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Primary tumors of the ribs; experience with 78 patients

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Abstract

A retrospective study of primary tumors of the ribs (PTR) was conducted to review their clinical, radiological, and pathological features, as well as the early and long-term results of surgical management. Seventy-eight patients (48 male, 30 female, age range: 7–80 years) with PTR were treated in our clinic between January 1998 and July 2008. Forty-nine (63%) of the patients had benign lesions and 29 (37%) had malignant lesions. In the group with malignant tumors, the main symptom was pain, but in the group with benign tumors, the main symptom was swelling. Eight cases in the benign group and four cases in the malignant group were asymptomatic (accidental radiological findings in recruitment centers). Most of the patients with PTR had benign lesions. All PTR should be considered as malignant until proven otherwise. Surgery must consist of wide resection with tumor-free margins in order to provide the best chance for cure in both benign and malignant lesions.
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Keywords: Primary tumors; Ribs; Thoracic wall

1. Introduction

Primary tumors of ribs (PTR) are uncommon, comprising 5–10% of all bony tumors. In this study, 78 patients who were operated for a primary rib lesion were retrospectively evaluated, and their postoperative pathological features were classified. In the literature it is reported that malignant neoplasms are significantly more common than are benign ones [1]. However, Hsu and his friends reported that the ratio of malignant neoplasms to benign neoplasms are similar with the ratio of 1:1 [2]. However, among the patients evaluated in our study, those with benign tumors were more common. Operative procedure was a wide resection for a malignant PTR and resection of only one affected rib due to a benign PTR diagnosed during the operation by frozen studies.

2. Materials and methods

Seventy-eight patients with primary rib lesions were operated in our clinic. The diagnosis of PRT was established through clinical and radiological studies conducted at Atatürk Chest Disease and Chest Surgery Training and Research Hospital, Ankara, between January 1998 and July 2008. The patients with lung, breast, and skin carcinomas that were invasive of the chest wall, hyperplasia due to known rib fracture, and histology findings consistent with callus appearance criteria were excluded from this retrospective study.

The diagnostic work-up consisted of a detailed history, physical examination, and postero-anterior and lateral chest radiographs, and thoracic CT-scan in all the patients. To rule out bone metastasis, radionuclide bone scan was conducted in 23 patients with malignant PTR and positron emission tomography (PET CT) was routinely conducted in six patients, whose lesions were reported to be malignant after the operation. Fine-needle aspiration biopsy was performed on 11 patients who had three or more rib lesions and infiltration sites through the lung parenchyma to distinguish whether the lesion was primary rib tumor or not. All the patients were operated on.

3. Results

Of the 78 patients included in this study, 49 (63%) had benign lesions. The patients with benign lesions ranged in age between 9 and 80 years (mean age: 38 years), and 29 (37%) had malignant lesions. These patients ranged in age from 7 to 77 years (mean age: 43.9 years). Of the 49 patients (30 male, 19 female) with benign PTR: 21 patients (26%) had osteochondroma; 13 patients (16%), enchondroma; 12 patients (15%), fibrous dysplasia (FD); 1 patient (1%), granular cell tumor, and 2 patients (2%), aneurysmal bone cyst (ABC). Of the 29 patients (19 male, 10 female) with malignant PTR, 5 patients (6%) had plasmacytoma; 2 patients (2%), chondrosarcoma; 12 patients (15%), osteosarcoma; 8 patients (10%), Ewing's sarcoma; and 2 patients (2%), hemangiosarcoma. There were 48 male (63%) patients and 30 female (37%) patients. The mean age of all the patients was 40.2 years (range: 7–80 years). The right side was affected in 33 patients (42%), the left side was affected

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in 45 patients (57%). Forty-seven patients (60%) had only one costae lesion and 31 patients (40%) had two or more costae lesions.

In the patient group with benign PTR, 40 patients (81%) complained of swelling. Twenty-three patients (46%) had pain and eight patients (16%) were asymptomatic. In the patient group with malignant PTR, 21 patients (72%) complained of swelling, 25 (86%) had pain, and four patients (13%) were asymptomatic. In the patient group with malignant PTR, the main symptom was pain, but in the patient group with benign PTR, the main symptom was swelling. Eight patients in the benign group and four patients in the malignant group were asymptomatic (accidental radiological findings in recruitment centers).

All the patients underwent operation except 11 patients with preoperative biopsy. These 11 patients had three or more rib lesions and infiltration sites through the lung paranchyma on the thorax CT. In order to determine whether the lesion was a primary rib lesion, fine-needle aspiration biopsy was planned.

Our treatment policy for PTR was generally as follows: a total excisional biopsy for patients with one or two rib lesions with the patients under general anesthesia, and if the lesion was proved to be benign during surgery by frozen studies, only the affected ribs were resected until tumor-free margins were achieved. When the lesion was proved to be malignant, a wide resection was performed. Wide resection of a malignant PTR included the affected rib with at least a 4–5 cm free margin proximal and distal to the tumor, also resection of portions of the ribs immediately above and below the tumor, the adjacent muscles, and the pleura. In addition, the tissues adherent to the tumor were excised. Histopathological specimens were reviewed recently in order to re-adjust the diagnoses according to current classification.

