CASE REPORT

A Teenage Girl with a Large Lung Mass

Liaw G[a], Nadarajan C[a], Siti K[b], Roudhotul N[b]

[a]Department of Radiology, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan
[b]Department of Radiology, Queen Elizabeth II Hospital, Kota Kinabalu, Sabah

Keywords
pulmonary, ewing Sarcoma, extraskeletal

Corresponding Author
Dr. Chandran Nadarajan
Department of Radiology, School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia.
Email: drchandran@usm.my

Received: 1st Dec 2021; Accepted: 27th Dec 2021

Doi: https://doi.org/10.31436/imjm.v22i2

ABSTRACT

Extraskeletal Ewing Sarcoma is rarely reported. A fifteen-year-old girl presented with the right-sided pleuritic chest pain, shortness of breath, and productive cough with whitish sputum associated with poor oral intake. She subsequently underwent contrast-enhanced Computerized Tomography thorax, which showed lobulated heterogenous hypodensity occupying the entrance of the right superior pulmonary vein, extending to the left atrium.

INTRODUCTION

Ewing's Sarcoma (ES) of the bone is the second most common primary malignancy of the bone in children and adolescents after osteosarcoma, and the fourth most common primary bone malignancy after multiple myeloma, osteosarcoma, and chondrosarcoma. It is commonly seen in patients with meta-diaphysis. Extraskeletal Ewing Sarcoma (EES) of the bone is rare compared to ES, which is between 15 to 20 % of that ES of the bone. It is seen in patients of ages ranging between 20 months and 30 years old, with the site most commonly reported at paravertebral, lower extremities, chest wall, retroperitoneum, pelvis, and hip, as well as in upper extremities.[1] EES is extremely rare to arise from the lung, [2] which is only a few reported cases in the literature. [3] Herein, we report a case of a young girl with primary ES of the lung.

CASE PRESENTATION

A fifteen-year-old girl, presented to the Hospital Queen Elizabeth I, Emergency and Trauma Department, with a history of right-sided pleuritic chest pain, shortness of breath, productive cough with whitish sputum associated with poor oral intake for one-day duration. Otherwise, she denies any history of fever, hemoptysis, loss of weight, or recent ill/pulmonary tuberculosis contact. She was alert during the presentation, cachexic, and tachypneic. She had reduced air entry over the right lung on auscultation, with dullness on percussion. The patient was started on antibiotics, intravenous Augmentin 1.2gram three times per day and tablet Azithromycin 500 milligram once per day, for community-acquired pneumonia with right pleural effusion. Subsequently, diagnostic pleural tapping was performed, hemorrhagic pleural fluid was drained. Then, chest tube insertion was performed. At this point, the diagnosis was revised to right exudative pleural effusion likely secondary to mediastinal mass, covering for community-acquired pneumonia. Due to an increase in oxygen requirement from nasal prong 2 to face mask 5 litre, low-grade fever, as well as persistent tachycardia on electrocardiography (ECG), the diagnosis of pulmonary embolism needed to be ruled out. Thereafter, Computerized Tomography (CT) pulmonary angiography (Figure 1) was done on day-five admission showing hemopneumothorax with intra-lobar hematoma. No evidence of pulmonary embolism.

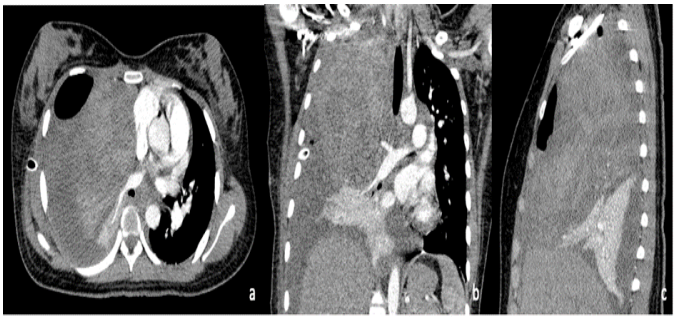

Figure1: above shows axial (a), coronal (b), and sagittal (c) images of CT Pulmonary angiography. These images showed a heterogenous hyperdense lesion confined to the right upper lobe, causing a mass effect to the right middle and lower lobes. Presence of heterogeneous hypodense collection with split pleural sign and pneumothorax, which has the attenuation of -73 to 133 Hounsfield Unit (HU).

