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## Ewing's sarcoma of the lung: A rare case

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### SUMMARY

#### Ewing's sarcoma of the lung: a rare case

The Ewing's sarcoma family of tumors (ESFT) incorporates both the well-recognized primary bone and the extraskeletal soft tissue sarcomas. Primitive neuroectodermal tumors (PNET) and ESFT have a similar neural phenotype and can be considered in the same entity. Here, we will present 28 years old patient with Ewing Sarcoma. Patient was admitted chest pain. Chest radiograph showed a suspicious lesion in the left paracardiac area. Computed tomography (CT) scan and positron emission tomography (PET)/CT result were compatible with malignant tumor. The patient was underwent surgical resection as the bronchoscopic result couldn't a malignant finding and pathological finding was detected as Ewing's sarcoma. Ewing's sarcoma should be considered in patients who are very fast growing in the lungs, are properly confined and suspected of malignancy in FDG-PET/CT.

**Key words:** Ewing's sarcoma; pulmonary

### ÖZET

#### Akciğerin ewing sarkomu: Nadir bir olgu

Ewing'in sarkom tümör ailesi (ESFT) hem iyi tanınan primer kemik hem de ekstra iskelet yumuşak doku sarkomlarını içerir. İlkel nöroektodermal tümörler (PNET) ve ESFT'de benzer bir nöral fenotip vardır. Yirmi sekiz yaşında, erkek hasta, göğüs ağrısı şikayetiyle başvurdu. Akciğer grafisinde sol parakardiyak bölgede şüpheli bir lezyon izlendi. Ewing sarkomu akciğerlerde çok hızlı büyüyen, FDG-Pozitron emisyon tomografi (PET)/bilgisayarlı tomografi (BT)'de düzgün şekilde ve SUV<sub>max</sub> değeri yüksek olan hastalarda maligniteden şüphelenilmeli erken tedavi uygulanmalıdır.

**Anahtar kelimeler:** Ewing sarkom; akciğer

## INTRODUCTION

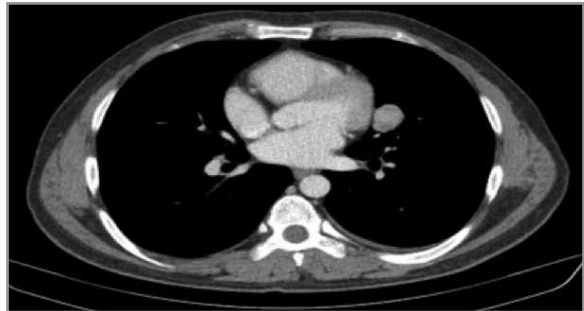
The Ewing's sarcoma family of tumors (ESFT) incorporates both the well-recognized primary bone and the extraskeletal soft tissue sarcomas. Primitive neuroectodermal tumors (PNET) and ESFT have a similar neural phenotype and can, therefore, be considered the same entity. First described in 1921 by American pathologist James Ewing, Ewing's sarcomas are neuroectodermal tumors characterized by monotonous small round cells that are arranged in sheets on histological examination. Although Ewing's sarcoma presenting with a lytic bone lesion is, in itself, uncommon, it is recognized as the second most prevalent primary bone tumor worldwide. Cases of primary pulmonary ESFT have been reported in recent years, but there are still only a few reports in the literature. We herein discuss both the clinical course and the therapeutic management of this disease and the associated family history (1). Patients with Ewing Sarcoma were at an increased risk in comparison to other patients and ranked third after survivors of retinoblastoma and primitive neuroectodermal tumors of the central nervous system (2). Over the last four decades, the survival has been improved from 10% with radiotherapy alone to near 70%, in localized disease, with the introduction of chemotherapy and multimodality approach. The multidisciplinary approach with risk adapted chemotherapy and local treatment (surgery and/or radiotherapy) has been used in recent times in many collaborative trials to minimize the over treatment and thus treatment related side effects with maintaining high cure rates (3).

## CASE REPORT

In September 2018, 28 year old a male patient was admitted to the hospital because of left chest pain and shoulder pain. No pathological findings were found on physical examination. A direct chest radiograph showed a suspicious lesion in the left paracardiac area (Figure 1). Therefore, Thorax computed tomography (CT) was performed on the patient. The CT of the patient revealed a well-limited nodular lesion with a soft tissue density of approximately 28 x 21 mm adjacent to the heart in the lingula inferior segment of the lung (Figure 2). Bronchoscopy was planned and bronchoscopy revealed no endobronchial lesion. Bronchoscopic lavage material was reported as benign. 18-FDG PET/CT was performed because the lesion was enlarged during chest X-Ray. 18-FDG PET/CT showed pathological increased 18F-FDG up to 1 cm in the left hemithorax. In the left lung lingula and apicoposterior segment was observed that 7 x 3 cm in diameter and it was not clearly differentiated with pericardium (Figure 3). Surgical intervention was planned upon rapid growth of the mass of the patient and acceptance as operative. During the opera-



