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Case Report
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A Rare Case of a Mediastinal Tumor, Located Under the Diaphragm

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Abstract

Chondrosarcoma is the most common malignant tumor of the bony thorax and the single most common malignancy of the sternum. Mediastinal chondrosarcoma is exceedingly rare and a cystic variant is an exception. Diagnostic imaging is from utmost importance for early detection, preoperative planning and for local recurrences screening. It is principally resistant to chemo and radiotherapy; therefore, radical surgical resection is the gold standard of treatment. We report a rare case of mediastinal cystic chondrosarcoma, originating from the xiphoid process of the sternum, partially located below the diaphragm, which was successfully operated in our institution.

Keywords: Mediastinal chondrosarcoma; Sternum; Chest wall; Diagnostic imaging; Operative treatment

Introduction

Chondrosarcoma is the third most common primary malignant bone tumor which is most frequently found in pelvis and long bones [1]. Mediastinal chondrosarcoma is exceedingly rare. The aim of the current study is to report a rare case of mediastinal cystic chondrosarcoma, originating from the xiphoid process of the sternum, which was successfully operated.

Case Presentation

We present a case of 45-year-old woman with complaints of pain, localized in the distal part of the sternal body and xiphoid process, dating from one week. Paraclinical studies were all within the normal range. A chest CT scan revealed a low attenuated anterior mediastinal formation (Figures 1a-1c). The lesion was with axial dimension of 40/38 mm and was localized outside the bone and inferior to the diaphragm. The conclusion of the CT scan was for a cystic formation in the anterior inferior mediastinum, that engaged xiphoid process of sternum. Vertical incision was performed above the distal part of the sternum and the proximal upper abdomen (Figures 2a, 2b). A cystic thick-walled lesion, that destructed the xiphoid process was found. A partial distal resection of the sternum together with about 2 cm of adjacent visibly intact part of the diaphragm and the surrounding adipose tissue was performed. The tumor was cystic with peripheral zones of densely-elastic cartilaginous tissue (Figures 3a, 3b). The histology revealed high-differentiated chondrosarcoma. The resection lines of the adjacent tissues were free of the tumor. Radiotherapy was proposed for local recurrence prevention control.

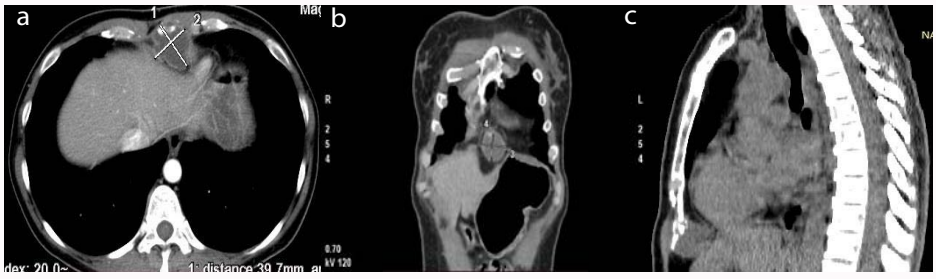

Figure 1(a,b,c): Preoperative CT scan in axial, coronal and sagittal projection.

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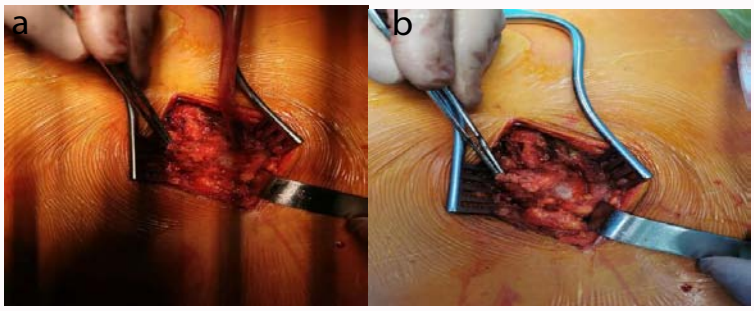

Figure 2(a,b): Intraoperative view.

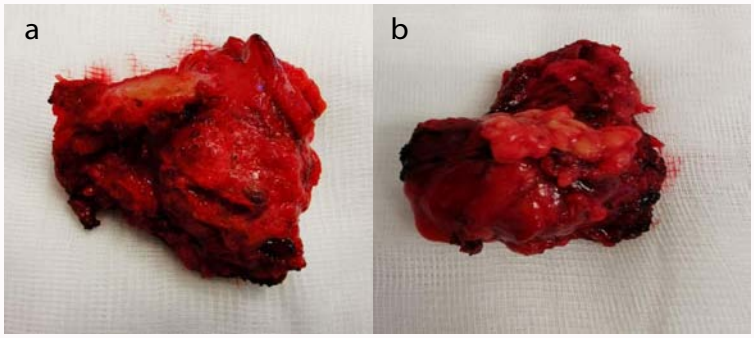

Figure 3(a,b): Macroscopic view of the removed chondrosarcoma.

Discussion

Chondrosarcoma is the most common malignant tumor of the bony thorax and the single most common malignancy of the sternum [2]. It is a rare lesion with an incidence of 0.5 chest 4 wall chondrosarcomas per million people per year [3]. It usually arises primarily within bones formed through endochondral ossification. Secondary chondrosarcoma arises from benign chondroma or osteochondroma. Usually a history of previous trauma, radiation therapy or presence of chromosomal mutations is available. Typical characteristics of chondrosarcoma include slow tumor growth, low-grade disease, extremely low rates of metastasis, and slightly male predominance [4]. Immunohistochemically the tumor shows positivity for SOX4, SOX9 and S100 protein. CT scan demonstrates a low-attenuating rounded mass with "ring and arc" calcification pattern, that is typical for chondroid matrix. Some of the tumors show cortical destruction, periosteal reaction or surrounding soft tissue invasion. Chondrosarcomas are of low to intermediate signal intensity on T1-weighted MR images and hyperintense on T2- weighted MR images. The "pepper surface sign" can be seen in T2WI, which is one of the most important imaging modalities that indicates chondrosarcoma [5]. It is interesting to note that not all malignant chondrosarcomas are PET positive and low-grade tumors are low or marginally PET avid, as it is described in a case by Østergaard et al. [1]. Our study shows that we should be aware that some anterior mediastinal cystic formations could be malignant, like in our case, and consequently an operative treatment needs to be proposed. Chondrosarcoma is chemo- and radioresistant tumor. This is most probably related to the abundance of chondroid matrix, low cellular grade, and relative hypovascularity [6]. R0 surgical resection is considered the gold standard and there is no consensus for the desired surgical margin. However, performance of radical resection is often combined with chest wall instability and therefore reconstructive operation is followed [2,7,8]. We decided to proceed with open approach surgery, because the formation was located on the border of two body cavities. The resultant defects in chest wall and diaphragm were 5 minimal and there was no need for reconstruction. After multi-board discussion radiotherapy was proposed despite radical resection for treating eventual residual tumor cells and for prevention of local recurrence. Surveillance consists of physical examination and thoracic imaging with either PA/lateral radiograph or CT scan every 3 to 6 months for the first 5 years and annually thereafter for a minimum of 10 years [6].

Conclusion

Mediastinal chondrosarcoma with subphrenic distribution is exceedingly rare malignant mesenchymal tumor. Diagnostic imaging is from utmost importance for early detection, preoperative planning and for local recurrence screening. Surgical resection is the gold standard for treatment.

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