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Askin Tumor Presents with Ewing Sarcoma of the Femur in Young Female Patient: Case Report and Literature Review

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Learning Point of the Article:

Consider metastatic Ewing sarcoma with an asymptomatic Askin tumor in children presenting with limb pain, emphasizing early comprehensive imaging and multidisciplinary management.

Abstract

Introduction: Askin tumor is a rare primitive neuroectodermal tumor originating in the thoracopulmonary region. It belongs to the Ewing sarcoma family and is known for its aggressive nature, typically arising from the chest wall. Ewing sarcoma is the second most common primary malignant bone cancer in children and adolescents, which arises from the neuroectoderm of bone and extra-skeletal soft tissue. Its characteristic chromosomal translocation between chromosomes 11 and 22 causes the EWS-FLI protein to develop in about 80–90% of patients, which might be helpful in targeted therapy in the future. However, Askin tumors are uncommon presentations of Ewing sarcoma of the femur at the time of diagnosis, exclusively in pediatric patients.

Case Report: A 12-year-old female patient presented with right thigh pain and swelling for 4 months associated with limping; the patient denied any respiratory symptoms. An ultrasound scan was done for both thighs and revealed Ewing sarcoma with bony metastasis as an initial diagnosis. At the same time, a computed tomography scan of the chest confirmed bilateral lung nodules. Subsequently, a needle biopsy from the right thigh and right posterior chest wall was completed for histopathological examination; the results showed that the cause of the thigh pain is Ewing sarcoma of the right thigh with bone metastases and Askin tumor.

Clinical Discussion: Ewing sarcoma is a highly metastatic form of sarcoma that ranks as the second most prevalent primary malignant bone tumor, constituting 10–15% of all soft-bone sarcomas. Around 20% of patients present with metastatic disease at the time of diagnosis, and among these cases, some have lung or pleural involvement. The presentation of Ewing sarcoma with bony metastasis as an Askin tumor is extremely rare. Almost all literature reviews reported each of these types separately. After an extensive review of the literature, we have not found any study reporting Ewing sarcoma of bone metastasis with Askin tumor at the time of diagnosis. This case uniquely shows their co-occurrence, a combination that has not been previously detailed in the literature. Individualized treatment plans tailored to pediatric patients' unique needs can significantly improve survival rates.

Conclusion: Careful monitoring of Ewing sarcoma should be prioritized, and appropriate treatment plans should be established during diagnosis to achieve satisfactory outcomes and further emphasize the significance of careful and early evaluation, even for typical clinical signs. This case underscores the importance of early diagnosis and a multidisciplinary approach to achieving favorable outcomes in rare tumors such as Ewing sarcoma and Askin tumor.

Keywords: Case report, Ewing sarcoma, Askin tumor, femur, bone metastasis, young age.

Introduction

Ewing sarcoma is an aggressive malignancy characterized by the rapid proliferation of small, round blue cells. It commonly metastasizes early to the lungs, bones, and lymph nodes, leading to a poor prognosis. It is described as the second most common tumor after osteosarcoma, mostly affecting 5–20-year-olds with male predominance, usually involving the long bones of the extremities, such as the femur 16.4%, the pelvis, and the ribs [1,2].

Ewing sarcoma family of tumors (ESFTs) consists of extraosseous Ewing sarcoma, skeletal Ewing sarcoma,

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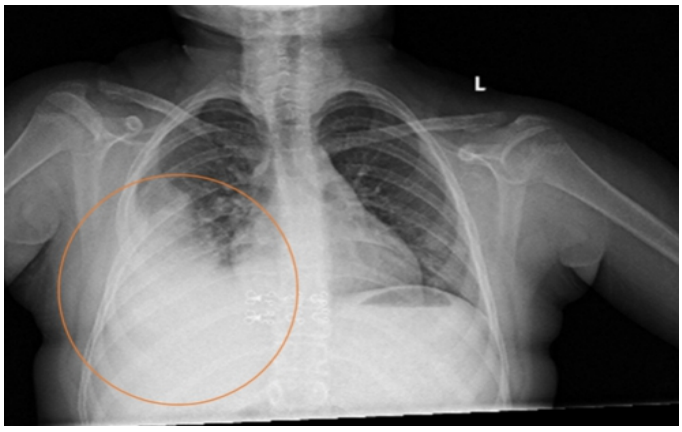

Figure 1: Chest radiograph showing a homogeneous radiopacity in the right lung.

primitive neuroectodermal tumor, and Askin tumor.