**Figure 1.** Suspicious lesion in the left paracardiac area.



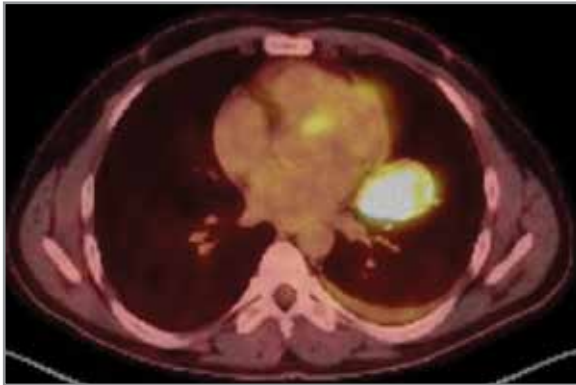
**Figure 2.** A well-circumscribed nodular lesion with a soft tissue density of approximately 28 x 21 mm, adjacent to the heart, in the inferior lingula of lung.

tion, frozen biopsy was studied and the result was reported in favor of malignancy. Therefore, the mass was removed from the pericardium and left lung upper lobectomy was performed. All material was sent to pathology. There was no metastatic lymph nodes in the pathology material. Initially, a small blue round cell high grade malignant tumor compatible with PNET/Ewing sarcoma and metastatic lymph nodes showed no metastasis. Vimentin (diffusely +), PanCK (focal +), CD56 (+), NSE (focal +), CD99 (focal +), WT1 (focal +), PAS (focal +), CD3 (-), CD20 (-), PAX5 (-), Synaptophysin (-), KromagraninA (-), TTF1 (-), Desmin (-), TdT (-), CK20 (-), S100 (-), Myoglobin (-), HMB45 (-) and the Ki67 proliferation index was reported to be 70% (Figure 4). The patient's general condition was good and control chest radiography was performed (Figure 5). Chemotherapy and radiotherapy were planned and the patient was discharged.

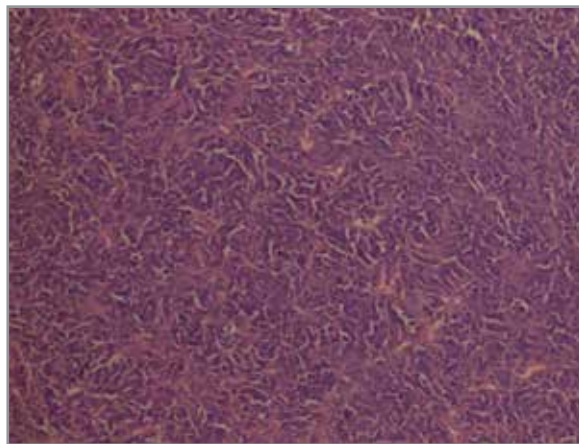
## DISCUSSION

Ewing sarcoma is a very rare malignant disease and generally occurs as a skeletal disease of the pediatric age group. Ewing sarcoma is much more rare in the adult age group (4). Extrasosseous Ewing's sarcoma is even more uncommon in adults: only approximately 16% of all Ewing's sarcomas are extrasosseous, and

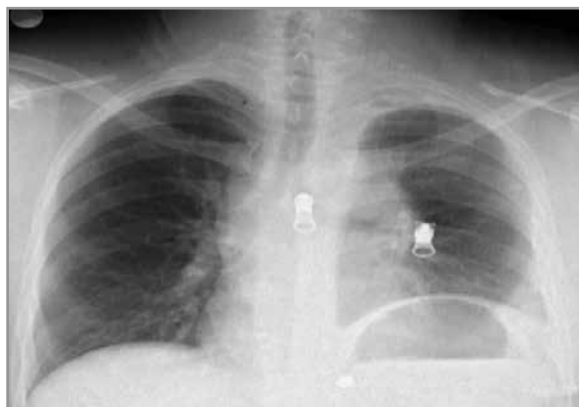
therefore, less than 1% of all such tumors are extraosseous tumors occurring in adults. Only 12 cases of primary pulmonary Ewing's sarcoma have been reported in the literature (5). In many studies, Ewing sarcoma and other sarcomas are performed by FDG-PET/CT (6). Since the radiological findings showed a rapidly growing lung lesion, PET/CT was performed. A lung lesion with dense FDG involvement was detected in PET/CT.



**Figure 3.** Properly limited mass with pathologically elevated 18F-FDG uptake.



**Figure 4.** Pathological preparation of the lesion.



**Figure 5.** Patient's post-operative control PA Chest X-Ray.

The pathological diagnosis of Ewing sarcoma may sometimes be incorrect. Because all of the pathological findings seen may not be specific to Ewing sarcoma. Therefore, Ewing sarcoma should be differentiated from small cell carcinoma, adenocarcinoma, squamous cell carcinoma and lymphoma. Recently, CD99, which has been used for the identification of Ewing's sarcoma, increased the correct diagnosis rate (4). CD99 was found to be focal in our case. Extraosseous Ewing's sarcoma is a curable disease. The disease-free survival rate has been significantly increased by managing these tumors with aggressive surgical resection in combination with multi-agent chemotherapy, with or without radiotherapy (5). Since CT findings, FDG-PET/CT findings were curable, lobectomy was performed. We planned to present our case because it is very rare and suitable for surgery. Ewing's sarcoma should be considered in patients who are very fast growing in the lungs, are properly confined and suspected of malignancy in FDG-PET/CT. The patient should be diagnosed very early and if it is curable, surgical treatment should be performed immediately.

#### CONFLICT of INTEREST

The authors reported no conflict of interest related to this article.

#### AUTHORSHIP CONTRIBUTIONS

Concept/Design: SE, CG

Analysis/Interpretation: SE, UÇ

Data Acquisition: All of authors.

Writing: SE

Critical Revision: SE, HY

Final Approval: All of authors.

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