At this point, the impression was right hemopneumothorax with intralobar hematoma, possible due to chest tube insertion. The chest tube was removed the day after it was not draining. The patient improved clinically with the reduction in oxygen requirement to room air. The patient was then transferred to the cardiothoracic team. In view that chest tube reinsertion there is not much output, contrast-enhanced CT thorax (Figure 2 and 3) was done on day 23 of admission.

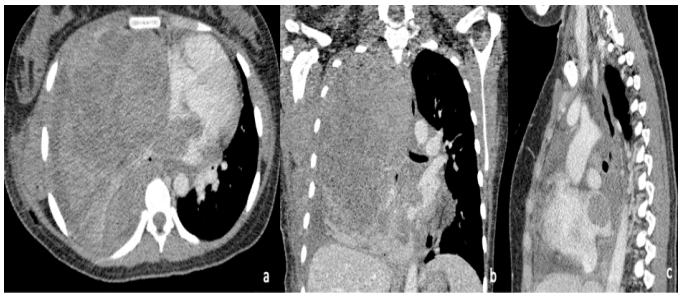

Figure 2: above shows axial (a), coronal (b), sagittal (c) of contrast-enhanced CT thorax. These images showed lobulated heterogenous hypodensity occupying the entrance of the right superior pulmonary vein, extending to the left atrium.

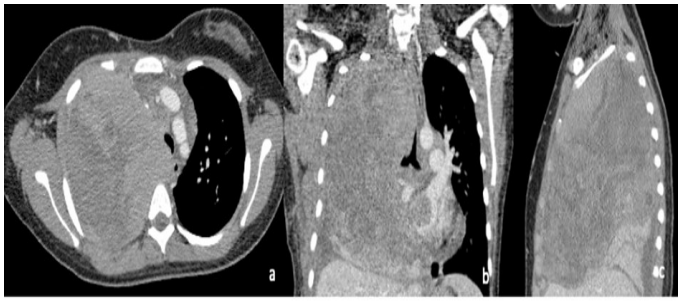

Figure 3 above shows axial (a), coronal(b), and sagittal (c) contrast-enhanced CT thorax images. These images showed minimal hemo- and pneumothorax at the right hemithorax. A large heterogeneous lobulated hypodense solid lesion is seen at the right upper and middle lobe, causing mass effect to the right lower lobe, trachea, right main bronchus, and pushes the heart to the left side.

At this point,our diagnosis was right lung hematoma was expanding with mass effect, and mass within the right superior pulmonary vein, extending to the left atrium and left appendages. This also raised the possibility of lung mass obscured by hematoma. Subsequently, the patient underwent right pneumonectomy and evacuation of left atrium mass. Histopathology report of the right lung mass and left atrium demonstrates high-grade small round cell tumor, suggestive of ES. However, parents refused chemotherapy, were then under palliative follow-up. Contrasted CT thorax, abdomen, and pelvis (TAP) shows no evidence of distant metastasis.

DISCUSSION

EES is a rare entity reported. It occurs in older age group patients, and usually at the trunk rather than limbs. Extremity involvement took up 32%, paravertebral including epidural 15%, cervical 11%, and chest wall 11%. [4] The primary EES of the lung is even rarer. In 2014, Hwang et al. reported that only 12 cases were found in the literature.[5] However, in 2015, Doekar et al. reported there were 16 cases reported thus far.[6] Pulmonary EES was reported that with its mean age onset around 20 years, as compared to 15 years in osseous ES. It is more common in males compared to females.[7] Although our patient falls in this age category, the patient is a female.

To diagnose as EES, a few criteria should be met, such as (a) no osseous involvement in Magnetic Resonance Imaging (MRI), (b) no increased uptake of bone or periosteum adjacent to tumor in static images of bone scintigraphy, (c) lesion consists of small round blue cells with no differentiation features under light microscopy, (d) demonstration of cytoplasmic glycogen.[1] Our patient has histological findings consistent with EES. However, MRI or scintigraphy was not performed for this patient. Imaging features of EES are not specific. Radiographic features may show soft tissue mass in 50% of the cases, adjacent bony involvement (25 to 42% of the cases), and calcification (25% of the cases). Angiography may show hypervascularity. Bone scintigraphy and FDG PET may show an increase in radionuclide uptake. FDG PET can be used to monitor metastases and recurrence.[1]

Pulmonary EES shows a solitary mass that is mostly well-defined in computerized tomography. It usually demonstrates hypo-enhancement with heterogeneous appearance and regions of low density, which may suggest necrosis. Intralesional calcifications and ipsilateral pleural effusion may be seen as in our patient. Adjacent invasion to the mediastinum or chest wall is infrequent.[7] In our case patient, EES was a rather ill-defined heterogeneous lobulated hypodense solid lesion with extension into the right superior pulmonary vein and left atrium.