Askin tumor is a rare malignant neoplasm of neuroectodermal origin, which develops from the soft tissues of the thoracopulmonary wall; it is predominantly seen in children, most commonly in males. The patients may have dyspnea, cough, Horner's syndrome, and regional lymphadenopathy. They usually present on computed tomography (CT) with a large, heterogeneous, extrapulmonary mass lesion with or without destruction of the ribs and frequently associated with pleural effusion. 10% of the patients usually have evidence of metastasis at diagnosis. Common sites of metastasis are the lungs, bone marrow, bone, and mediastinal lymph nodes. The treatment consists of neoadjuvant chemotherapy followed by surgical excision with or without adjuvant radiotherapy [3, 4]. It is associated with poor prognosis and short survival because local recurrence and metastases are frequently seen in Askin tumor. The reported overall survival is approximately 60% at 5 years [3, 5]. In an attempt to further define prognostic factors, appropriate management and treatment outcome of this rare neoplasm, we reviewed our experience with this relatively rare round cell tumor. Here, we report a case of extraosseous Ewing sarcoma with Askin tumor in a 12-year-old female.

Case Report

We present a case of a 12-year-old white Middle Eastern female with no significant past medical history who presented to the outpatient clinic complaining of right thigh pain and swelling associated with limping for 4 months. With normal regular pulses and normal vital signs. She has no family history of Ewing sarcoma or Askin tumor, and she denied any history of recent trauma, prolonged immobilization, or infection. Furthermore, denying any history of back pain, chest pain, chest tightness or discomfort, no fever, night sweats, or weight loss.

A timeline, including symptom onset, diagnostic steps, treatment interventions, and follow-up, is provided in Table 1.

On examination, it revealed an immobile, tender, tense swelling in the right thigh without any local increase in temperature. The range of motion of the joint and distal neurovascular status were normal. No palpable inguinal lymph nodes.

On investigation, as shown in Table 2, white blood cells, erythrocyte sedimentation rate, and C-reactive protein were elevated. Fig. 1 revealed that a chest X-ray was done on the day of admission as a clinical protocol of the hospital.

A right thigh X-ray was done, which revealed that there was about 7.5 × 5 × 5 soft-tissue lesion, also confirming a lytic bone lesion. Fig. 2 shows magnetic resonance imaging findings revealing a heterogeneous intramedullary mass lesion of the right femoral shaft with a soft-tissue component extending over a distance of 21cm in length, which appears hypointense on T1 and hyperintense on T2-weighted sequence with heterogeneous post-contrast enhancement and heavy diffusion restriction, which was associated with diffuse cortical bone destruction and large soft-tissue extraosseous extension surrounding the affected bone. Similar enhanced abnormal intramedullary signals were seen in the left iliac bone and distal right femoral metaphysis, with the largest measures up to 17 × 16 mm in the left iliac bone. Suggestive of Ewing sarcoma with bony metastasis.

Fig. 3 shows a CT scan with contrast for lung, mediastinal and bone were done which presented bilateral lower lung lobe

Table (1): Timeline of Clinical Events

Event	Timeline
Symptom onset	4 months before admission
Initial Imaging (X-ray, MRI, CT)	At admission
Biopsies performed	Shortly after imaging
Diagnosis confirmed	Post-biopsy
Start of chemotherapy	After diagnosis (VDC protocol)
Surgical resection	Post-chemotherapy
Post-op chemo/radiotherapy	Following surgery
Bone marrow biopsy	Post-treatment

Table (2): Laboratory Results upon Admission Day

Test Name	Normal Range (children)	Results
White blood cells (WBCs)	4.6-10.2mg/dl	19.6
Neutrophils	2-6.9 mg/dl	15.5
Erythrocyte sedimentation rate (ESR)	0-20 g/dl	81
C-Reactive Protein (CRP)	8-10 mg/L	257
Calcium	86-10.2mg/dl	9.5
Alkaline phosphatase (ALP)	40-480unit/L	380.9

