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PRIMARY MALIGNANT BONE TUMOR CHONDROSARCOMA OF THE STERNUM

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ABSTRACT

The primary sternal tumor is rare, with its incidence representing 0.6-0.9% of all primary bone tumors. The current study aimed to report a rare case of chondrosarcoma in the body of sternum, which was successfully treated.Chondrosarcoma is a primary malignant bone tumor that is composed of cartilage-producing cells or chondrocytes. It is one of the most challenging bone tumors to diagnose and treat. These cartilage lesions may have inconclusive features, necessitating close follow-up or biopsy. On radiographs, the radiologist should look for aggressive features such as focal erosion with abrupt margins, periosteal reaction or cortical breakthrough. In our case, none of the typical radiographic changes of chondrosarcoma appeared on chest imaging studies, and the diagnosis was based on biopsy. Surgery is the best therapeutic option for a sternal tumor. In some cases, radiation and or chemotherapy may be used pre- or post-operatively. Chondrosarcoma is a malignant cartilage-forming tumor of bone. It is rarely found in the sternum, making the case presented herein rare and the diagnosis to be complicated. Multidisciplinary management of this case is necessary to make diagnosis and treatment.
Keywords: *Chondrosarcoma, Sternum, Sternal tumor, rare bone tumor, Primary malignant bone tumor, Tumor diagnostic, Tumor histopathology*

ABSTRACT

Tumor sternum primer jarang terjadi, dengan insidensinya sekitar 0,6-0,9% dari semua tumor tulang primer. Artikel ini bertujuan untuk menunjukkan kasus chondrosarcoma yang langka di badan sternum, yang berhasil ditangani.Chondrosarcoma adalah tumor tulang ganas primer yang terdiri dari sel-sel penghasil kartilago atau kondrosit. Ini adalah salah satu tumor tulang yang paling sulit untuk didiagnosis dan diobati. Lesi tulang rawan ini mungkin memiliki penampakan yang tidak jelas, sehingga seringkali diperlukan biopsi dan pengawasan berkala yang ketat. Pada radiografi, ahli radiologi harus mencari fitur agresif seperti erosi fokus dengan tepi yang tidak jelas, reaksi periosteal atau kortikal. Dalam kasus kami, tidak ada perubahan radiografi khas dari chondrosarcoma yang muncul pada hasil dari pencitraan dada, dan diagnosis didasarkan pada biopsi. Pembedahan adalah pilihan terapi terbaik untuk tumor sternum. Dalam beberapa kasus, radiasi dan atau kemoterapi dapat digunakan sebelum atau sesudah operasi.Chondrosarcoma adalah tumor pembentuk tulang rawan ganas. Jarang ditemukan di sternum, membuat kasus yang disajikan di sini jarang dan diagnosis menjadi rumit. Manajemen multidisiplin dari kasus ini diperlukan untuk membuat diagnosis dan perawatan.

Kata kunci: Chondrosarcoma, Sternum, Tumor sternum, Tumor tulang langka, Tumor tulang ganas primer, Tumor diagnostik, Tumor histopatologi

INTRODUCTION

Chondrosarcoma is a malignant tumor composed of cartilage-producing cells. Chondrosarcomas account for 3.5-9% of primary bone tumor and approximately 30% of primary bone malignancies. They occur in the 4th to 7th decade of life and are slightly more common in men. The most common locations are the pelvis, ribs, proximal femur and proximal humerus. Only 1% of primary bone cancers arise in the sternum [1,2,3].

Tumors of the sternum are rare. They can be primary, metastatic or arise from adjacent tissues. A careful radiological investigation is necessary because of the mass evident on examination is often part of a much larger tumor invading the sternum. Most neoplasms of the sternum are metastatic lesions. Histologically they can be malignant, benign, or inflammatory [2,4].

The primary sternal tumor is a rare type of bone and soft tissue tumor. Incidence of sternal tumors represent 0.6-0.9% of all primary bone tumor. Chondrosarcoma is one of the most common types of primary sternal tumor, followed by osteosarcoma, myeloma and malignant lymphoma, [4,5] while some differential diagnosis of benign tumors of the chest wall are enchondroma, chondroma, osteochondroma, osteoid osteoma, and fibrous dysplasia [6]. Table below represents the differential diagnosis of tumors of the bony chest wall.

Table 1. Classification of tumors of the bony chest wall [6].

