CLINICAL REPORT

Costal chondrosarcoma. Report of five cases

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Summary
We report five cases of costal chondrosarcoma (CS) in four women and one man between 28 and 49 years of age. In four cases, the tumor had spread and infiltrated the adjacent structures (soft tissues, thoracic vertebrae, mediastinum, etc.). CS was diagnosed based either on wide surgical resection specimens in three patients, or on tumor biopsies in two cases. The CS was grade I in one patient, grade II in three cases, and grade III in one case. Wide surgical resection which was performed in three patients was associated in one case with adjuvant radiotherapy and chemotherapy. In the three cases, the disease natural history appeared favorable after a follow-up duration ranging from 1 month to 4 years. Two patients died, one after radiotherapy and chemotherapy done immediately after diagnosis as a result of the large size and invasive nature of the tumor. Coastal CS is characterized by a distinct potential for locoregional and distant metastasis. Diagnosis remains anatomopathological. Wide surgical resection is the only demonstrated curative treatment, even for high-grade CS. © 2009 Elsevier Masson SAS. All rights reserved.

Introduction
Malignant primary bone tumors of the thoracic wall are rare. They account for 4.5 to 8% of all bone tumors. Most often, they are located in the ribs and are predominantly chondrosarcoma (CS), which alone accounts for 40% of all malignant rib tumors [1]. Its diagnosis is suspected based on imaging studies, with definitive diagnosis based on histology. CS is a malignant tumor with cartilaginous differentiation, with no bone or osteoid production, but it can contain foci of tumor-induced or enchondral ossification. It is characterized by a high potential for locoregional invasion and recurrance after treatment [2]. Five cases of costal CS are reported (Table 1).

Observations
Observation No. 1
A 41-year-old female patient was hospitalized for a left-sided cervicothoracic mass that had been evolving for
<table>
<thead>
<tr>
<th>Case</th>
<th>Sex/Age</th>
<th>Circumstances at discovery</th>
<th>Imaging</th>
<th>Size/grade</th>
<th>Treatment</th>
<th>Follow-up/progression</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F/41 years</td>
<td>Left cervicothoracic mass + cervicobrachial neuralgia</td>
<td>Parietal tissue mass, anterior-external, centered on 1st rib. Central necrosis and peripheral contrast agent uptake. Parietal, mediastinal infiltration and invasion of large venous trunks</td>
<td>15 cm/GII</td>
<td>Radiotherapy + chemotherapy</td>
<td>Coastal and vertebral extension + lung metastases. Death</td>
</tr>
<tr>
<td>2</td>
<td>F/42 years</td>
<td>Chest pain</td>
<td>Apical opacity flaring anterior 1st rib with cortical rupture</td>
<td>4 cm/GI</td>
<td>Resection of superior arc of 1st rib</td>
<td>3 years/good</td>
</tr>
<tr>
<td>3</td>
<td>M/28 years</td>
<td>Swelling under axilla fistulized at skin</td>
<td>Expansive tissue process infiltrating soft tissue and wall + multiple rib fractures</td>
<td>20 cm/GIII</td>
<td>Peritectomy</td>
<td>1 month/lost to follow-up</td>
</tr>
<tr>
<td>4</td>
<td>F/49 years</td>
<td>Right cervicobrachial neuralgia</td>
<td>Posterior and superior mediastinal mass infiltrating thoracic vertebra and surrounding vessels</td>
<td>6 cm/GII</td>
<td>En bloc resection planned but not performed</td>
<td>4 months/death</td>
</tr>
<tr>
<td>5</td>
<td>F/31 years</td>
<td>Chest pain</td>
<td>Mediastinal mass 5 cm posterior, with lumpy calcifications in periphery + fracture of 6th rib</td>
<td>8 cm/GII</td>
<td>Surgery + chemotherapy and radiotherapy</td>
<td>4 years/good</td>
</tr>
</tbody>
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Figure 1  Axial thoracic CT: large tumor involving the left first rib with homolateral mediastinal and soft tissue extension, with a high peripheral contrast-enhancement and a central necrosis (case 1).