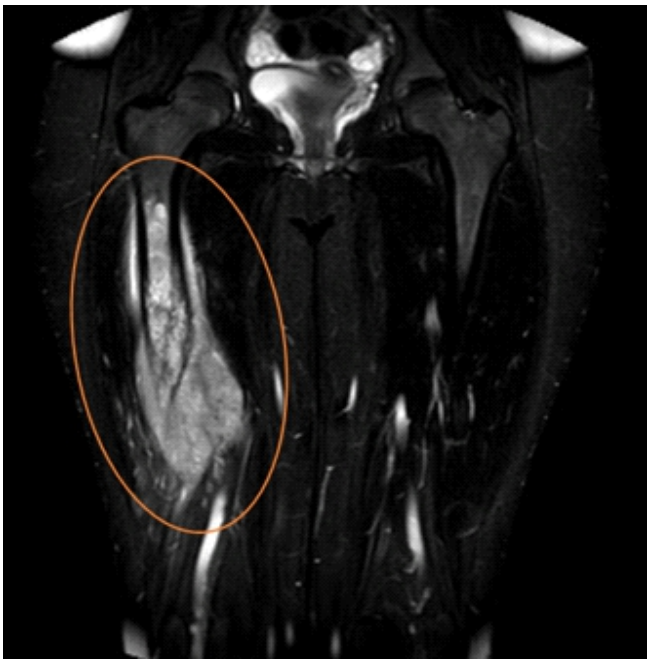

Figure 2: Magnetic resonance imaging of the thighs, revealing an aggressive right femoral bone malignancy.

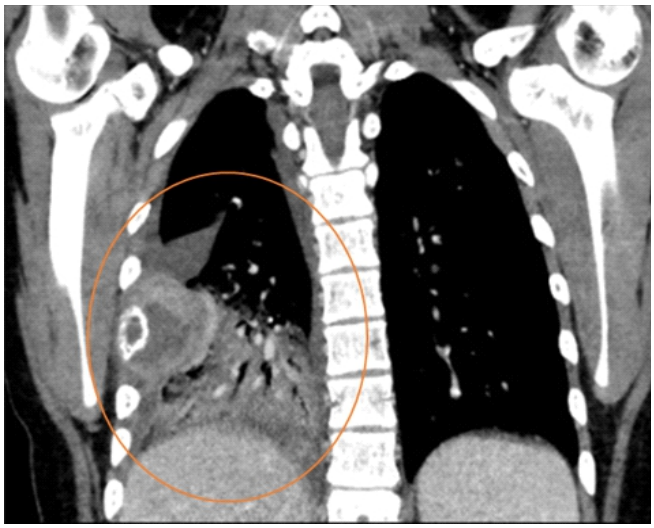

Figure 3: Computed tomography with contrast showing a heterogeneous mass in the right lung with a large soft-tissue component and pleural effusion.

nodules: The largest on the right measures 2 cm and on the left measures 1.5 cm and permeative destructive lesion in the right 5th rib associated with large soft tissue component measuring 9 cm and subsequent consolidation of the right lower lobe, with rim of right sided pleural effusion, which raises suspicion for Askin tumor, also a lytic bone lesion in the iliac side of the left sacroiliac joint measuring 2 cm.

Under sedation and ultrasound guidance, a needle biopsy was performed on the right thigh and posterior chest wall. Histopathological examination revealed microscopically small round neoplastic cells with hyperchromatic nuclei and minimal to focally moderate vacuolated cytoplasm. Mitotic activity was observed. Areas of tumor necrosis are seen; the background shows a desmoplastic reaction.

On immunohistochemistry, the tumor cells were positive for CD99 (membranous), whereas desmin, myogenin, CD45, and SOX10 were negative. A final diagnosis of Ewing sarcoma of the right thigh with bone metastases and Askin tumor.

Based on the patient's history, examination findings, and previous investigation, she was scheduled for pre-operative chemotherapy management, as a vincristine, doxorubicin, cyclophosphamide protocol was done over 6 cycles of chemotherapy. Re-evaluation chest CT and X-ray demonstrated a significant decrease in the previously described right 5th rib destructive lesion with residual soft-tissue component. Subsequently, the patient underwent a radical resection of the femur tumor as follows: an intra-operative procedure. After segmental bone resection, it underwent extracorporeal radiation; fixation and reaming were done for the proximal intramedullary canal and distal intramedullary canal with preservation of the growth plate area, then the reduction and fixation of bone were checked under C-arm image; the procedure was successfully completed. Post-operative chemoradiation was done. Bone marrow biopsy and aspiration were repeated and confirmed a normocellular marrow with trilineage hematopoiesis to ensure that the patient was in good health.

