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Case report

Radiological and pathological diagnosis of an incidental Askin tumor ☆,☆☆,★,★★

Brian Covello, MD[a,*], *Steven Hartman, MS*[b], *Sean Kaufman, MD*[c], *Orlando Enrizo, MD*[a]

[a] *Department of Interventional Radiology, Kendall Regional Medical Center, 11750 SW 40th St, Miami, FL 33175, USA*

[b] *Dr. Kiran C Patel College of Allopathic Medicine, Nova Southeastern University, 3200 S University Dr, Davie, FL 33328, USA*

[c] *Department of Pathology, Kendall Regional Medical Center, 11750 SW 40th St, Miami FL 33175 USA*

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ABSTRACT

A 53-year-old male with no significant past medical history presented with an acute traumatic fracture of his thumb. Preoperative chest radiograph before K-wire fixation demonstrated an incidental 9 cm opacity of the left lung. Chest computed tomography revealed a 6.3 cm aggressive appearing pleural-based mass with erosion and destruction of the underlying rib. The patient underwent percutaneous biopsy with interventional radiology, and pathology revealed a small round blue cell tumor with positive CD99 staining and a FUS-ERG chromosomal translocation. The patient was diagnosed with Askin tumor, a peripheral primitive neuroectodermal tumor of the thoracopulmonary region belonging to the Ewing sarcoma tumor family. Computed tomography and magnetic resonance imaging of Askin tumors may show features such as a heterogeneous soft tissue mass, pleural effusion, rib destruction, hemorrhage, necrosis, and cystic degeneration. Askin tumors typically exhibit the EWS-FLI1 fusion mutation, although FUS-ERG chromosomal translocation has been described. Both rarity and variability of Askin tumors present a diagnostic challenge for clinicians. Collaborative effort amongst radiologists and pathologists is essential for diagnosis.

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* Corresponding author.
E-mail address: brcovello@gmail.com (B. Covello).

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Introduction

Askin tumors belong to the Ewing sarcoma and primitive neuroectodermal tumor (PNET) family of tumors, representing malignant small round cell tumors of the thorax [1]. Askin tumor, first described by Askin and Rosai in 1979, is a rare disease that is most commonly diagnosed in children and young adults [2]. Extraskeletal Ewing sarcoma, soft-tissue PNET, and Askin tumor commonly share chromosomal and histological similarities, including the cytogenetic marker t(11;22)(q24;q12) [1]. An estimated 85% of reported cases of Ewing sarcoma are shown to have an *EWS-FLI1* fusion between chromosomes 11 and 22 [3]. As such, their pathological appearance is identical to Ewing sarcoma of the bone. Radiologically, Askin tumors can have a wide variety of presentations on imaging. Computed tomography (CT) typically shows a large heterogeneously attenuating chest wall mass with associated pleural effusions [1]. On CT, Askin tumors may also show intrathoracic and extrathoracic extension, rib erosions, necrosis, nonuniform pleural thickening, or cystic degeneration [1,4]. Because Askin tumor is rare, identification via a variety of imaging diagnostic modalities is important, as patients may be asymptomatic at initial presentation. Askin tumors carry a poor prognosis, and early diagnosis helps initiate treatment through referral to a multispecialty team. Herein, a case of an incidentally discovered anterior chest wall Askin tumor with a rarely seen *FUS-ERG* chromosomal translocation is described.

Case report

A 53-year-old male with no significant past medical history presented to the emergency department with an acute traumatic fracture of his right thumb. Orthopedic surgery was consulted, and the patient was planned for K-wire fixation of his fractured thumb.

A preoperative chest radiograph revealed a 9 cm. rounded opacity overlying the periphery of the left mid lung (Fig. 1). Follow-up CT of the chest showed a 6.3 × 6.5 × 5.4 cm aggressive appearing pleural-based mass along the left anterior chest wall with erosion and destruction of the underlying left third rib (Fig. 2). Physical exam noted a palpable diffuse density in the left hemithorax. Cardiothoracic surgery was consulted by the medical team and recommended biopsy with interventional radiology (Fig. 3).

A follow-up biopsy of the mass revealed a small round blue cell tumor (Fig. 4). Diagnostic workup with immunohistochemistry revealed positive staining for CD99. Further molecular studies detected a FUS-ERG chromosomal translocation. The patient was subsequently diagnosed with an Askin tumor of the chest wall by pathological confirmation.

Throughout his hospital stay, the patient experienced no cardiopulmonary symptoms. After undergoing K-wire fixation of his right thumb with orthopedic surgery, he was subsequently discharged and referred to a facility with a multidisciplinary oncology team for further treatment of his Askin tumor and follow-up.