9 months, with persistent symptoms of cervicobrachial neuralgia. The physical examination found a 15-cm mass at its widest point filling the left clavicular fossa. The chest X-ray showed an opacity at the apex of the left lung, measuring 8 cm at its longest axis, extending to the mediastinum and the lateral thoracic wall. The thoracic computerized tomography (CT) scan demonstrated a parietal tissue mass, anteroexternal, substantially enhanced in its periphery after injection of contrast agent and delimiting central necrotic zones. This mass was centered on the cartilage of the first left rib with its mid and posterior arc fractured. This mass presented wide ipsilateral parietal and mediastinal extension with invasion and thrombosis of the left innominate venous trunk and the left jugular vein (Fig. 1). Abdominal ultrasound and CT showed no secondary lesions. The histological exam of the ultrasound-guided mass biopsy showed grade II CS. Given the extent of the tumor's locoregional invasion, surgical resection was not planned. Antalgic radiotherapy at a dose of 20 Gy on the chest wall associated with four courses of palliative chemotherapy with ifosfamide, Uromitexan®, dacarbazine, and doxorubicin were administered. Clinical progression was marked by a slight reduction in the tumor's size in the left supraclavicular fossa, likely secondary to the resorption of the peritumoral edema. Nevertheless, the follow-up thoracic CT scan showed tumor extension with osteolysis of the second rib and the first thoracic vertebra as well as the appearance of secondary lung nodules. A new course of chemotherapy was decided but the patient died from the disease after the 12th round.

Observation No. 2

A 42-year-old woman consulted for chest pain on the right side, which had appeared 5 years before, irradiating to the ipsilateral upper limb. X-rays of the thorax at this time showed a right apical opacity, 5-cm at its widest point, oval with clear limits (Fig. 2). The radiological diagnosis argued in favor of a benign lesion. The patient was lost to follow-up for 5 years. The follow-up chest X-ray revealed a slight 0.5-

cm increase in the size of the lesion at its largest diameter. The thoracic CT showed an expansive lesion of the internal extremity of the anterior arc of the first right rib, flaring the cortex, which was broken in several spots. The anterior arc of the first rib was resected via the axilla using a Roos approach: an incision between the external edge of the pectoralis major muscle in front and the external edge of the latissimus dorsi muscle in back, done carefully with the patient in the lateral decubitus position and the arm maintained at 90°. Macroscopically, the rib segment measured 6 cm in length, the seat of an osteolysis shaping a recess partially occupied by a tumor nodule measuring 4 cm at the widest axis, with a shiny chondroid aspect. On histological examination, this nodule had the aspect of well-differentiated grade I CS, extending in a fine line along the compartment wall. The limits of the resections were not infiltrated. Follow-up thoracic CT and bone scintigraphy, done 3 years after surgery because of the onset of thoracic pain on the right side, were normal with no signs indicating recurrence or metastasis.

Observation No. 3

A 28-year-old male, who had been operated on 4 years before for a hydatid cyst of the eighth left rib, consulted for swelling under the left axilla. The 20 × 15-cm tumor was painless, firm, and fistulized at the skin by an orifice suppurating purulent secretions. The scar of the thoracotomy was good quality. The thoracic X-ray showed a left-sided paracardiac opacity, oval in shape, heterogenous, and poorly limited, 16 cm at the largest diameter, associated with costal lysis. The rib cage showed loss of bony segments in the posterior arcs of the sixth and seventh left ribs and a fracture of the eighth left rib (Fig. 3). The CT showed an expansive process infiltrating the soft tissues of the left axillary region, with a lateral parietal extension, without affecting the parenchyma of the lung, associated with fracture of the sixth, seventh, and eighth ribs. During surgery, the tumor was found to extend to the spinal cord up to the anterior angle of the ribs. A peritectomy removing the posterior and lateral arcs of the sixth, seventh, and eighth
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Figure 3  Plain roentgenogram shows bone destruction involving the posterior sixth and seventh ribs with osteolysis of the eighth rib, adjacent to a left pericardial heterogeneous mass (case 3).

ribs was performed. On macroscopic examination, the surgical specimen weighed 1350 g and measured 25 × 16 cm. The whitish-grey tumor with a few necrotic-hemorrhagic reorganizations, was infiltrating the three ribs. The limits of the surgical resection were healthy. The histological examination showed that the tumor corresponded to grade III CS. The immediate and delayed postoperative course was uneventful. The patient was discharged on the tenth postoperative day, with a satisfactory follow-up chest X-ray. Seen 1 month after the intervention, the patient was doing well and has since been lost to follow-up.

Observation No. 4

A 49-year-old female patient consulted for cervicobrachial neuralgia on the right side occurring over several months. The clinical examination showed nothing in particular. The frontal chest X-ray showed a rounded right-sided paratracheal opacity, with well-defined limits, measuring 6 cm at its widest point. The thoracic CT showed a heterogenous mass of the upper and mid sections of the posterior mediastinum, the seat of nodular and arciform calcifications (Fig. 4). Thoracic magnetic resonance imaging (MRI) showed that this mass was centered on the posterior arc of the second right rib, invading the second thoracic vertebra with no intraluminal extension and pushing the trachea and the right subclavicular artery toward the left. Cervical and abdominal ultrasound did not bring out secondary locations. Several biopsies of the tumor were done by cervicotomy. On histological examination, certain biopsy fragments showed the aspect of grade II myxoid CS that was focally necrotic. En bloc resection of the tumor by median sternotomy enlarged by a cervicotomy was planned but the patient died before the surgery, 4 months after the biopsy, because of substantial invasion of the trachea.

