SUNTEXT REVIEWS

SunText Review of Case Reports & Images
ISSN: 2766-4589

Open Access
Case Report
Volume 5:5

Askin-Rosai Tumor; Exteremly Rare Oncology Case Report

Abuhajar RM[1,*], Elshahoubi A[2] and Alqawi O[3]

[1]*Department of Radiology, National Cancer Institute/Misrata-Libya*
[2]*Department of Pediatric Oncology, National Cancer Institute/Misrata*
[3]*Department of research, National Cancer Institute, Misurata-Libya*

[*]**Corresponding author:** Abuhajar RM, Department of Radiology, National Cancer Institute/Misrata-Libya; Tel: +218 91 6339496; E-mail: raabuhajar@gmail.com

Received date: 15 August 2024; **Accepted date:** 22 August 2024; **Published date:** 29 August 2024

Citation Abuhajar RM, Elshahoubi A, Alqawi O (2024) Askin-Rosai Tumor; Exteremly Rare Oncology Case Report. SunText Rev Case Rep Image 5(5): 140.

DOI: https://doi.org/10.51737/2766-4589.2024.140

Abstract

Askin-Rosai tumor is a peripheral primitive neoroectodermal tumor of thoracopulmonary region. The diagnosis of the tumor is difficult and often missed due to the rarity and confused with other tumors because it shares the other small round blue cells tumors. The tumor is aggressive and had been poor prognosis with short survival. Early diagnosis is imperative to start specific treatment immediately. We report a case of Askin-Rosai tumor in a 14 years old female patient presented with chest pain and admitted in pediatric oncology department/NCI-Misrata, Libya. Underwent chest CT scan for the patient which established early diagnosis with other full investigations including histopathology, immunochemistry as well as the routine laboratory analysis performed for confirmation and starting treatment.

Keywords: Ewing sarcoma family; Askin tumor; Imaging; Pediatric; Rarity

Introduction

Primitive neoroectodermal tumor (PNET) are identical to Ewing sarcoma and very rare malignant tumors originating from neoroectoderm. The incidence of Ewing sarcoma is about 4% of all sarcomas and the annual incidence in USA population is one case per million. PNET can be differentiated into central (cPNET) and peripheral tumors (pPNET). The central PNET are tumors of central nervous system (CNS) and mostly found among children and young adults, also the tumor can occur outside CNS such as pelvis or limbs [1-3]. Askin tumor that belonged to Ewing sarcoma family, arisen from bone or soft tissue in thoracopulmonary region. Askin-Rosai tumor firstly described in 1979, who reported 20 patients with mean age of 14 years old. The tumor is very rare and more found among children and adolescence than adults, it is predominant in males and nine times to be Caucasian race [4-7]. Its incidence in literature is extremely low and in Dickenson et al study the Askin tumor prevalence is 0.2 cases per million [4,5]. The tumor histopathology shows specific small round blue cells similar to Ewing sarcoma and PNET cells [8]. Askin tumor usually develops in chest wall as soft tissue and bony mass and occasionally develops in periphery of lung or mediastinum. The tumor represents with respiratory symptoms such as chest pain, dyspnea, palpable mass, weight and apatite loss [5,9]. The disease is highly malignant and aggressive that is why it has been poor prognosis and short survival; reported overall 60% at 5 years [10].

The diagnosis is difficult and highly misdiagnosed due to rarity of the disease with a lack of histopathology and immunochemistry analysis, so it is easily confused with the other round small blue cells tumors [11]. Imaging modalities are important and play vast majority in early diagnosis to start proper treatment. Non-invasive CT scan and MRI describe location, size, morphology, margin blood supply of the tumor, and possible invasion of pleura and lungs as well as presence of distant metastases. Correlation of imaging examinations, clinical, histopathological and immunochemistry analysis is the way to reaching the specific diagnosis of Askin-Rosai tumor [12]. Treatment dependent on size of tumor and metastases. Small size localized tumor without metastases can be resected surgically with chemotherapy and/or radiotherapy. A large tumor with metastases can be treated with chemotherapy and radiotherapy, also surgical excision may be required depending on tumor size, location and associated

symptoms. Unfortunately the recurrence of sarcoma and Askin tumor is common and carry poor prognosis despite combination of chemotherapy, radiotherapy and surgical treatment. Very few patients are still alive although patients received the treatment [2,6].

