, with no additional foci of increased radiotracer activity to suggest metastatic disease (Fig. 1C).

Review of the final pathology of the original biopsy specimen at our institution demonstrated densely packed neoplastic cells in a background of fibromyxoid stroma with patchy hemorrhage, fibrosis, and geographic necrosis (Fig. 2). The neoplastic cells showed strong membranous CD99 (O-13) immunoreactivity, characteristic of Ewing’s sarcoma/primitive neuroectodermal tumor (PNET). The EWSR1 gene rearrangement was demonstrated by fluorescence in-situ hybridization studies. The patient was initially started on chemotherapy as per Ewing sarcoma BAT pro-
Primary extraosseous Ewing sarcoma of the lung: Case report and literature review

tocol. After six cycles of chemotherapy, there was significant decrease in size of the mass (Fig. 3A), and the patient underwent right upper lobectomy, right mainstem bronchus resection, right sixth rib resection, and mediastinal tumor resection. Chemotherapy was resumed, along with radiation. It has now been six months since initial diagnosis, and followup CT imaging demonstrates post-surgical changes with no evidence of disease recurrence (Fig. 3B).

Discussion
The Ewing sarcoma family of tumors (ESFT), characterized histologically by primitive small round cells of neuroectodermal origin, includes classic osseous Ewing sarcoma, PNET, Askin tumor (Ewing sarcoma of the chest wall) and extraosseous (soft-tissue) Ewing sarcoma (6). Extraskeletal Ewing sarcoma represents a small subset of the ESFT, though the exact incidence has not yet been ascertained (7). While more frequent sites of ESFT include the retroperitoneum, paravertebral space, and chest wall (5), specific organs of involvement described in the literature also include the kidneys, pancreas, colon, uterus, and ovaries (6). The prognosis for treated patients has been reported to be similar to that for Ewing sarcoma patients (8). The lung represents a rare organ of primary
Involvement, to our knowledge being described about twelve times in the literature. Initially reported in 1989 (9), it occurs in a wide age distribution spanning the pediatric and adult age range, with the mean age of onset around 20 years as compared to 15 years for osseous Ewing (5), and occurring more commonly in males than in females (6, 10).

Imaging characteristics of extraskeletal ESFT have been described in the literature, although characterization of pulmonary Ewing sarcoma has been limited.

In an attempt to identify common radiological features of pulmonary Ewing sarcoma, we reviewed the CT imaging findings described in prior case reports, or present on the images provided, and summarized the results in the table.

In all cases, a solitary mass was predominantly circumscribed or well-defined in nature. In the majority of cases (7 out of the 11 cases in which CT findings were described or could be evaluated), the lesion demonstrated a heterogeneous appearance, with regions of low density suggesting necrosis. For the majority of patients, a contrast-enhanced
CT was performed, with the mass usually showing a hypo-enhancing pattern. Amorphous calcifications (as seen in our case) were also described in two previous cases. Ipsilateral pleural effusion was noted in three cases, while adjacent invasion (either mediastinal or chest wall involvement) was seen in two cases. In summary, primary pulmonary Ewing sarcoma most commonly presents as a circumscribed solitary mass with heterogeneous CT appearance both on noncontrast as well as contrast-enhanced CT. Occasionally, intrallesional calcifications or an ipsilateral pleural effusion may be seen. Infrequently, a mass may demonstrate evidence of invasion of adjacent structures.

As for the differential diagnosis of solitary lung lesions, primary pulmonary neoplasms in children are rare; secondary malignancy, from primary tumors such as Wilms tumor, osteosarcoma, or rhabdomyosarcoma, are far more common (11)—however, these usually present with multiple lesions. Primary pulmonary neoplasms in children are more frequently malignant than benign (12). Carcinoid represents the most common primary pulmonary malignancy in children. Bronchogenic carcinoma and pulmonary blastoma represent the second and third most frequent pediatric primary pulmonary cancers, respectively. Sarcomas represent an even rarer subset of primary pulmonary malignancy in children. In all age ranges, the most frequently encountered thoracic sarcomas include angiosarcoma, rhabdomyosarcoma, sarcomatoid mesothelioma, and leiomyosarcoma, but those known to affect the pediatric population include rhabdomyosarcoma, Ewing sarcoma, PNET, and endobronchial fibrosarcoma (13).

While the diagnosis of extraosseous pulmonary Ewing sarcoma relies on pathologic features, awareness of this clinical entity can help the radiologist broaden the differential diagnosis of primary lung masses, leading toward early diagnosis, comprehensive staging, and prompt treatment.

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