Figure 4  Axial thoracic CT: large tumor arising from the posterior mediastinic space with arciform calcifications (case 4).

Observation No. 5

A 31-year-old female was hospitalized with thoracic pain on the left side with intercostal irradiation. The physical examination found pain on pressure of the left sixth costal-vertebral joint. At the sixth rib’s posterior arc, the chest X-ray revealed a heterogenous opacity, with clear external limits and indistinct internal limits. The thoracic CT showed a posterior mediastinal mass, hypodense, 5 cm at its widest point, with nodular lumpy calcifications in the periphery. The tumor seemed to thicken the surrounding epidural spaces and did not enhance after injection of contrast agent. It was associated with a fracture of the posterior arc of the sixth rib. The thoracic MRI demonstrated an oval posterior mediastinal mass at the left costal-vertebral groove, with regular contours (Fig. 5). This mass presented an intermediate signal on the T1-weighted sequences and a slightly heterogenous signal on the T2-weighted sequences, with mostly heterogenous peripheral enhancement after injection of gadolinium. This aspect suggested a nerve sheath tumor that had developed at the posterior mediastinum.

Figure 5  Sagittal MRI reveals a well-circumscribed mass of the posterior mediastinic space (case 5).
Bone scintigraphy showed heterogenous hyperfixation at the posterior arc of the left sixth rib. A left posterolateral thoracotomy was performed. It demonstrated a tumor formation centered on the posterior arc of the sixth rib. The tumor was resected with dislocation of the head of the sixth rib and resection of the posterior arc of the fifth rib and the sixth intercostal space. On macroscopic examination, the sixth rib was totally occupied by a bilobed tumor measuring 8 cm at the widest point, with a multilobular and translucent aspect. On histological examination, this aspect argued in favor of grade II CS. The resection limits were not infiltrated.

After the intervention, five courses of chemotherapy and 27 sessions of radiotherapy were given. An X-ray of the rib cage taken 1 year later showed radiation-induced costal lesions, with no evolving tumoral lesion. A thoracic CT scan taken after 4 years showed no signs of recurrence or metastasis.

Discussion

Primary tumors of the ribs are rare and malignant in 29% of cases. They are dominated by CS, which accounts for 40% of all malignant tumors of the ribs [1]. The first five ribs are the most often affected and the lateral costal location seems to be more frequent than anterior and posterior locations [3]. Rare before the age of 20 years, CS affects most particularly middle-aged men, between the fourth and fifth decades [4]. This male predominance was not found in our series, which included four women and one man. The time lapsed between the discovery of the lesions and consultation is usually long. In observations 1, 2, and 4, the delay was, respectively, 9 months, 5 years, and a few months. This slow progression is not a criterion indicating benign lesions because these tumors usually progress in this way [5].

CS is the most often symptomatic and the major symptom is pain, as was the case in four of our five patients. Swelling is also a frequent occurrence when the condition is discovered, observed in two of our patients. The pain involved and the adherence to deep structures suggests malignity. A pathological rib fracture, hemithorax, or metastasis can bring out the diagnosis. In more than 90% of cases, CS appears spontaneously de novo, with no preexisting lesion. In rare cases, these lesions develop on a preexisting benign lesion, such as a chondroma or an osteochondroma, or in Paget disease. External thoracic irradiation and injuries can play an etiopathogenic role in the origins of CS [6].

Biological exams contribute little useful information. Imaging plays an important role in the diagnosis and management of chest wall tumors. The chest X-ray can orient the diagnosis of costal CS by showing a seemingly parietal opacity, a costal bone bulge, lumpy calcifications that are irregularly shaped and distributed, and a rib fracture [7]. Thoracic CT and MRI provide a better definition of the tumor’s seat and extension. CT demonstrates a well-limited, hypodense, rounded or multilobated parietal mass, with clumps of calcifications, which are sometimes nodular, predominating at the tumor’s periphery. Arc-shaped or ring-shaped calcifications can also be found, as illustrated by observation 4. These calcifications are not constant. Costal or vertebral fractures as well as infiltration of the surrounding soft tissues can be observed. The thoracic CT scan is generally sufficient to decide on surgical resection. MRI, less specific than CT, shows a parietal mass, often with clear contours presenting an intermediary signal on T1-weighted sequences and a hyperintense signal on T2-weighted sequences. The tumor looks heterogenous because of the signal loss in areas of calcification. MRI provides a better configuration of the cortical destruction and the periosteal reaction [3].