Discussion

Ewing sarcoma is a highly metastatic form of sarcoma that ranks as the second most prevalent primary malignant bone tumor, constituting 10–15% of all soft bone sarcomas, predominantly afflicting males between 10 and 15 years of age. Extraosseous Ewing sarcoma and Askin tumors are types of ESFT that are characterized by the presence of non-random chromosomal translocation producing fusion genes 11; 22-q24 [2]. Ewing sarcoma classically presents with leg pain, stiffness, or swelling; more than 50% of patients with Ewing sarcoma have intermittent pain that worsens at night. Around 20% of patients present with metastatic disease at the time of diagnosis, and among these cases more than 20% have lung or pleural involvement [6].

Askin tumors maintain a low incidence regardless of the new diagnostic technique. They are more common in males, and 80% of patients are children and young adults; however, there are reported cases of elderly patients. Usually, patients come with ambiguous symptoms including a palpable mass in the thorax, either painful or painless (the most common finding),

Table (3): Selected reported Askin Tumor Cases in Literature

Study	Treatment Approach	Survival Outcome
Zhang et al. (2016)	Surgery + Chemotherapy	~60% 5-year OS
Laskar et al. (2011)	Neoadjuvant chemo, surgery, radiation	Variable, high recurrence
Singh et al. (2016)	Chemo + Radiotherapy	Poor prognosis, rapid recurrence

Table (4): Differential Diagnosis Comparison

Condition	Distinguishing Features	Diagnostic Tools
Ewing Sarcoma	Small round blue cell tumor; CD99+, EWSR1 translocation	Histology, IHC, FISH/PCR
Askin Tumor	Chest wall origin, same histologic features as ES	Imaging, location, IHC
Osteomyelitis	Infection markers, fever, elevated CRP	Blood tests, imaging
Lymphoma	Lymphadenopathy, systemic symptoms	Biopsy, immunophenotyping
Neuroblastoma	Adrenal origin, elevated catecholamines	Urine catecholamines, MIBG scan

pleuritic pain, dyspnea, fever, cough, and weight loss [3, 4].

Our 12-year-old female patient presented with extraosseous Ewing sarcoma (Ewing sarcoma) with bony metastases and an Askin tumor at the time of diagnosis, with an unusual presentation of limping with mild swelling in the right thigh, pain and tenderness, but she denied any fever, night sweats, weight loss, or chest symptoms. During the investigation, we incidentally discovered two masses in the lung with minimal right-sided pleural effusion. This is different from typical presentations of Askin tumor, which often manifest with respiratory symptoms such as dyspnea and chest pain. The lack of thoracic symptoms in our patient underscores the importance of comprehensive imaging in cases of unexplained limb pain and swelling.

The presentation of Ewing sarcoma with bony metastasis as an Askin tumor is extremely rare. Almost all literature reviews reported each of these types separately, with only 17 cases identified in the literature reviewing Askin tumor. Table 3 presents a summary of selected reported cases, their treatment approaches, and outcomes. After an extensive review of the literature, we have not found any study reporting Ewing sarcoma of bone metastasis with Askin tumor at the time of diagnosis. This case uniquely shows their co-occurrence, a combination that has not been previously detailed in the literature.

The co-occurrence of Ewing sarcoma with Askin tumor, while extraordinarily rare, can be better understood by examining their shared histogenetic and molecular characteristics. Both are part of the ESFTs, defined by the presence of specific chromosomal translocations, most commonly t(11;22)(q24;q12), resulting in the EWSR1-FLI1 fusion gene. This fusion gene acts as an aberrant transcription factor that drives oncogenesis by dysregulating gene expression involved in cell proliferation, differentiation, and apoptosis. The identification of this translocation in both intraosseous and thoracopulmonary tumors supports the hypothesis of a shared origin from primitive neuroectodermal cells, albeit arising in different anatomical locations.

From a radiological perspective, positron emission tomography-CT (PET-CT) has become an essential tool in the staging and restaging of Ewing sarcoma and related tumors. It provides valuable information on metabolic activity, enabling detection of both skeletal and soft-tissue involvement with high sensitivity. In this case, although PET-CT was not initially used, its incorporation in follow-up evaluations could provide superior assessment of treatment response and early detection of recurrence.