Chest wall resection and reconstruction with synthetic polypropylene mesh and local muscle flaps can be performed as a safe, effective one-stage surgical procedure for a variety of major chest wall defects [3–6]. Chest wall closure was achieved without difficulty in all the patients except in six patients (20%) in the malignant group who had three or more than three affected ribs. To cover large defects of the lateral chest wall in the resection of three or more ribs, especially in these six patients with malignant tumors, partial transposition of the latissimus dorsi muscles or pectoralis major muscles was used. No synthetic prostheses were used. The middle axillary line was the borderline to distinguish the anterior from the posterior chest wall, and the anterior from the posterior parts of a rib.

All malignant PTR were under chemotherapy and radiotherapy. No survival analysis was conducted.

4. Conclusion

Chondrosarcomas, the most common malignant primary tumors of the chest wall, usually occur in the anterior part of the wall, arising from the costochondral arches or sternum [7–9]. Nevertheless, in our patients, the incidence rate of chondrosarcoma in all the primary rib tumors was 2%, and 7% in all the malignant tumors. Two peak periods of prevalence have been identified, the first at <20 years of age, and the second at >50 years of age. These tumors occur twice as often in men as in women [10]. As seen, our two patients with chondrosarcoma were older than 50 years and both were male.

Osteosarcomas are malignant mesenchymal neoplasms that rarely occur in the thorax. The rib, scapula, and clavicle are the most frequent sites of origin [10]. Osteosarcoma is the second most frequent malignant rib tumor (Fig. 1f). The incidence rate of osteosarcoma in all primary rib tumors is 15% and 41% in all malignant rib tumors. In our patients, the incidence rate of osteosarcoma in all the primary rib tumors was 15%. Osteosarcoma is more common in males. Similarly, in our study, there was a male predilection, and the mean age of the patients was 41.6 years with a range of 7–75 years.

Ewing's sarcoma is a relatively rare malignant bone neoplasm that usually occurs in children and young adults. The ribs are frequent sites of primary Ewing tumor accounting for about 10–12% of all cases. The lesions may be predominantly lytic or sclerotic or show a combination of bone destruction and secondary reactive changes (Fig. 2b) [11]. In our study, there was a male pre-dilection and it was determined in 33% of all the malignant rib tumors and in 10% of all the primary rib tumors. The mean age of the patients with Ewing tumor was 23.6 years (range: 12–41).

Osseous angiosarcoma commonly occurs in long bones and few occur in the ribs. These lesions are characterized by solid sarcomatous areas, abortive vessel formation, and vascular areas exhibiting anastomosing channels (Fig. 1d). In our study, there were two cases of hemangiosarcoma with an equal gender distribution and with a mean age of 65.5 years (range: 56–77 years). In our series, osseous angiosarcoma was detected in 2% of all the PTR.

Localized solitary plasmacytoma of the bone (SPB) is a rare disease and is characterized by only one or two isolated bone lesions with no evidence of disease dissemination (Fig. 2c). In the literature, the ratio of male to female patients was approximately 1.3:1. The average age on presentation was 59.5 years with a range from 39 to 77 years [12]. In our study, there was a male predilection. It was determined in 17% of all the malignant rib tumors and in 6% of all the primary rib tumors (Fig. 1e). The mean age of the patients with this lesion was 69.8 years (range: 60–76 years).

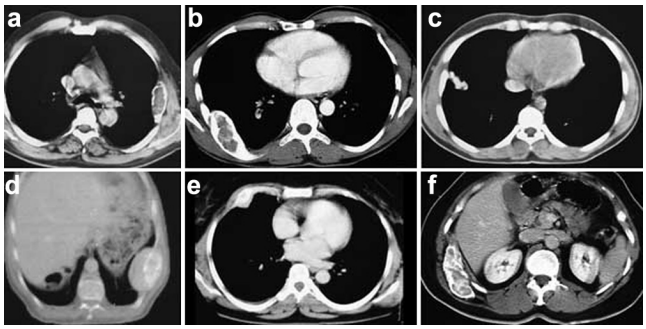


Fig. 1. (a) Aneurysmal bone cyst, (b) fibrous dysplasia, (c) osteochondroma, (d) osseous angiosarcoma, (e) plasmacytoma, (f) osteosarcoma.

