Case Report

Chondrosarcoma from chondroblastoma in young: A rare event

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ABSTRACT

Among bone tumors, chondrosarcoma (CS) is the second most common bone tumor affecting adults in their fifth decade of life. Although the appendicular skeleton is the site of occurrence, chest wall involvement has also been reported. They can be primary if it develops *de novo* or secondary if originates from a pre-existing benign lesion. Osteochondroma, enchondroma, synovial chondromatosis, and Paget's disease are the common precursor lesions. A 24-year-old male with a history of chest wall tumor for which excision was done 11 years back presented with a recurrence at the same site. A past biopsy reported it to be chondroblastoma. Radiological investigations revealed the recurrence likely to be a CS. The patient underwent left thoracotomy and excision of the mass along with a segment of the sixth and seventh ribs from where the mass was arising. The biopsy reported well-differentiated CS. Since it was low-grade and most were resistant to chemotherapy and radiotherapy, no adjuvant therapy was given. Although uncommon for age and site, CS should be suspected even if the earlier excision biopsy was a benign cartilaginous tumor. Complete excision is the best treatment.

Key words: Chest wall tumor, Chondroblastoma, Chondrosarcoma

Chondrosarcoma (CS) is the second most common malignant bone tumor [1], accounting for almost 27% of bone cancers reported in the International Agency for Research on Cancer database [2]. CS primarily affects adults, with over 70% of cases presenting after 40 years of age. During the first two decades, CS forms only 3% of bone cancers [3-5]. CS most commonly involves the appendicular skeleton and pelvis, and the chest wall is involved rarely [6]. CS forms the most common malignant neoplasm of the chest wall although the overall incidence is very low [7]. We could not find any case of CS developing from a chondroblastoma (PubMed, Scopus, Google Scholar, and Lancet; from 1990 to July 2024). Here, we report the case of secondary CS of the chest wall arising from a chondroblastoma in a young patient is presented.

CASE REPORT

A 24-year-old male presented with complaints of swelling on the left chest wall for the last 10 months, which progressed from 1 cm × 1 cm to 2 cm × 2 cm over the same period.

On examination, there was a 2 × 2 cm hard, fixed, non-tender swelling of the left lateral chest wall at the level of the 6th rib. The patient had a history of a similar swelling in the same region 11 years back which was excised in another institution and was reported as chondroblastoma. The rest systemic examination was normal except for slightly deviated cardiac apex medially and decreased breath sounds in the left lower zone of the chest. No palpable lymphadenopathy was observed.

Chest X-ray (Fig. 1) showed a radio-opacity in the left lateral chest with lower lobe collapse. Computed tomography (CT) chest confirmed a large heterogenous hypodense lesion measuring 11.2 × 10 × 13 cm in the left hemithorax with coarse calcification within it. There was erosion of the left 6th rib and also the likely origin of the tumor. The left lung was compressed by the tumor (Fig. 2a and b). Magnetic resonance imaging (MRI) chest showed a relatively well-defined solid mass measuring 10.8 × 9 × 13.7 cm with lobulated margins, extending from D5 to D10 vertebral levels, isointense on T1-weighted image, and heterogeneously hyperintense on T2-weighted/short-tau inversion recovery images. On the post-contrast sequence, the lesion had peripherally enhancing walls with multiple enhancing foci within, showing ring and arc configuration, suggestive of CS (Fig. 2c and d). An image-guided core needle biopsy was inconclusive.

The patient underwent left thoracotomy and excision of the mass along with a segment of the 6th and 7th ribs from where the mass was arising. Since the mass was large and could not be removed through the thoracotomy incision in one piece, it was divided into smaller chunks and removed. Gross removal of all

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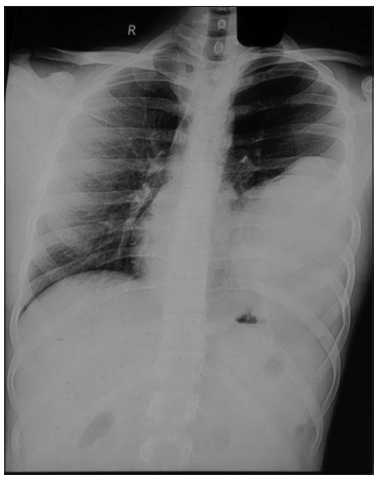

Figure 1: Radio-opacity in left lower zone with expansion and lucency of 6th rib

tumors was achieved. The resultant defect in the chest wall was closed with the interposition of a composite mesh (proceed) and sutured with surrounding ribs. The chest tube was removed after 2 days, after confirming full expansion on X-ray.

The final biopsy (Fig. 3) reported hypercellular sheets of chondrocytes with a lobular growth pattern. Chondrocytes had plump hyperchromatic nuclei with hyperchromatic nucleoli with features suggestive of well-differentiated CS. The patient was referred to a dedicated sarcoma clinic. Since it is a low-grade sarcoma, no adjuvant therapy was planned. The patient followed up at monthly intervals. The last visit was 3 months after surgery and was doing well.

DISCUSSION

CS is a malignant cartilaginous, matrix-producing neoplasm, primarily affecting adults. Although reported in children as well, secondary CS tends to be more common than primary.

Huvos and Marcove [6] reviewed 79 patients with CS under 21 years of age and found the appendicular skeleton to be the most common site of involvement, followed by the pelvis, with only one case arising from the chest wall. Although the chest wall forms a rare site for primary malignant tumor, CSs are the most common [7].

