CASE REPORT

Massive pleural effusion in an 18-year-old girl with Ewing sarcoma

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Ewing sarcoma is a bone tumour that commonly appears between ages five and 10 in the diaphysis of the long bones and predominantly presents with pain and swelling. The case of an 18-year-old girl who presented with back pain, cough, dyspnea, weakness and fever is described. Chest radiography showed a homogenous density in the middle and inferior zones of the right hemithorax. Thoracic computed tomography revealed a diffuse pleural effusion and a 6.99 cm×4.45 cm solid mass composed of lobulated, small cystic lesions and calcifications in the right hemithorax. Biochemical analysis of pleural fluid showed hemorrhagic effusion and exudate. A pleural needle biopsy demonstrated solid uniform tumour cells with narrowed cytoplasm, round nuclei and uncertain nucleoli. All of the tumour cell cytoplasmstained with CD99. The pathological examination supported Ewing sarcoma. Three-phase Tc-99m methylene diphosphonate scintigraphy of the whole body showed pathological tracer uptake in a broad area of the eighth costal bone and in smaller areas of the ninth and 10th costal bones. This case is reported because Ewing sarcoma is a rare cause of pleural effusion in clinical practice among younger adults.

Key Words: Adolescent; Ewing sarcoma; Pleural effusion

CASE PRESENTATION

An 18-year-old girl was referred with back pain for three months, cough, dyspnea and weakness for 10 days, and fever for three days. Her medical history had no relevant data. Family history was significant only for asthma in her mother. Physical examination revealed diminished breath sounds in the middle and inferior posterior right hemithorax, and dullness to percussion over the corresponding areas. The physical examination was otherwise unremarkable. Laboratory examination revealed moderate anemia (hemoglobin 85 g/L, hematocrit level 25%) and a high erythrocyte sedimentation rate of 8.77 µmol/L to 27.02 µmol/L and blood iron binding capacity 43.73 µmol/L (normal range 44.75 µmol/L to 76.07 µmol/L). An erect frontal chest radiograph demonstrated a homogeneous density in the middle and inferior right hemithorax (Figure 1).
There was blunting of the right costophrenic angle, displacement of the cardiac shadow and opacification to the level of the second rib. This was in keeping with a large right-sided pleural effusion. Thoracic computed tomography examination revealed diffuse right-sided pleural effusion, a solid mass composed of lobulated, small cystic lesions, and calcifications in the right hemithorax (diameter of 6.99 cm×4.45 cm) (Figure 2). In addition, there were erosive changes in the adjacent ribs and atelectasis in the right lung. Spine radiographs did not show any pathological changes. Biochemical analysis of pleural fluid indicated an exudate (Table 1). The cytology of pleural fluid demonstrated abundant red blood cells, mesothelial cells and histiocytes. No malignant cells were observed. Pleural biopsy, with an Abrahams needle, demonstrated a solid uniform tumour, with the cells having narrowed cytoplasms, round nuclei and uncertain nucleoli. Necrosis or mitosis was not determined and rosette formation was not observed. The tumour cell cytoplasm stained uniformly with CD99, which is a MIC2 product (a specific surface membrane protein highly sensitive but not necessarily specific for Ewing sarcoma). The tumour tissue did not stain with keratine (CK 7), which is highly positive for synovial sarcoma. After the two procedures described above and other immunohistochemical procedures, the patient was diagnosed with Ewing sarcoma. Three-phase Tc-99m methylene diphosphonate scintigraphy of the whole body showed pathological tracer uptake in a broad area of the right eighth costal bone and in smaller areas of the ninth and 10th costal bones.

DISCUSSION
Localized pain or local swelling is typically the first symptom reported by pediatric patients with bone tumours. Fever is present in 28% of patients with Ewing sarcoma at the time of diagnosis (3). Metastases are present in approximately 26% of patients at initial diagnosis. The most frequent sites of metastases are the lungs and other bones. Multiple pulmonary metastases may produce respiratory insufficiency, or paraplegia may develop secondary to a vertebral body metastasis (6,7).

Massive pleural effusion is an unusual finding in young persons. Because of the high prevalence of tuberculosis in Turkey, most of the pleural effusions in the younger age groups are related to tuberculosis. Generally, hemorrhagic effusion or exudates are diagnosed as tuberculosis without a histopathological examination. Thoracentesis, and radiological and histopathological examinations are usually required to distinguish the etiology (8). In the present patient, a tumour of the ribs with
The involvement of soft tissues could have produced the pleural effusion. In our opinion, it is more likely that hemorrhagic effusion was secondary to diffuse pleural metastasis because the specimens from pleural surface and the solid tumor suggested the findings of Ewing sarcoma.

It is known that metastatic disease, tumor size (greater than 8 mm in diameter) and high serum lactate dehydrogenase levels are poor prognostic factors for Ewing sarcoma. On the other hand, an indicator of prognosis is a radiographical and histological response to initial treatment. According to the literature, after the six-drug regimen treatment of the present patient, the expected five-year, event-free survival would be 68% (9).

A case reported by Wolf et al (10) had similar characteristics to the present patient, except for the size of the tumor and pain localization. As Ewing sarcoma is a bone tumor of childhood (commonly develops between the ages of five and 10), our patient was older than usual, as was the case reported by Wolf et al (10). A massive pleural effusion with dyspnea as the first sign of Ewing sarcoma is the second important peculiarity of the present case.

### REFERENCES


### TABLE 1

The results of chemical analysis of synchronous blood and pleural fluid

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Blood</th>
<th>Pleural fluid</th>
</tr>
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<tbody>
<tr>
<td>LDH (U/L)</td>
<td>218</td>
<td>450</td>
</tr>
<tr>
<td>Total protein (g/L)</td>
<td>76</td>
<td>75</td>
</tr>
<tr>
<td>Albumin (g/L)</td>
<td>38</td>
<td>31</td>
</tr>
<tr>
<td>Glucose (µmol/L)</td>
<td>5.55</td>
<td>4.66</td>
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</tbody>
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LDH Lactate dehydrogenase