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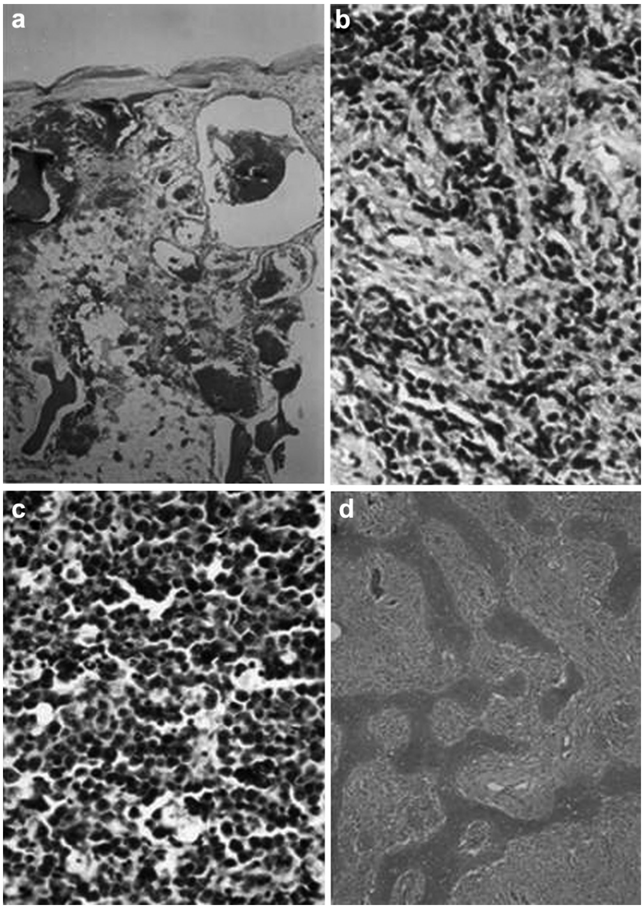


Fig. 2. (a) Aneurysmal bone cyst, (b) Ewing's sarcoma, (c) plasmacytoma, (d) fibrous dysplasia.

Enchondroma is a cartilaginous tumor that develops in the medullary cavity. Radiographs typically show a lobulated, well-demarcated, and osteolytic lesion that demonstrates mild expansion and well-defined, sclerotic margins. Enchondroma of the ribs accounts for 2–12% of primary rib tumors [13]. In our study, the rate of enchondroma in all benign rib tumors was 16% with a slight male predilection. The majority of patients are males between the age of 20 and 50 years.

An osteochondroma is a hamartomatous cartilage capped bony growth that projects from the surface of the affected bone and most commonly presents in the second decade of life. In the ribs, these tumors occur with particular frequency at the costochondral junction. The tumors are characteristically pedunculated or sessile osseous protuberances arising from the surface of the parent bone [9] (Fig. 1c). They account for 2–8% of primary rib tumors [13]. Likewise, in our study, there was a male predilection. The mean age of the patients with osteochondroma was 57.1 years (range: 13–75 years). In our series, the rate of this lesion was 26% in all the PTR.

FD is a developmental skeletal anomaly in which osteoblasts fail to undergo normal morphological differentiation and maturation, leading to replacement of normal marrow and cancellous bone by immature bone and fibrous stroma (Fig. 2d) [13]. FD is the most common benign rib tumor accounting for ~30% of benign tumors of the chest wall [13]. Approximately 70–80% of cases are monostotic and 20–30% are polyostotic. The age range of patients with monostotic disease is 10–70 years, but recognition is most frequent at 20–30 years of age. Patients are usually asymptomatic, although pathologic fracture resulting from FD can cause pain [9]. In our study, FD accounted for 15% of all the primary rib tumors, with the patients ranging in age from 25 to 68 years (mean age: 42.4 years). There was also a male predilection. Radiographically, FD shows unilateral fusiform enlargement and deformity with cortical thickening and increased trabeculation of one or more ribs (Fig. 1b).

An ABC is an expansile, osteolytic lesion consisting of blood-filled cystic spaces separated by connective tissue septa containing bone or osteoid and osteoclast giant cells (Fig. 1a). An ABC involving the rib is very rare (Fig. 2a). Forty-four cases have been reported in the literature [14]. ABC accounts for 5% of primary rib lesions. In our series, there were two patients with ABC, accounting for 2% of all the primary rib tumors. Most ABC occur in patients under the age of 30 years [14]. The mean age of the patients was 42.2 years (range: 26–59 years).

As a summary, 78 patients were included in our study. Our limited experience in the management of primary rib tumors showed that the number of the patients with benign lesions were much more than from the patients with malignant lesions. The mean age of the patients with malignant tumors is generally higher than that of the patients with benign rib lesions [10], as in our patients (43.9 years vs. 38 years). Generally, the rate of benign lesions is reported to show equal gender distribution with a slight male pre-dilection [2, 15]. Similarly, it was determined in 63% of the male patients in our study. The left side ribs were affected much more than according to the right side ribs. Most of the patients had only one costae lesion. In the patient group with malignant PTR, the main symptom was pain, but in the patient group with benign PTR, the main symptom was swelling. Asymptomatic patients were much more in the benign group than according to the malignant group. All PTR should be considered malignant until proven otherwise. Surgical treatment of primary malignant tumors of the ribs, when confirmed, is wide resection of the tumor. If malignancy is not proved, surgery must consist of wide resection with tumor-free margins in order to provide the best chance for cure in both benign and malignant lesions. All malignant PTR must be given under chemotherapy and radiotherapy.

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