Tissue	*Benign Tumors*	*Malignant Tumors*
Cartilage	Enchondroma Chondroma Periostial chondroma Osteochondroma	Chondrosarcoma
Bone	Osteoid osteoma	Osteosarcoma
Fibrous	Fibrous dysplasia	Malignant fibrous histiocytoma
Vascular	Hemangioma	Hemangiosarcoma
Marrow	Eosinophilic granuloma	Plasmacytoma
Osteoclast	Giant cell tumor Aneurysmal bone cyst	
Unknown		Ewing's sarcoma

CASE REPORT

A 42-year-old female presented to our Orthopaedic Clinic RSUP Sanglah Denpasar with major complaints of lump and pain in the anterior chest wall since March 2018. On physical examination, a palpable mass was identified on the inferior part of manubrium sternum, which was tender on palpation. Laboratory test showed no abnormalities. From the chest x-ray, there was a blastic and sclerotic lesion with sternum enlargement, which led to the suspicion of metastatic bone disease.

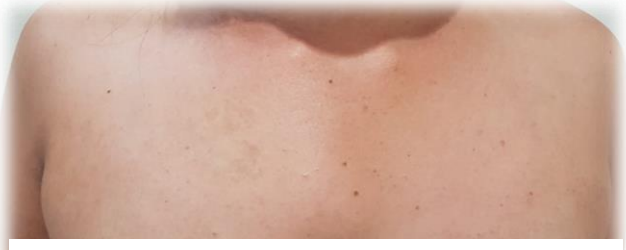

Figure 1. Clinical Picture of the Sternal Mass

Chest CT-scan demonstrated osteoblastic, osteolytic and destruction of the sternum with normal manubrium. No periosteal reaction with minimal soft tissue mass also led to the suspicion of a mixed type of sternal metastatic bone disease. However, the bone survey showed no lesions other than the one in the sternum

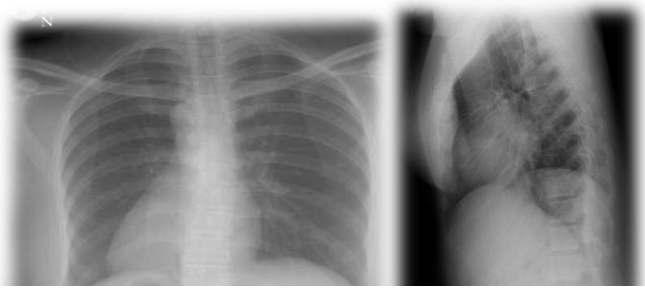

Figure 2. The Chest X Ray from AP view (A) and lateral view (B) showing sternum enlargement with minimal soft tissue mass

Therefore, we performed an open biopsy, and histological analysis of the surgical specimen revealed malignant cartilaginous cell with a variation of size and shape. The tumor cells which located in the medullary part of the sternum caused bone expansion and destruction of the cortex. From all of the examinations we had done, chondrosarcoma of the sternum remained the primary suspect of the patient's complain. Even though chondrosarcomas rarely occur in the sternum. The current study aimed to report a rare case of chondrosarcoma in the body of the sternum, which was successfully treated.

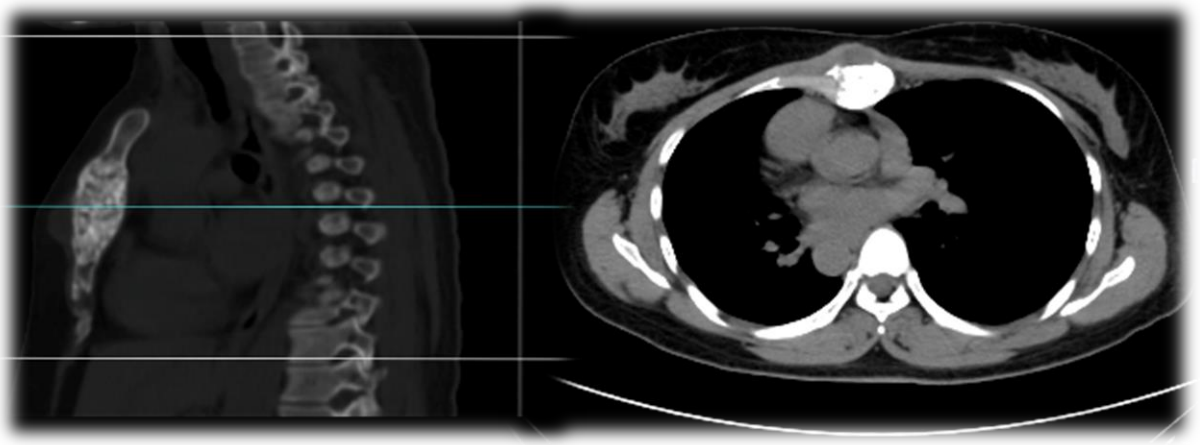

Figure 3. Computed tomography of sagittal section (A) and axial view (B) demonstrating the tumor of sternum with blastic and sclerotic lesions