Case Presentation

14 years old female patient presented with upper left limb mild weakness and pain at left side of chest. Clinical examination revealed palpable non-mobile, hard mass at anterosuperior left side of the chest, normal function of respiratory, cardiovascular systems and other systems. Normal routine laboratory investigations. Underwent CT scan revealing a large mass 7.5 x 6.0 cm with well demarcated margin and heterogonous density causes destruction of 2nd rib of the left side of the chest. The mass bulged posteriorly and causes severe compression on the ipsilateral lung parenchyma, but no evidences of detectable invasion or infiltration. No pleural collection, pulmonary, hepatic, bone or other distant metastatic changes, no lymphadenopathy. CT scan play role in the establishment of early diagnosis of the tumor. CT scan guided biopsy and histopathologic study, hematoxylin eosin stained the specimen after sliced into sections and showed small round blue cells which are not deferent from neoblastic and rhabdomyoblastic cells. The tumor cells are uniform and rarely fusiform showed an aspect of Ewing's sarcoma. The immunohistochemistry study showed same picture of Ewing sarcoma family. Correlation of clinical, imaging morphologic, histopathologic and immunochemistry invitations confirm the diagnosis of Askin-Rosai tumor involved 2nd rib of left side of the chest. Patient received preoperative 6 cycles of neoadjuvant chemotherapy and later underwent thoracotomy with complete resection of the left 2nd rib. The mass at the rib is hard and not invading the surrounded structures and closely attached to the underlying pleural membrane. Delicate dissection of the mass and completely separated from the underling pleural membrane. Patient is very well and it is planned to receive complete 14 cycles of chemotherapy.

Discussion

Askin–Rosai tumor is a member of Ewing sarcoma family/primitive neoroectodermal tumors which occurs at thoracopulmonary site and it is founded mainly in children and adolescence.

Askin tumor shares PNET and Ewing sarcoma in imaging morphology, chromosomal translocations, morphology, immonohistochemistry and ultrastructure.

Diagnosis of Askin tumor is not easy because a limited number of cases have been documented and its microscopic appearance is similar to Ewing sarcoma, neuroblastoma and rhabdomyosarcoma histopathology. Our case is a girl 14 years old presented with thoracopulmonary symptoms; chest pain, associated with palpable chest wall mass. Establishment of an accurate diagnosis in imperative for instituting specific therapies. CT scan chest showed a large mass with heterogeneous density destructed anterior 2nd rib of left side of chest bulging posteriorly causes severe compression on the adjacent part of the lung, but no invasion or infiltration occurred. The clinical information, imaging morphology, histopathology and immunochemistry with site of the mass confirm the diagnosis. No evidences of local or distant metastasis and no lymphadenopathy. The treatment included neoadjuvant chemotherapy, complete surgical resection of the 2nd rib of chest left side. Patient received cycles of vincristine, doxyurudicin and cyclophosphamide before surgical resection and later patient underwent complete surgical excision. At present time it is planning for adjuvant chemotherapy is planned.

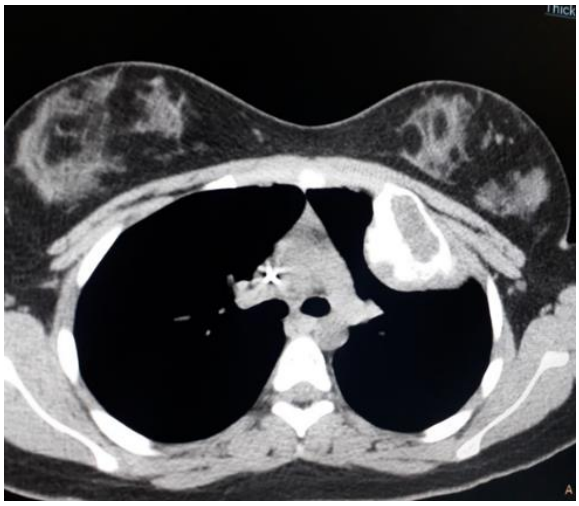

Figure 1: *Axial section of chest CT scan showed a large heterogamous mass at left chest wall.*

