

Tumours of the ribs: experience with 47 cases

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Abstract

Objectives: To emphasise the existing difficulties in differentiating benign from malignant rib tumours, and especially the problems that a clinical doctor encounters when dealing with a hyperplastic rib. **Methods:** Forty-seven patients with rib tumour underwent surgery in a period of 12 years (1984–1996). In 40 cases (85%), the lesion was benign and in seven (15%) was malignant. Twenty-one benign tumours originated from cartilage and bone, seven were inflammatory, six originated from the bone marrow, and minor percentages (2.5–5%) had vascular, neurogenous, degenerative or miscellaneous origin. Three of the malignant tumours were primary chondrosarcomas and two were metastatic from kidney. The rest were metastatic from stomach (adeno-Ca), and skin (melanoma). The mean age in the benign group was 25.2 years and in the primary malignant group was 20.7 years. Related symptoms were pain (47%) and swelling (42.5%). One-third (32%) of the patients were asymptomatic and the lesion was accidentally found during routine chest radiography. All patients were treated surgically with wide excision of the tumour and the diagnosis was established histologically. **Results:** Resection was complete and curative in all cases without recurrence. **Conclusions:** Since the likelihood of malignancy cannot be excluded, all rib tumours should be considered malignant until proven otherwise. Therefore, prompt intervention is necessary and wide and radical initial excision of the involved rib is advocated. © 1999 Elsevier Science B.V. All rights reserved.

Keywords: Thorax; Chest wall; Malignant; Benign; Rib hyperplasia; Costal cartilage

1. Introduction

Rib tumours are uncommon comprising 5–10% of all bony tumours [1–3]. Previous reports have stressed that malignant neoplasms are significantly more common than are benign ones [3,4]. Our series comes from a large military hospital treating mainly youngsters. Bearing this in mind, we analysed our cases and reviewed our experience.

2. Materials and methods

We operated on 47 Caucasians, admitted, investigated and treated for a rib lesion (clinically or radiologically identified) at the Army General Hospital, Athens from 1984 to 1996. Cases meeting any of the following criteria were excluded from this retrospective study: breast, skin and lung carcinomas invading the chest wall, hyperplasia due

to known rib fracture, histology findings consistent with callus appearance.

Finally, 47 patients were included in this study and are analysed. Forty (85%) had benign lesion (Table 1) and seven (15%) had malignant (Table 2). There were 45 (96%) male patients. The average age was 28.1 years (range 19–74). The left side was affected in 28 cases (60%). Twenty-five lesions (53%) were located in the upper six ribs. The tumour was anterior in 22 patients (47%), lateral in 16 (34%), and in the paravertebral portion in nine (19.1%). Twenty-two patients complained of pain, 20 had palpable swelling and 15 (32%) were asymptomatic. Computed tomography scan (CT-scan) was performed in all but 15 older cases and in very few, fragmented periosteum was shown. Isotopic bone scan was performed in 27 patients (positive in 21). All patients underwent radical dissection without preoperative biopsy. No chest reconstruction was needed. Histopathological specimens were reviewed recently in order to readjust the diagnoses according to current classification.

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Table 1
Histologic analysis of benign rib tumours

Origin	Type of tumour	No. of patients
Cartilage and bone	Chondroma	10
	Osteochondroma	9
	Chondroblastoma	1
	Chondromyxoid fibroma	1
Marrow	Eosinophilic granuloma	4
	Reactive hyperplasia	1
	Lipoma	1
Inflammatory or infectious	Necrotizing granulomatosis	3
	Osteomyelitis	2
	Tietze's Syndrome	2
Vascular	Hemangioma	2
Neurogenous	Neurilemoma	2
Degenerative	Hyperparathyroidism	1
Miscellaneous	Myositis ossificans	1
Total		40

2.1. Benign tumour analysis

Nineteen out of 40 patients with benign lesions experienced local pain, 17 had a palpable swelling and 13 (32.5%) were asymptomatic (accidental radiological findings in recruitment centres). Eleven patients had a radiological enlargement of the rib and eight had rib expansion; only four of them had fragmented periosteum seen on CT-scan.