Histologically, both tumors consist of small, round blue cells. Immunohistochemistry plays a critical role in differentiating them from other small round cell tumors. In our case, tumor cells showed diffuse membranous positivity for CD99, which, although not specific, is highly sensitive for ESFT. The negative markers (desmin, myogenin, CD45, SOX10) helped exclude rhabdomyosarcoma, lymphoma, and other mimics. In addition, newer markers such as NKX2.2, FLI1, and molecular confirmation by fluorescence in situ hybridization or reverse transcription-polymerase chain reaction for EWSR1 rearrangement can enhance diagnostic accuracy, especially in ambiguous cases [7].

The other differential diagnoses are inflammatory disorders and other malignancies. Clinical symptoms of Askin tumor mimic inflammatory disorders such as osteomyelitis, empyema, and TB, all of which can present with bone pain and systemic symptoms. On the side of malignancies in children include lymphoma (lymphadenopathy, splenomegaly), osteosarcoma, and neuroblastoma (elevated urinary catecholamines) [3, 5, 8]. A comparative overview of these conditions is presented in Table 4, highlighting their distinguishing clinical features and diagnostic modalities.

Given the aggressive nature and high metastatic potential of both tumors, a multimodal approach involving neoadjuvant chemotherapy, surgical resection, and adjuvant radiotherapy remains the cornerstone of treatment. Future therapeutic directions may include EWS-FLI1-targeted therapies and immune checkpoint inhibitors, which are currently under investigation in clinical trials.

Due to the disease's rarity, there are not many small-scale, single-

institution research studies and no established treatment guidelines. Emerging therapies, such as proton therapy, offer targeted approaches to minimize damage to surrounding healthy tissue, especially in pediatric cases. Furthermore, clinical trials exploring checkpoint inhibitors and molecular inhibitors for EWS-FLI1 fusion proteins may provide new hope for aggressive and metastatic disease management [9, 10]. The successful management of this case underscores the importance of a multidisciplinary approach, integrating oncology, surgery, and radiology expertise. Individualized treatment plans tailored to pediatric patients' unique needs can significantly improve survival rates. This plays a major role when combined with early clinical suspicion, especially in resource-limited settings, to overcome the challenging diagnoses of rare tumors such as Askin tumor and metastatic Ewing sarcoma due to limited access to advanced imaging and genetic studies.

Beyond medical management, addressing the psychosocial impact of such a diagnosis is critical, particularly in pediatric patients. Comprehensive care should include psychological support, rehabilitation, and counseling for both the patient and their family to improve overall quality of life. Furthermore, detecting a disease in its early stages often allows us an improved prognosis, reducing complications and engaging the patient with daily activities. This also reduces the need for more intensive and expensive interventions later. Specialists from various fields collaborate to develop a personalized treatment strategy that optimizes outcomes.

This case reinforces the need for heightened clinical suspicion, especially when initial symptoms are non-specific, and advocates for integrating advanced imaging and molecular diagnostics in early workup to facilitate timely and precise management [1,11].

Conclusion

This case highlights the rare co-occurrence of Askin tumor and metastatic Ewing sarcoma at initial diagnosis in a pediatric patient. Early diagnosis, comprehensive imaging, and a multidisciplinary approach are critical in managing such complex presentations. Future studies should explore the molecular relationship between these entities to inform targeted therapies and improve outcomes.

Askin tumor, also called Ewing sarcoma of the chest wall, is an extremely rare disease and is often misdiagnosed. Hence, careful monitoring should be prioritized. Appropriate and early multimodal treatment plans should be established during diagnosis to achieve satisfactory outcomes and to point out the significance of careful and early evaluation, even for typical clinical signs. This case highlights the importance of considering early diagnosis for intervention to occur quickly and to obtain a favorable outcome and optimal survival for the patient. Hence, the physicians should maintain a high index of suspicion when encountering patients, particularly children and young adults. It also emphasizes the importance of a multidisciplinary approach to ensure accurate diagnosis, effective treatment, and improved outcomes.

Clinical Message

Unexplained limb pain in children warrants thorough evaluation for metastatic Ewing sarcoma, including asymptomatic thoracopulmonary involvement, to facilitate early diagnosis and multidisciplinary treatment.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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