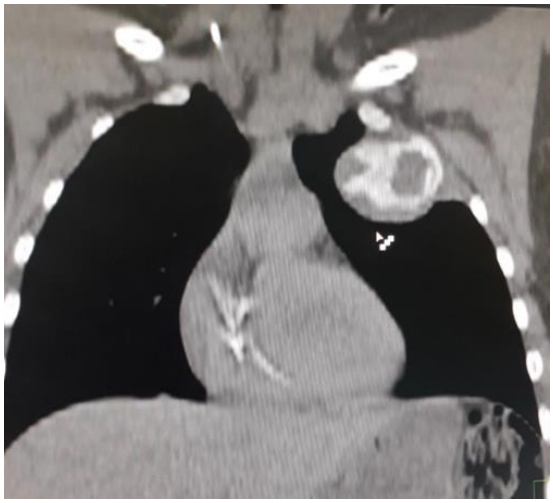

Figure 2: *Coronal section of chest CT scan showed a large heterogamous mass destructed the 2nd left chest rib.*

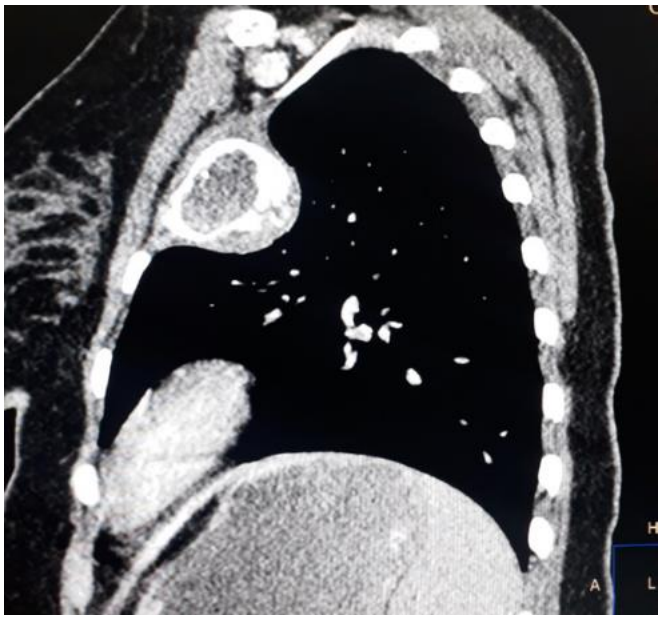

Figure 3: *Sagittal section of chest CT scan showed a large heterogamous mass bulging posteriorly compression on the adjacent pulmonary part.*

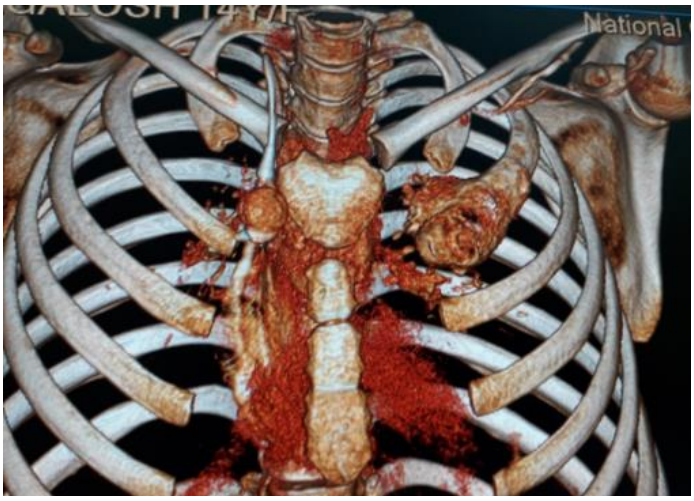

Figure 4: *Chest CT scan (3D) showed a mass in anterior 2nd rib of left chest side extended posteriorly along the rib.*

The Askin-Rosai tumor has predilection to recurrence and the prognosis is very poor, whereas few cases still alive after treatment and follow up. We hoop that our patient will be recover and recuperate after receiving combined treatment and resultant in optimal outcome (Figures 1-5).