In our series, the radiological and clinical aspect suggested a benign costal tumor in one case and a posterior mediastinal neurologic tumor in another case. The definitive diagnosis can only be established by anatomopathological examinations. Immediate wide resection of the tumor is the best course whenever possible, allowing analysis of the entire lesion and minimizing the risk of recurrence and dissemination [5]. Fine-needle biopsy of the tumor often gives only poorly abundant material that is not highly representative of the tumor [8]. It should only be done in the case where resection is not immediately feasible or at the price of substantial sequelae, as illustrated by observation 1. In these cases, it can eliminate tumors whose treatment is not exclusively surgical, such as Ewing sarcoma [5].

Macroscopically, the tumor is grayish in color, opaque, soft in consistency, with a gelatinous aspect, sometimes pseudocystic. The cartilaginous islets are often voluminous, coalescent, and poorly limited, with at most the loss of the lobulated aspect of cartilaginous tumors. Histologically, CS is a malignant tumor with cartilaginous differentiation with no bone or osteoid production, but tumor-induced or endochondral ossification is observed. According to the World Health Organization (WHO) classification, CS are divided into three grades of increasing aggressivity based on cellular density, the degree of anisokaryosis, and nuclear hyperchromatism [9]:

- grade I represents moderate cellular density. The nuclei are hyperchromatic and uniform. Occasionally, a few binucleation aspects are noted. The cytological aspect is close to that of benign chondroma;
- grade II is characterized by higher cellular density. The nuclei are more atypical and hyperchromatic. The binucleate or multinucleate cells are observed more frequently. Mitoses are rare (Fig. 6);
- grade III is densely cellular. The tumor cells are pleomorphic and highly atypical. Mitoses are frequent (Fig. 7).

CS progresses spontaneously toward locoregional spread and the appearance of primarily pulmonary metastases, as illustrated by observation 1 and the death.

Surgery is the only curative treatment [8], with the quality of the surgery an essential prognostic factor. Resection should be wide, taking the involved rib en bloc as well as the two intercostal spaces above and below, with a safety margin of at least 4 cm, which can be less when there is contact with the vital organs but should also be greater than 2 cm. The fine-needle biopsy puncture trajectory should also be excised. If the neighboring organs such as the lung and the muscles are invaded or adherent to the tumor, they must be resected [5]. Wide excisions raise the problem of wall reconstruction, which should never make the surgical act palliative and should restore the morphology and physiology of the thoracic wall. Many thoracic wall reconstruction
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Figure 6 Grade II CS: mesenchymal tumor with high cellularity in a fibrous and myxoid matrix. The cells show variation in size and shape (HE × 100) (case 1).

Methods exist, using autogenous biological material or a synthetic material such as polypropylene and polytetrafluorethylene [6,10]. Radiotherapy and chemotherapy have a limited effect. They are undertaken in cases of a voluminous tumor that cannot be resected at once in an attempt to reduce tumor size, as illustrated by observation 1. Adjuvant treatments after surgery have not proven to be effective [4]. In observation 5, the place of these adjuvant treatments could not be clearly evaluated because the absence of recurrence and metastasis 4 years after surgery could be explained by the complete excision of the tumor. Treatment of recurrences is also surgical [8].

The prognosis for CS is determined by its degree of histological differentiation and by the quality of the surgical excision. Other prognostic factors have been suggested, such as tumor size and the onset of metastasis [5]. According to some authors, cases of CS of the trunk have poorer prognosis than CS affecting the limbs [11]. This may be explained by the larger tumor size of CS located in the trunk at the time of diagnosis, because their growth often goes unnoticed in the chest wall or in the pelvic cavity, and because it can be difficult to remove the tumor completely with healthy margins in cases of deep CS. Other authors consider that the survival and local recurrence rates of CS located in the chest wall are comparable to the rates of CS located elsewhere, provided that the surgical margins are healthy [12]. Recurrences and metastases are frequent. Generally, recurrences occur within 3 years of surgical resection but can occur after a longer period in 37% of cases. Postoperative clinical follow-up is imperative every 3 months for the first 3 years and every 6 months for at least 10 years, the duration after which a malignant tumor of the chest wall is considered cured [10,7].

Conclusion

Costal CS are rare tumors, characterized by a high potential of locoregional invasion, recurrence after treatment, and metastatic dissemination. Treatment is based essentially on surgery, which can only be curative in tumors with limited extension that are diagnosed early. This warrants surveillance of benign bone lesions as well as rigorous and prolonged follow-up of treated CS so that recurrence or metastasis can be detected early.

References