Twenty one male patients had tumours of cartilage and bone origin (Table 1). Ten had chondroma (Fig. 1), nine osteochondroma (Fig. 2), one had chondroblastoma and another chondromyxoid fibroma. Of the remaining 19 patients, six had tumour of marrow origin (Fig. 3), and seven of infectious (Fig. 4). Vascular and neurogenous tumours were each encountered in two patients. Finally, one patient had degenerative lesion (hyperparathyroidism) and another had myositis ossificans.

2.2. Malignant tumour analysis

Seven male patients (four left-sided), aged 20–74 years, had rib carcinoma (Table 2). Three had primary chondrosarcoma and four had metastatic carcinoma.

2.2.1. Chondrosarcoma

Three patients belonged to the 20–22-year group and in two, the tumour was left-sided. One patient complained of pain and one had swelling. The third was entirely asymptomatic. All three tumours radiologically produced expanding

Table 2
Histologic analysis of malignant rib tumours

Type of tumour	No. of patients	
Primary	Chondrosarcoma	3
Metastatic	Renal cell carcinoma	2
	Gastric adeno-Ca	1
	Melanoma	1
Total		7

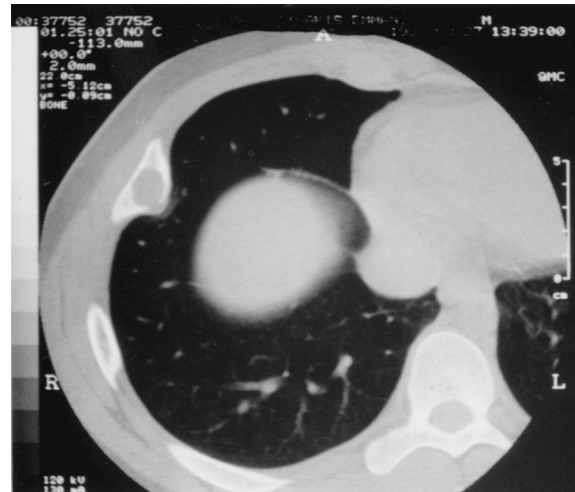


Fig. 1. Chest CT-scan of a 18-year-old man with chondroma of the anterior segment of the right 7th rib.

lesions, and positive isotope bone scan. The patients underwent radical resection including the pleura and the adjacent soft tissues. As soon as histological diagnosis was made, the patients were led to re-operation in which one rib above and one below were resected. No synthetic materials were placed and no chemotherapy or radiotherapy was given postoperatively. All patients are free of disease 13, 11 and 7 years after surgery.

2.2.2. Metastatic carcinoma

Metastatic carcinoma was found in four patients. One aged 74, presented with two right-sided rib lesions, visible in isotope bone scan. The patient had a history of recurring episodes of gastric ulcer and in one occasion adenocarcinoma was suspected on gastroscopic grounds, however, was



Fig. 2. Chest CT scan of a 20-year-old man with osteochondroma of the left 3rd rib.