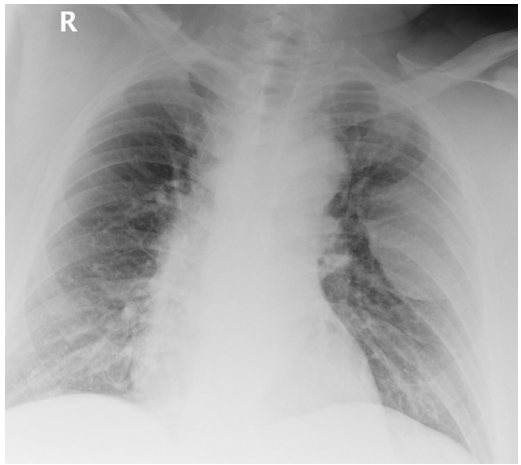

Fig. 1 – Anteroposterior portable chest radiograph shows a 9 cm rounded opacity overlying the left mid lung zone. The anterior portion of the left third rib is obscured by this opacity

Discussion

Askin tumors are of primitive neuroectodermal origin and typically arise from the soft tissues of the thoracopulmonary region. Most patients with Askin tumor are asymptomatic at clinical presentation, although fever, anorexia, and weight loss have been reported [1]. The current patient was asymptomatic, and a preoperative chest radiograph before K-wire fixation of a traumatic thumb fracture ultimately led to the discovery of an incidental Askin tumor.

Askin tumors have been reported in the chest wall, rib periosteum, lung, and anterior mediastinum [4]. Imaging characteristics are nonspecific; however, these tumors are most often seen as a heterogeneous soft-tissue mass in the thorax. CT and magnetic resonance imaging (MRI) findings can often show rib erosions, bone destruction, and nonuniform thickening of the pleura [1]. Askin tumors associated with hydropneumothorax, enhancing nodular pleural deposits, calcifications, necrosis, and cystic degeneration have been reported [2,4,5].

As Askin tumors typically present in childhood, the imaging differential includes primary Ewing's sarcoma of the rib, neuroblastoma, Langerhans's cell histiocytosis, Wilms tumor, and non-Hodgkin lymphoma [4-6]. Askin tumors less commonly present in older adults, as seen in the current case. Older age at presentation further broadens the differential diagnosis to more likely include rhabdomyosarcoma, chondrosarcoma, lymphoma, neurofibroma, as well as a metastatic disease [4]. When the mass is mediastinal or paraspinal and in a child, neuroblastoma should be considered. Elevated catecholamine levels seen in neuroblastoma differentiate it from Askin tumor [6]. In addition, neuroblastomas are known to have early enhancement, invade surrounding vasculature, and in contradistinction to the typical Askin tumors, may show coarse calcifications [7].

MRI typically shows a large heterogeneous mass with intermediate T1 and high T2 signal [1]. Areas of hemorrhage

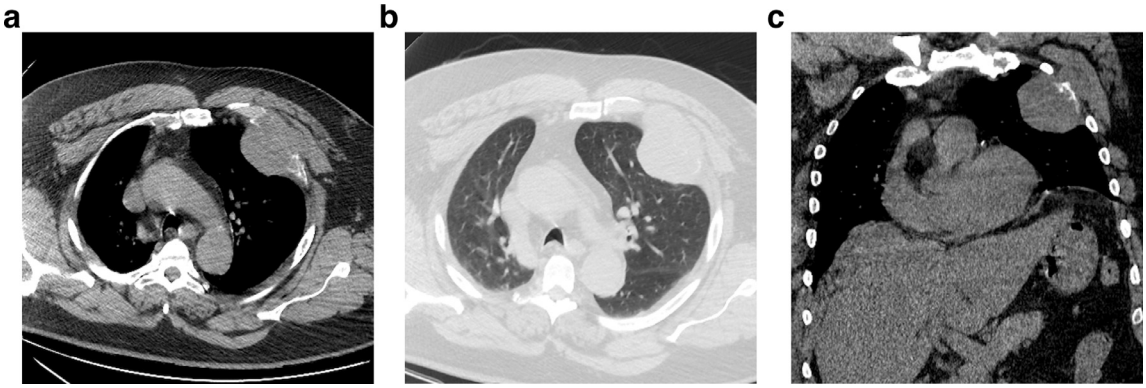

Fig. 2 – (A) Axial slice computed tomography of the chest reveals an aggressive appearing large pleural-based mass in the left anterior chest wall underlying the left anterior third rib. Osseous erosions of the third rib are present. These findings are consistent with malignancy until proven otherwise. (B) Axial slice computed tomography of the chest in lung windows confirms that the mass is pleural based. No other lung masses are noted. (C) Coronal reformatted computed tomography of the chest and abdomen helps confirm the size and location of the pleural based mass, which measures 6.3 x 6.5 x 5.4 cm