Based on origin, CS can be primary if it develops *de novo* or secondary if originates from a pre-existing benign lesion. Osteochondroma, enchondroma, synovial chondromatosis, and Paget's disease are the common precursor lesions for secondary CS [8]. Almost 80% of such lesions arise from osteochondromas [1]. Altay *et al*. [8] performed a retrospective analysis of 627 patients with cartilage-forming bone tumors, in which 32 patients had malignant transformations. None of the 32 patients with chondroblastoma developed malignancy.

The true incidence of secondary CS is difficult to report as many patients are asymptomatic with their benign lesions. The incidence of secondary CS reported is 0.4–2% and 1–5% in patients with solitary and multiple osteochondromas, respectively [9,10]. Secondary CS tends to occur in younger patients, with a mean

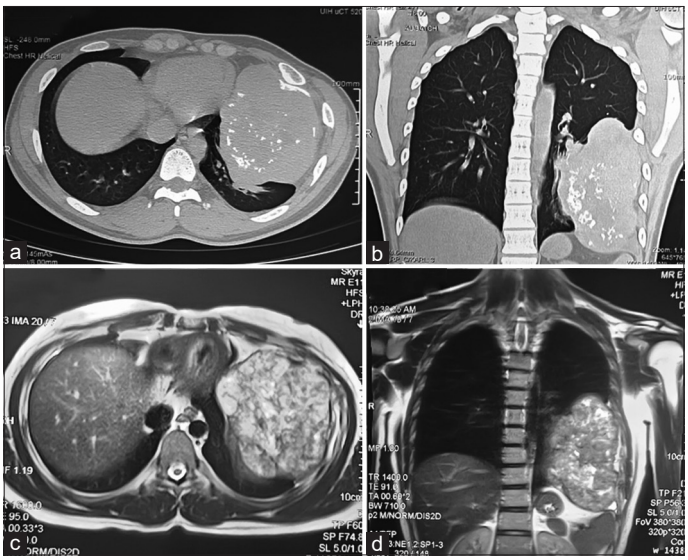

Figure 2: Cross-sectional imaging (a and b) is high-resolution computed tomography image showing well-defined large heterogenous hypodense lesions with coarse calcifications within eroding the cortex of the 6th rib; (c and d) is magnetic resonance imaging showing well-defined lobulated margins, hyperintense on T2-weighted/short-tau inversion recovery, with enhancing foci within (corresponds to calcifications on computed tomography)

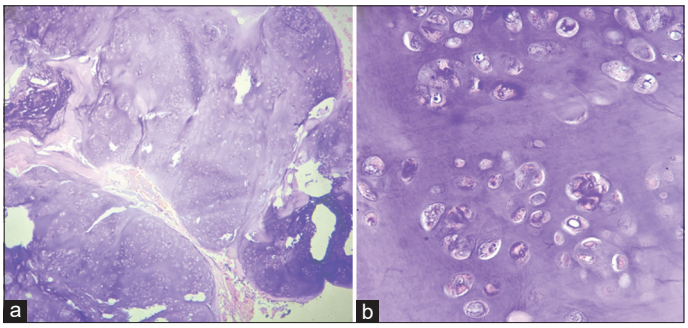

Figure 3: (a) H and E staining (×100) shows hypercellular sheets of chondrocytes with lobular growth pattern; (b) H and E staining (×200) shows bilobed chondrocytes with hyperchromatic nuclei

age of 35 years. In general, secondary CS tends to be low grade, and the difficulty in management is mostly due to a difficult location [9]. Any patient suspected to have CS should undergo a plain radiograph followed by cross-sectional imaging, either CT or MRI.

For any type of CS, primary or secondary, the gold standard of treatment is complete surgical excision [11,12]. It helps reduce the rate of local recurrences (LR) [13]. Most are resistant to chemotherapy and radiotherapy. Poor vascularity of the tumor and a low percentage of dividing cells are thought to be the cause of this resistance [11,14]. Treatment scenario changes with a change in the grade or location of the tumor, as more aggressive surgical resection is needed for more aggressive tumors. LR has not increased even with intralesional excision of grade I central CS [15]. A wide local excision is generally needed for grade II and III CS. Cartilaginous lesions of the pelvis and sacrum show a higher recurrence, so wide excision is recommended for any grade in such locations [16]. Although CS is relatively chemo-resistant, mesenchymal CS shows some chemo-sensitivity and can be

considered for adjuvant or neo-adjuvant chemotherapy. Radiation therapy is indicated only in cases of incomplete excision of high-grade or mesenchymal CS (as they are more radiosensitive than others) to maximize local control or in cases where resection is not feasible due to difficult anatomical location [17].

Following the initial treatment, regular follow-up is essential to look for any signs of LR. For low-grade CS, initial follow-up is 6 monthly for 2 years with an X-ray of the primary site with or without cross-sectional imaging, followed annually or as clinically indicated. For high-grade CS, more extensive follow-up with 3–6 monthly X-rays of the primary site with a plain chest X-ray with or without cross-sectional imaging for the first 5 years, annually thereafter, or as clinically indicated [18].

LR is the most important factor influencing prognosis. LR depends on various factors, and a high LR is associated with pelvic location, high grade, dedifferentiated subtype, and surgical margins (<1 mm for grade 1 and <4 mm for high grade) [19]. The lowest rate of LR is seen in peripheral CS (5–13%) and highest in dedifferentiated CS (42–55%). Grade 1 shows an LR of 12% whereas grades 2 and 3 show an LR of 26% and 37%, respectively. Most LR develops in the first 2 years following the surgery. Development of LR in the 1st year following surgery and age over 50 years are predictors of poor survival [20,21].

CONCLUSION

CS is an uncommon tumor of the chest wall and should be suspected even if the earlier excision biopsy was a benign cartilaginous tumor. Complete excision is the best treatment.

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