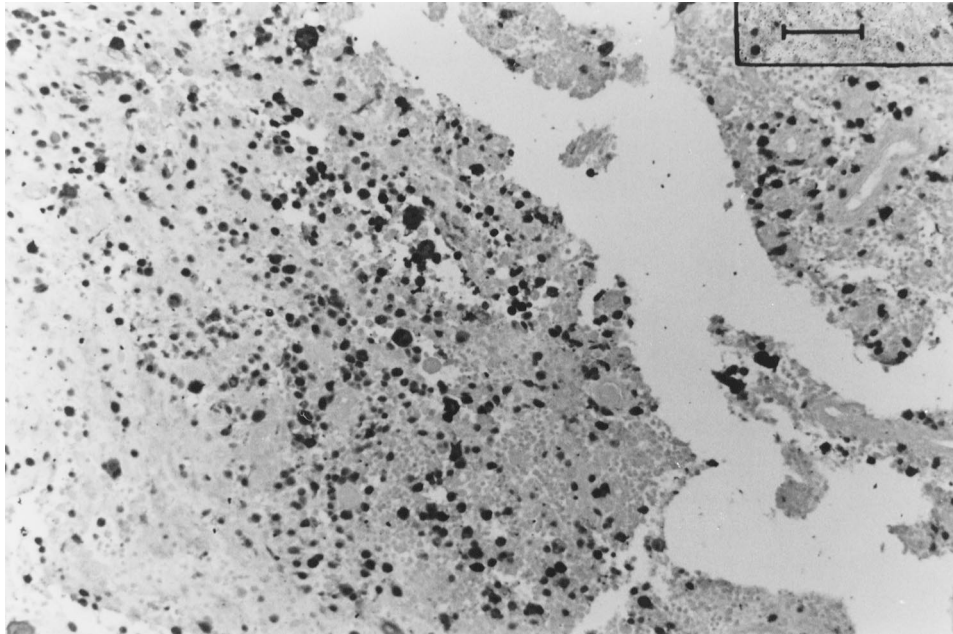


Fig. 3. Histologic appearance of eosinophilic granuloma. Anti-S-100 immunostaining reveals several histiocytes (brown staining), APAAP method, DAB staining $\times 100$.

not firmly diagnosed nor treated. The second patient aged 47, previously had a melanoma of his back resected and 2 years later developed rib metastasis. The other two patients had had a nephrectomy for renal cell carcinoma (Grawitz type) and 5 years later developed rib metastasis (Fig. 5). All four patients underwent wide resection of the metastatic lesions. The patients with Grawitz tumour are disease-free, for 7 and 6 years and the one with melanoma 2 years, respectively. The adenocarcinoma case was lost to follow-up 3 years postoperatively.

3. Discussion

The proportion of benign versus malignant rib tumours is reported 43–52% [4,6,10]. However, depending on the material and the nature of the Hospital involved, the benign lesions' proportion may raise up to 78% [3], as happened with our cases (85%). Most authors report equal sex distribution with a slight male predilection [9]. The average patient age is generally higher in malignant than in benign rib lesions [4]. However chondrosarcoma being the most common malignant tumour [4,6,8,11–13] involves young people aged 16–30 years (like our cases) most probably arising in pre-existing osteochondroma [10].

Pain and/or swelling, provides no safe guide to the nature of a rib lesion [9,10] since 61% of benign and 48% of malignant tumours may present with pain [4,10]. In our series, 47.5% and 42.5% of benign lesions presented with pain and swelling respectively. One in three of our patients with chondrosarcoma, was asymptomatic. Symptomless patients are reported to be as high as 31% [4]. The pain can be either pleuritic (growth within the pleural space), or neuritic (intercostal nerves invasion) [2].

Roentgenography is useful, not always resulting in visualisation of an existing tumour. CT-scan is helpful sometimes demonstrating fragmented periosteum suspicious of malignancy. Irregular or destructed ribs, sometimes with calcific mottling, indicate malignancy [3].

Preoperative diagnosis is difficult. Rib tumours should be considered malignant until proven otherwise [6,8,9,17]. Needle biopsies cannot provide enough material [9,14], and the identity of the tumour will be obtained only after excisional biopsy. CT and Isotope bone scan are necessary,



Fig. 4. Chest CT scan of a 33-year-old man showing a lytic lesion of the left 2nd costal cartilage due to salmonella osteomyelitis.

