Giant cell tumors (GCT) are rare, benign but locally aggressive tumors of bones. They account for about 5% of all primary bone tumors and approximately 20% of benign bone tumors. They usually occur between the 2nd and 4th decades of life.\(^1,2\) GCTs mainly occur in the meta-epiphyseal regions of long bones, and the most common site is the distal end of the femur. Ribs are the least frequently affected site, seen in less than 1% of all cases and most of them occur at the posterior arc.\(^3,4\)

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GCTs mainly occur in the meta-epiphyseal regions of long bones, and the most common site is the distal end of the femur. Ribs are the least frequently affected site, seen in less than 1% of all cases of GCT.\(^3,4\) Most of them arise from the posterior arch of the rib, and the anterior arch of origin is also extremely rare in adults. The typical histologic feature of GCTs is proliferation of multinucleated giant cells resembling osteoclasts with a stroma of spindle-shaped mononuclear cells.\(^5\) The tumor has to be differentiated from other lesions containing benign giant cells, such as aneurysmal bone cyst (ABC) or brown tumors of hyperparathyroidism.\(^2\) Detailed search of old and recent series and cases of GCT or rib lesions revealed that there are a few pediatric cases with GCT in the rib and no children had the involvement of the anterior arc (Table I).\(^5,12\) Therefore, we aimed to present our case with unusual GCT location to include GCT in the list of differential diagnoses of chest wall tumors and to emphasize the necessity for \textit{en-bloc} resection and close follow up in children.

Case Report

An otherwise healthy 12-year-old girl presented with swelling and slight pain on the left inferior-anterior chest wall for two years. She had no respiratory symptoms. Past medical history was unremarkable.
Physical examination revealed a giant, hard and fixed mass on the left chest wall. The overlying skin was normal. Hematological and biochemical test results were in normal limits except slight elevation of alkaline phosphatase (ALP) level (359 U/L, normal range; 51-332 U/L). Parathyroid hormone (PTH) level was also normal (46.8 pg/ml, normal range; 12-88 pg/ml). Plain chest X-ray revealed a radiopaque area in the left lower hemithorax (Fig. 1). Computed tomography (CT) of the chest showed a large expansive mass and lytic lesion with internal calcification arising from the anterior part of the 7th rib and atelectasis of the left lower lobe (Fig. 2a, b). Dimensions of the mass were 7×10×10 cm. The differential diagnosis included ABC, primitive neuro-ectodermal tumor (PNET), and less likely chondroid matrix neoplasms; but was not specified on the CT. Tru-cut biopsy was performed but it was not possible to differentiate GCT and ABC.

The patient was operated via a left thoracotomy incision at the 6th intercostal space. En-bloc resection was performed including the 6th-8th ribs and a small part of the diaphragm, which was attached to the tumor. Chest wall reconstruction was performed with expanded-polytetrafluoroethylene (ePTFE) dual-surface (Dual Mesh, W.L. Gore & Associates, Flagstaff, AZ).

The diameter of the tumor (as measured from the gross surgical specimen) was 11×10×6.5 cm and three ribs were 9 cm in length. Final pathological evaluation revealed GCT of bone and the surgical margin was clear (Fig. 3). Serum ALP levels decreased to 230 U/L and 176 U/L at 2nd and 11th months respectively following

<table>
<thead>
<tr>
<th>Case no</th>
<th>Author, year &amp; Sex</th>
<th>Age (year) &amp; Sex</th>
<th>Sign &amp; Symptoms</th>
<th>Side and location of GCT</th>
<th>Size (cm)</th>
<th>Initial approach</th>
<th>Surgery</th>
<th>Pathology</th>
<th>Outcome</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>Locher GW, et al. (3), 1975</td>
<td>14, M</td>
<td>Pain</td>
<td>R, 6th rib, lateral arc</td>
<td>NA</td>
<td>Resection</td>
<td>En-bloc resection &amp; bone graft</td>
<td>GCT - ABC</td>
<td>NED, 3 years</td>
</tr>
<tr>
<td>2</td>
<td>Locher GW, et al. (3), 1975</td>
<td>13.5, M</td>
<td>Pain, mass</td>
<td>L, 8th rib, posterior arc</td>
<td>NA</td>
<td>Resection</td>
<td>En-bloc resection</td>
<td>GCT - ABC</td>
<td>NED, 5 years, scoliosis</td>
</tr>
<tr>
<td>3</td>
<td>Locher GW, et al. (3), 1975</td>
<td>4.5, F</td>
<td>No</td>
<td>R, 4th rib, posterior arc</td>
<td>NA</td>
<td>Resection</td>
<td>En-bloc resection*</td>
<td>GCT - ABC</td>
<td>NED, 1 year</td>
</tr>
<tr>
<td>4</td>
<td>Schütte HE, et al. (4), 1993</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>GCT</td>
<td>NA</td>
</tr>
<tr>
<td>5</td>
<td>Athanassiadou F, et al. (7), 2003</td>
<td>12, F</td>
<td>Mass</td>
<td>R, lower rib, posterior arc</td>
<td>8x4x4</td>
<td>Needle biopsy</td>
<td>En-bloc resection</td>
<td>GCT</td>
<td>NED, 1 year</td>
</tr>
<tr>
<td>6</td>
<td>Özyüksel G, et al. (present case), 2020</td>
<td>12, F</td>
<td>Pain, mass</td>
<td>L, 7th rib, anterior arc</td>
<td>7x10x10</td>
<td>Tru-cut biopsy</td>
<td>En-bloc resection &amp; chest wall reconstruction</td>
<td>GCT</td>
<td>NED, 14 months</td>
</tr>
</tbody>
</table>

M: male, F: female, NA: not available, R: right; L: left; *En-bloc resection for recurrent tumor previously underwent incomplete resection another center, GCT: giant cell tumor, ABC: aneurysmal bone cyst, NED: no evidence of disease

Fig. 1. Chest X-ray demonstrates destructive rib lesion with soft tissue mass (arrow).
surgery, respectively. She had no symptoms of recurrence or metastasis at the one-year follow-up. Written informed consent was obtained from the patient for publication of this case report and accompanying images.