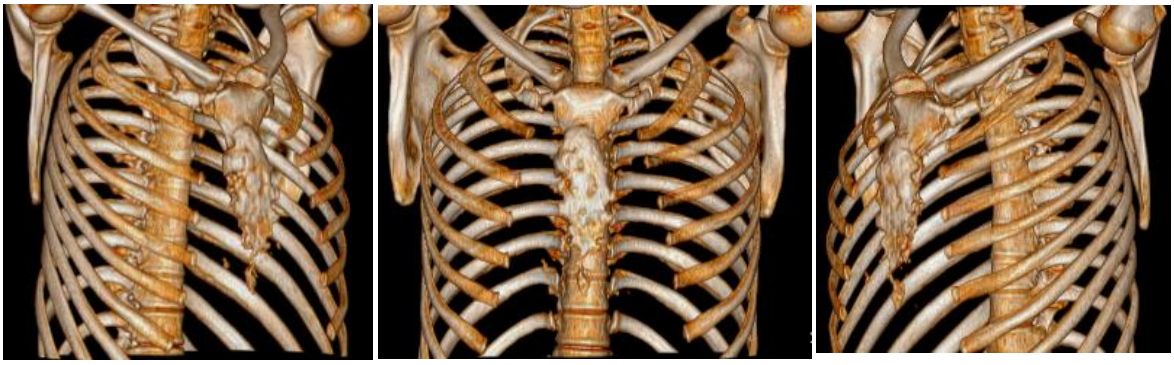

Figure 4. 3D Reconstruction demonstrating the sternal tumor

DISCUSSION

The current study reports a case of an extremely rare tumor that presented in an uncommon location and was successfully treated. Chondrosarcoma is a malignant tumor composed of cartilage-producing cells or chondrocytes. It

is one of the most challenging bone tumors to diagnose and treat. Most common locations are pelvis, proximal femur, distal femur and scapula. In this case presented the chondrosarcoma in a unique location: sternum.

Chondrosarcoma is one of the malignant transformations of osteochondroma and enchondroma. Chondrosarcoma shows a broad spectrum of appearances on imaging depending on the grade of underlying tumor and duration of the disease. Imaging interpretation can be complicated and fraught with subjectivity. Often given cartilage lesion may have inconclusive features necessitating close follow up or biopsy. On radiographs, the radiologist should look for aggressive features such as focal erosion with abrupt margins, periosteal reaction or cortical breakthrough. The decisions to biopsy a cartilage lesion can be difficult due to the broad spectrum of appearances for cartilage tumor [1,2,4].

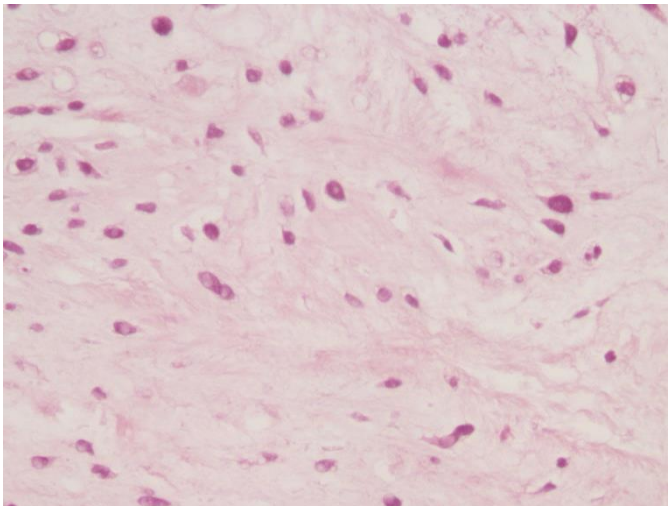

Figure 5. Histopathology examination with Hematoxylin Eosin staining under 100x magnification

The incidence of the sternal tumor has not been established. Primary sternal tumor is uncommon and account for only 1% of primary bone neoplasm worldwide. Chondrosarcoma is the most common tumor of the sternum, followed by osteosarcoma, myeloma and malignant lymphoma. Among benign chondrosarcomas, osteoblastoma and haemangioma have been described. Two thirds of the sternal tumor are metastatic or locally invasive. Most frequently, there is breast cancer invasion, lung or pleural malignancy invasion or rarely solitary metastasis from the kidney or thyroid. The diagnosis was obtained by surgical biopsy. However, even surgical biopsies may be uncertain when the cortex of the sternum is not involved [2,4,5].