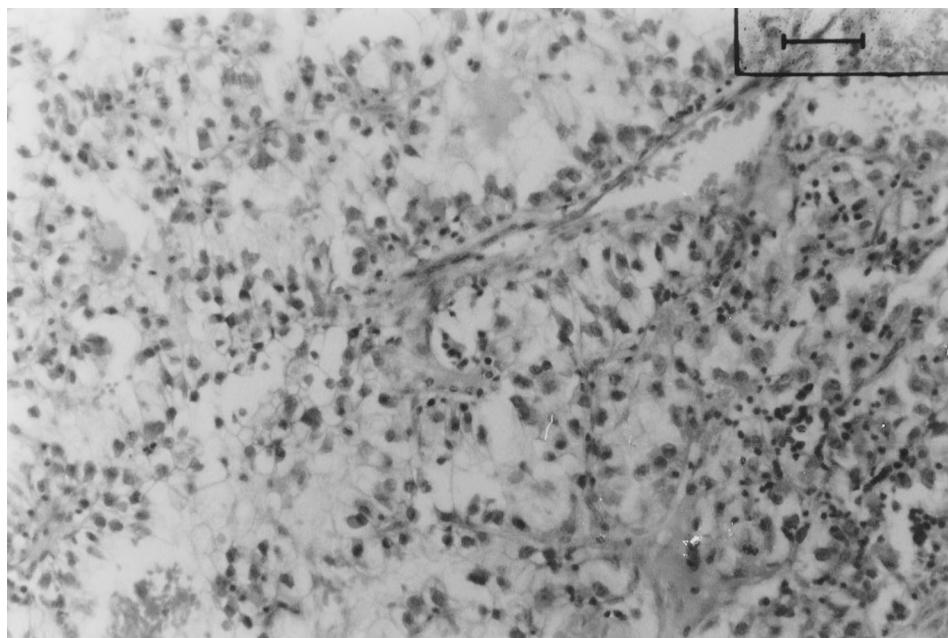


Fig. 5. Histologic appearance of metastatic foci of renal cell carcinoma, in the rib. H and E $\times 200$.

because all primary malignant rib neoplasms can metastasise to bones, or, a second primary tumour may be discovered [15].

3.1. Benign rib tumours

Cartilaginous tumours, are the most commonly (50%) encountered rib lesions [4,9,16] with chondroma and osteochondroma being the most frequent. The majority are males below the age of 21. Chondroma is more often situated near the costochondral junction [1,5,9,11,17] as in our cases (90%). They are likely to become malignant (11%) after years of benign growth [18]. This incidence is lower (1–2%) for osteochondroma and may raise in familial syndromes of multiple osteochondromas [19].

Other cartilaginous benign neoplasms are chondroblastoma (less than 1% of 2900 bone tumours reviewed at the Mayo Clinic [20], affecting young people, like our 23-year-old patient) and Chondromyxoid fibroma (2% of benign bony tumours [20], affecting persons aged 10–30 [21]).

Osteoma, osteoid osteoma and osteoblastoma, all rarely located in the ribs, comprising the benign bony group (not encountered in our series).

Lipoma and eosinophilic granuloma are tumours arising within the marrow. Lipomas of bone are uncommon. Rib involvement is extremely rare. Our case was diagnosed only after rib resection and careful microscopic study. Eosinophilic granuloma is a solitary lytic lesion arising from the reticuloendothelial system, and ribs are frequent locations (12.4%) [22]. Its pathogenesis is unknown and it occurs from infancy to the 6th decade. The majority of cases appear before the age of 30 with a slight male preponderance [2]. Dominant symptom is pain of short duration. It is not neces-

sary for these patients to have peripheral eosinophilic counts.

Fibrous tissue tumours within the rib are xanthoma, fibromyxoma, ossifying or nonossifying fibroma, fibrous dysplasia, and desmoplastic fibroma. They are very rare, and apart from fibromyxoma these tumours affect the younger age group. Neurilemoma (schwannoma) is the main neurogenous rib tumour, arising from the nerve sheath. It has to be distinguished from neurofibroma in which encapsulation does not exist. Neurilemoma occurs in the 4th decade of life, shows no sex predilection and presents with pain and/or swelling. Pathologic fracture occasionally occurs [23]. Hemangioma is the main vascular benign lesion and can be diagnosed by its 'soapbubble' or 'honeycomb' appearance on the radiograph.

3.2. Malignant rib tumours

Fifteen percent of the patients with primary malignancy, are asymptomatic at the time of diagnosis and this incidence surpasses 40% with metastatic neoplasms [4]. Radiologically, it is difficult to distinguish benign from malignant rib tumours, and even more difficult to differentiate primary rib cancers from adjacent tumours involving the rib [2].