**Discussion**

We presented an extremely rare site of GCT in childhood. GCT is a rare tumor that usually affects the bones around the knee joint. GCT originating from the ribs is an extremely rare condition and usually arise from posterior ribs. In this sense, the presented case is unique for its location in anterior arc. Giant cell tumor of bone represents around 4-5% of all primary bone tumors and approximately 20% of benign primary bone tumors. It is seldom seen in skeletally immature individuals. In the pediatric population, GCT of bone is seen in 2-5% of all reported cases. The most common area of the tumor is proximal tibia, followed by vertebrae and pelvis. A detailed review of case reports, clinical and surgical series revealed that GCT of the rib has been encountered in only six pediatric cases including ours and there is no pediatric case with the GCT arising from the anterior arc of the rib.

It is known that GCT of bone has a slightly higher female-to-male incidence in all age groups. According to novel pediatric reports, female predominance may be seen in the GCT of bone. Of note, our patient was female as well.

Serum ALP may be elevated at the time of diagnosis and normalize after surgery. ALP is not a specific marker for GCT of bone, as some stromal cells express ALP. However, normal level of ALP rules out brown tumor of hyperparathyroidism. Our patient had slightly
Giant Cell Tumor Arising from the Anterior Arc of the Rib

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18 Elevated serum ALP level in the preoperative period, and decreased to normal in the postoperative period. Additionally, PTH level was in normal limits in the current case. Serum acid phosphatase (AcP) level has also been suggested as a useful marker for GCT of bone, because a correlation was revealed previously between the tumor volume and serum AcP level. The serum AcP was not examined in the present case.

Radiologically, the tumors generally appear as expansive osteolytic defects on X-rays, eventually leading to significant local bone destruction. A cross-sectional imaging provides a better evaluation of cortical destruction, calcification, penetration, and metastases. While CT is the best choice to describe the grade of cortical destruction, magnetic resonance imaging (MRI) may be useful to evaluate the invasion of soft tissues. Furthermore, MRI can detect hemosiderin deposition seen in GCT of bone.

In a child with a chest wall mass, histopathological diagnosis should be performed, besides physical and radiological examination findings, to exclude a more likely malignancy. Therefore, tru-cut biopsy is useful in preoperative diagnosis. Similarly, in the current case, malignancy was ruled out by the tru-cut biopsy, but the differential diagnosis between GCT of bone and ABC was not possible due to the limited amount of tissue.

The first choice of treatment is surgery in GCT of the bone. There are two options: en-bloc excision with reconstruction or extended intralesional curettage followed by filling the defect with bone cement or graft. Wide excision to ensure clear margins is recommended because of 25-50% of local recurrence rates following intralesional curettage. Although no statistical significance between local recurrence rates following wide resection or intralesional curettage has been reported for GCT in pediatric case series, it is remarkable that patients who undergo wide resection may have a greater chance in avoiding local recurrence. En-bloc resection of GCT should be preferred in every patient unless surgery causes impairment of joint function, mobilization or esthetics. We preferred en-bloc excision to eradicate all neoplastic tissue with clear margins in our patient.

Histologically, GCT of bone is characterized by a large number of multinucleated giant cells as well as macrophage-like and stromal cell-like mononuclear cells. It may contain areas of cystic degeneration, hemorrhage, hemosiderin deposition, occasional mitotic figures or increased spindle cell stroma. It is important to make the differential diagnosis of other lesions containing giant cells, such as ABC, fibrous metaphyseal defects, chondroblastoma, brown tumor in hyperparathyroidism and giant cell-rich variants of osteosarcoma. In addition, GCT of bone and ABC may be seen together in the same case since GCT of bone is the most common precursor lesion of ABC.

Malignant transformation occurs in less than 1% of GCT. Furthermore, radiotherapy should be avoided in these patients which may lead to malignant transformation. Metastases on GCT occur to the lungs with an incidence of 2-6% of patients, in 3-4 years after the primary diagnosis. They have usually a benign character and some of them regress spontaneously. A small group of these metastases is progressive and causes mortality. According to a wide study on 2315 patients with GCT of bone, 5-year survival rate for malignant GCT of bone was 87%, with a total mortality rate of 16%.

In conclusion, we present a unique case of GCT arising from the anterior arc of the rib in a 12-year-old adolescent girl. As such, we propose that differential diagnoses of chest wall tumors in childhood should comprise GCT as well. Due to its recurrence and malignant potential, complete wide resection with chest wall reconstruction must be the first choice of treatment for this tumor. Lastly, it is crucial to follow up these patients postoperatively for recurrent or metastatic disease.
Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

The authors confirm contribution to the paper as follows: study conception and design: İK, GÖ; data collection: GÖ, BA; analysis and interpretation of results: GÖ, BA; draft manuscript preparation: İK, GÖ, BA, HNÖ, GG, AV. All authors reviewed the results and approved the final version of the manuscript.

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Conflict of interest

The authors declare that there is no conflict of interest.

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