Conclusion

Askin-Rosai tumor is extremely rare aggressive malignant tumor of thoracopulmonary region, more common in children and adolescence than adults. Histopathology study reveal similar cells in Askin tumor as in Ewing sarcoma and PNET family. We reported proved Askin-Rosai tumor.

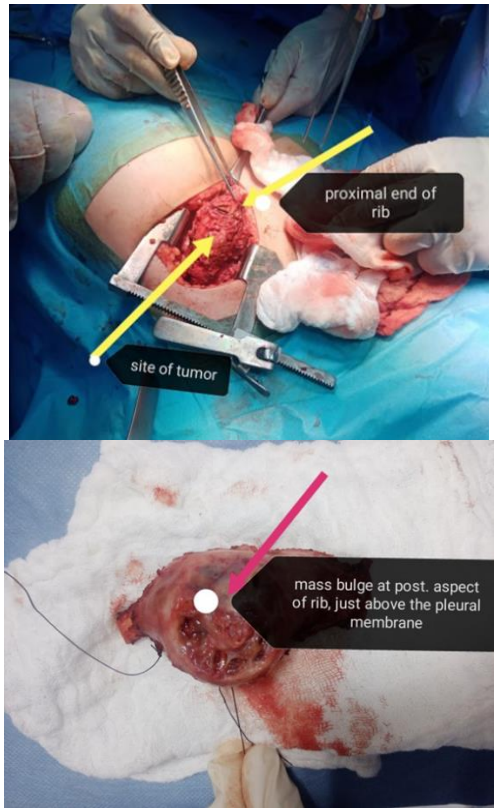

Figure 5: *Askin tumor; surgical resection.*

References

1. Khan S, Abid Z, Haider G, Bukhari N, Zehra D, Hashmi M, et al. Incidence of Ewing's sarcoma in different age groups, their associated features, and its correlation with primary care interval. Cureus. 2021; 13.
2. Korol T, Blazhko S, Rudenko H, Khromykh K. Principles of diagnosis and treatment of Askin's tumor in children: case report. Wiad Lek. 2022; 75: 551-554.
3. Tan Y, Zhang H, Ma G-L, Xiao E-H, Wang X-C. Peripheral primitive neuroectodermal tumor: Dynamic CT, MRI and clinicopathological characteristics-analysis of 36 cases and review of the literature. Oncotarget. 2014; 5: 12968-12977.
4. Covello B, Hartman S, Kaufman S, Enrizo O. Radiological and pathological diagnosis of an incidental Askin tumor. Radiol Case Rep. 2021; 16: 1245-1248.
5. Benbrahim Z, Arifi S, Daoudi K. Askin's tumor: a case report and literature review. World J Surg Oncol. 2013; 11: 10.
6. Keehn B, Jorgensen SA, Towbin AJ, Towbin AJ, Towbin R. Askin tumor. Applied Radiol. 2017; 32-33.
7. Peng L, Deng H-Y, Yang Y. Askin tumor in the chest wall. Thoracic cancer. 2021; 12: 407-408.
8. Mathur SK, Jain P, Singh S, Rattan KN, Dahiya N, Singh S. Askin tumour: A rare childhood thoraco-pulmonary malignancy of Ewing's family of tumours. Cumhuriyet Med J. 2012; 34: 352-355.

9. Tingting X, Yubao G, Yongxin C, Jingxu L. Askin tumor: CT and FDG-PET/CT imaging findings and follow-up. Med. 2014; 93: 1-6.
10. Singh A, Abhinay A, Kumar A, Prasad R, Amrita Ghosh A, Mishra OP. Askin tumor: A rare neoplasm of thoracopulmonary region. Lung India. 2016; 33: 196-198.
11. Zhang K, Lu R, Zhang P, Shen S, Li X. Askin's tumor: 11 cases and a review of the literature, Oncol Lett. 2016; 11: 253-256.
12. Berrada K, El Harass Y, El ouali I, Latib R, Omor Y. Radiological diagnoses of an incidental Askin tumor: About one case. Radiol case rep (RCR). 2023; 18: 4137-4139.