Diagnosis of Chondrosarcoma often requires correlation among clinical, imaging and histologic features, and histologic grading is necessary to determine optimal management. Radiographic findings in the sternal abnormalities are often nonspecific, showing appearances from a localized benign lesion to an aggressive lesion as seen with infections and malignant neoplasms.[7] From plain radiography, a lobulated mass arising in the medullary portion of the rib or sternum can be seen. Typically, the cortical bone was destroyed, and the margins of the tumor were poorly defined. Mineralization of the tumor matrix was common and produced a mottled type of calcification.[8] However, because conventional radiographic findings are not reliably accurate for diagnosis and staging this tumor, a specific diagnosis of sternal abnormalities can be suggested based on CT and MR characteristics.[2,5,7] Radiographic evaluation includes entire bone radiography. CT is more sensitive for detecting matrix mineralizarion, provides better visualization of the chondroid matrix and can delineate both the intraosseous and extraosseus components of chondrosarcoma. MRI can be particularly useful for demonstrating features that help distuingish a chondrosarcoma from a benign chondroid lesion. Positron-emission tomography can distuingish benign from malignant neoplasm, but it cannot differentiate tumor types. Because of the hypovascularity and slow rate of mitosis in chondrosarcoma, they are resistant to chemotherapy and radiation therapy [2,5].

From this case, we reported the chondrosarcoma stage II that occurred in the inferior manubrium sternum. On the X-Ray and the CT scan, the diagnose of Chondrosarcoma is still cannot clear. After an open biopsy and do the histopathology examination, we can see the structure of the cell is Chondrosarcoma Stage II. Clinico Pathological Conference (CPC) was held to confirm the diagnose and planning the definitive treatment. From the literature review, the gold-standard treatment for this condition is the surgical resection of the lesion, the only curative option, due to the therapeutic resistance to chemotherapy and radiotherapy. The sternal tumors present themselves as an excellent therapeutic challenge on account of the local aggressiveness of these tumors and their high rate of recurrence, making it difficult to the lesion resection without compromising the stability and reconstruction of the chest wall.[3] For our patient here, curettage and bone cement were performed, and the

complaint is well treated. However, chondrosarcomas of thoracic wall typically relapse locally, and if not treated well, it could evolve with metastases. Therefore, the long term outcome of this patient requires further investigations.

CONCLUSION

Chondrosarcoma is a malignant cartilage-forming tumor of bone. It is rarely found in the sternum, making the case presented herein rare and the diagnosis to be complex. Diagnosis of Chondrosarcoma often requires correlation among clinical, imaging and histologic features. Meanwhile, histologic grading is necessary to determine optimal management.[1,4,5] Clinico Pathological Conference (CPC) was held to confirm the diagnosis and to plan the definitive treatment.

REFERENCES

1. Kozak K, Łochowski MP, Białas A, et al. Surgical treatment of tumours of the sternum – 10 years' experience. *Polish J Cardio-Thoracic Surg* 2016; 3: 213–216.
2. Yousefi Y, Sadrizadeh A, Sadrizadeh S. Huge sternal chondrosarcoma: a case report. *Asian Cardiovasc Thorac Ann*. Epub ahead of print 2018. DOI: 10.1177/0218492318803813.
3. Ji F, Fracasso JI, Macedo GB De, et al. American Chondrosarcoma of the Sternum Treated with En Bloc Resection : A Case Report. 2018; 6: 1–6.
4. He B, Huang Y, Li P, et al. A rare case of primary chondrosarcoma arising from the sternum: A case report. *Oncol Lett* 2014; 8: 2233–2236.
5. Le H V, Wadhwa R, Theodore P, et al. Excision of Thoracic Chondrosarcoma: Case Report and Review of Literature. *Cureus* 2016; 8: e708.
6. Somers J, Faber LP. Chondroma and chondrosarcoma. *Semin Thorac Cardiovasc Surg* 1999; 11: 270–277.
7. Marulli G, Hamad AM, Cogliati E, et al. Allograft sternochondral replacement after resection of large sternal chondrosarcoma. *J Thorac Cardiovasc Surg* 2010; 139: e69–e70.
8. Unni KK, Bergstralh EJ, Payne WS, et al. Chondrosarcoma of the Chest Wall: Factors Affecting Survival. *Ann Thorac Surg* 2010; 40: 535–541.