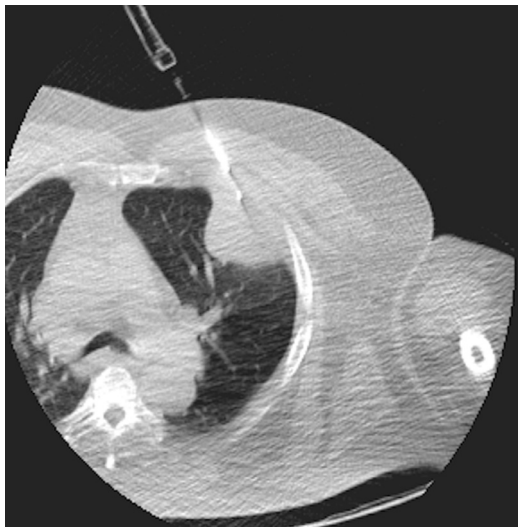

Fig. 3 – Axial slice computed tomography of the mass during biopsy with interventional radiology shows the appropriate placement of the needle tip within the mass

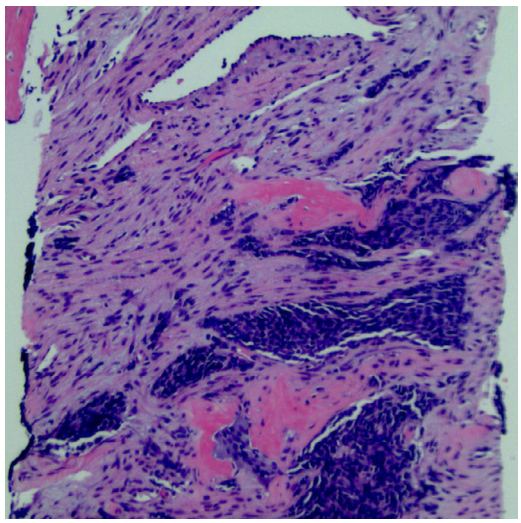

Fig. 4 – 10x magnification histological slide with hematoxylin and eosin staining showing small blue cell tumor.

and necrosis may be visualized on MRI through a high signal on T1 and T2 weighted images [1]. Vascular enhancement after intravenous gadolinium may occur and indicates neoplastic hypervascularity [1]. Fluorodeoxyglucose-positron emission tomography/CT may also help provide clarification, as Askin tumors have presented as fluorodeoxyglucose-avid masses [7]. Nevertheless, no set of imaging characteristics are pathognomonic for Askin tumors. Therefore, close coordination amongst radiologists, pathologists, and clinicians is critical for diagnosis.

Extraskeletal Ewing sarcoma, soft-tissue PNET, and Askin tumor commonly share chromosomal and histological similarities, including the cytogenetic marker t(11;22)(q24;q12), with a resulting reciprocal translocation of chromosome 11, which codes for EWS transcription factor, and chromosome 22, which codes for FLI1 [1,3]. In total, 85% of Ewing sarcoma family tumors exhibit this EWS-FLI1 fusion mutation [3]. In rare cases such as the one described, FUS substitutes for the EWS gene, with translocation t(16;21)(p11;q24) producing the FUS-ERG fusion [3]. This mutation was initially described in an Ewing sarcoma in 2007 and represents <5% of Ewing sarcoma family genetic mutations [3].

Immunohistochemistry staining for CD99, a membranous protein expressed on most Ewing sarcomas, round cell sarcomas, low-grade fibromyxoid sarcoma, and synovial sarcoma, provided an early clue as to the neuroectodermal origin of the mass in the current case presentation [8]. Further molecular studies allowed for the detection of a *FUS-ERG* chromosomal translocation, which provided the actual confirmation of the Ewing sarcoma family tumor. Ewing family tumors have

been shown to exhibit a range of chimeric transcripts created from specific chromosomal translocations [3]. This unusual *FUS-ERG* translocation may serve as a pathologic point of reference to any future cases with similar translocations and add to the growing knowledge of the characterizations, treatment outcomes, and prognostic predictions of these malignancies.

Due to the interdisciplinary treatment team approach, Askin tumor survival has somewhat improved from a median survival of 8 months in 1979 to a 2-year survival of 38% [1]. Askin tumor recurrence and metastases are common, occurring in over 50% of patients [1]. In cases of recurrence, CT is the modality of choice for the evaluation of pulmonary metastases. Less commonly, the hepatic and adrenal metastatic disease has been described [1].

A consequence of the rarity of this disease is a limited number of small-scale, single-institution studies and a lack of standardized treatment protocols. Current mainstay treatment methods may include chemotherapy and surgery, with new studies showing favorable results of neoadjuvant chemotherapy [9]. Further studies are needed to help guide the diagnostic and treatment process. An interdisciplinary approach to both diagnosis and treatment remains critical to improving survival.

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