It is associated with ipsilateral pleural effusion. CT TAP has shown no distant metastasis. Primary lung malignancy in teenagers is rare, and congenital lung tumours and metastatic disease are more common. Differential diagnoses include adenocarcinoma of the lung and NUT midline carcinoma. Adenocarcinoma presented with a median age of 15 years old. Its imaging features range from focal ground-glass nodules to mass-like consolidation. It is associated with invasion of the pleura, chest wall, or diaphragm. NUT midline carcinoma presents with a median age of 22 years. It manifests as a central or lower lobe large heterogeneously enhancing mass that invades the adjacent structures like the mediastinum, heart, airway, and oesophagus. Pleural effusion and lymphadenopathy are also common.[8] There are cases of pulmonary EES reported,[3,9,10] with age ranging from 19 to 36 years old, with CT showing unilateral lung mass, and histo-pathologically proven to be EES.

Multimodality had shown one case with no bony involvement via CT, bone scan, and bone marrow aspiration,[3] two cases showed no distant metastasis in metastatic workup,[9] one case has shown no metastatic lymph node via tissue biopsy.[10] Histologically, the tumor consists of a proliferation of small round cells with scanty and clear cytoplasm, round to oval nuclei, finely granular chromatin, and inconspicuous nucleoli. It also has periodic acid Schiff positive due to the presence of cytoplasmic glycogen and strong reactivity to CD 99/MIC-2 and vimentin.[6] In our patient, the histological findings was showing a high-grade small round cell tumor with the presence of glycogen demonstrating Periodic acid Schiff positive. This is also supported by moderate to a strong reaction to CD99 and patchy strong positivity to vimentin. These morphology, special stain, and immunohistochemical findings are consistent with ES.

CONCLUSION

Primary pulmonary EES is a rare entity. However, it should be considered in young patients who presented with large lung mass with no other evidence of extrathoracic involvement.

REFERENCES

1. Murphey MD, Senchak LT, Mambalam PK, Logie CI, Klassen-Fischer MK, Kransdorf MJ (2013) From the Radiologic Pathology Archives: Ewing Sarcoma Family of Tumors: Radiologic-Pathologic Correlation. https://doi.org/101148/rg333135005 33:803–831
2. Asker S, Sayir F, Bulut G, Sunnetcioglu A, Ekin S, Yavuz A (2015) Primitive neuroectodermal tumor/ ewing sarcoma presenting with pulmonary nodular lesions. Case Rep Oncol Med. https:// doi.org/10.1155/2015/957239
3. S A, AR M, G P (2020) Primary Ewing's sarcoma of the lung: a rare case report. Indian J Thorac Cardiovasc Surg 36:409–411
4. Patnaik S, Yarlagadda J, Susarla R (2018) Imaging features of Ewing's sarcoma: Special reference to uncommon features and rare sites of presentation. J Cancer Res Ther 14:1014–1022
5. Kim DK, Hwang SK, Park S-I, Kim Y-H, Kim R (2014) Primary Ewing's Sarcoma of the Lung. Korean J Thorac Cardiovasc Surg 47:47–50
6. KK D, NG K, S G (2015) Primary ewings sarcoma of the lung. J Clin Diagn Res 9:XD01–XD03
7. Shet N, Stanescu L, Deutsch G (2013) Primary extraosseous Ewing sarcoma of the lung: Case report and literature review. Radiol Case Reports 8:832
8. III JPL, Biko DM, Carter BW, Pavio MA, Huppmann AR, Chung EM (2018) Primary Lung Tumors in Children: Radiologic-Pathologic Correlation From the Radiologic Pathology Archives. https:// doi.org/101148/rg2018180192 38:2151–2172
9. Sobh E, El-Sheshtawy WH, Anis SE (2017) Primary pulmonary extraskeletal Ewing sarcoma/Primitive neuroectodermal tumor: Two case reports. Egypt J Bronchol 2017 112 11:161–164
10. S E, U Ç, C G, R E, H Y (2019) Ewing's sarcoma of the lung: A rare case. Tuberk Toraks 67:146–148