Chondrosarcoma is the most common primary malignant rib tumour [1,4,6,8,12], arising either de novo in a rib (90%) or it is superimposed on a pre-existing cartilaginous neoplasm. Most commonly encountered in the 5th decade and before the age of 20, this tumour occurs more frequently in men [2]. Although this tumour may involve any part of a rib, it usually arises at the costochondral junction, as happened in two of our three cases. The 5-year survival figures vary from 50 to 76%, and 10-year rates from 35 to

69% [24]. Five-year survival for patients undergoing complete resection is 69%, compared to 50% for those with incomplete and 20% for the ones who had no resection [15].

Osteosarcoma is the second most frequent malignant rib tumour. It is relatively rare comprising 3% of all osteosarcomas. More common in males in the twenty to thirty age bracket. In later years, this tumour is often associated with Paget's disease, frequently presenting itself as a fracture. Bone irradiation can also cause osteosarcoma. The 5-year survival rates range from less than 10–16.5% [15].

Ewing's sarcoma is a highly malignant and relatively uncommon (6.5%) rib tumour [25]. It predominantly affects young males presenting itself as pain and swelling in the tumour area. Anaemia, increased sedimentation rate and leukocytosis may be present. Prognosis is poor and the 5-year survival rates vary from 0 to 8% [26].

Fibrosarcoma is a relatively rare malignant primary rib tumour originating from fibrous elements of the medullary cavity. It usually occurs in the twenty to forty age group resembling radiologically to chondrosarcoma [2].

Angiosarcoma, plasma cell myeloma, reticulum cell sarcoma, liposarcoma, leiomyosarcoma, giant cell tumour, hemangioendothelioma and chordoma are extremely infrequent malignant tumours which are rarely encountered in the ribs.

3.3. Treatment of rib tumours

It is generally advocated that all primary rib neoplasms should be treated by complete en bloc excision [2,4,8,9]. As it is impossible in most instances to determine with certainty, whether the tumour is benign or malignant by radiographs alone, the policy should be to resect all mono-ostotic tumours radically because of the likelihood of malignancy. Excisional wide rib resection is recommended. This must include resection of the involved rib, the corresponding costochondral arches, and several partial ribs above and below the neoplasm [27]. Four to five centimetres of intact bone should be removed on both sides of the tumour as recurrences in the rib stumps, particularly of chondrosarcoma, are fairly common [2]. Furthermore, nodules of malignant cells are often found in the periosteum some distance from the main mass. If there is infiltration of the soft tissues, pleura, lung parenchyma or diaphragm, one should not hesitate to excise the involved portions. The size of the tumour should not be contraindication for radical removal. The main and major therapeutic error in handling rib tumours, is failure to perform a radical excision. Resection of large segments of the chest wall is well tolerated and usually results in little functional impairment [4]. Reconstruction includes stabilisation of the thorax rigidity and coverage of any soft tissue defect [4,28]. Prognosis of benign tumours after surgical excision is uniformly excellent. For malignant neoplasms, adequacy of surgical exci-

sion is an important determinant of the incidence of recurrence and survival.

For chondrosarcoma, the treatment of choice is complete resection. There is currently no effective chemotherapy for this tumour. On the contrary, rib osteosarcoma is responsive to chemotherapy. Burt (1994) recommends resection followed by chemotherapy [15]. Ewing's sarcoma is also responsive to chemotherapy. There is a tendency to treat Ewing's tumours with preoperative chemotherapy, followed by either resection, if possible, or irradiation, the results of surgery being superior to that of radiation therapy [15]. Solitary rib plasmacytoma is sensitive to radiation and in most modern series this type of therapy is the primary treatment modality [7,29]. Local control was achieved in 62–100% of patients with radiation doses from 3000 to 4600 cGy.

The present study and our review of the literature indicate that accurate diagnosis is fundamental in successful management of any rib tumour, since types of treatment vary for each neoplasia.

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