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Chondrosarcoma Arising in the Posterior Part of a Rib with Dumb-Bell Shaped Extension to the Adjacent Intervertebral Foramen;

A Rare and Unusual Presentation

Abstract: Chondrosarcomas of thorax are unusual tumors categorized as axial malignancies that also invade and destroy the adjacent bone. These tumors are known to displace soft tissue structures coming in their way without invasion. They destroy adjacent organs violently. In this report, we present a 44-year-old man with a large, expansive mass in the posterior mediastinum invading the spinal canal and epidural space through an intervertebral foramen and displacing the thecal sac. To our knowledge, this is the second case in the literature describing a chondrosarcoma of a rib with dumb-bell-shaped extension to the adjacent intervertebral foramen.

Keywords: chondrosarcoma, rib, dumb-bell shaped

Introduction

Chondrosarcomas are malignant cartilaginous tumors of usually poor prognosis, that represent the second most common primary malignant spindle cell tumors of bone after osteosarcomas.[1,2] Chondrosarcomas form a heterogeneous group of tumors whose basic neoplastic tissue is cartilaginous without evidence of direct osteoid formation.[3] The most common sites are the pelvis (31%), femur (21%), and shoulder girdle (13%). In the thorax, chondrosarcomas are the most common malignant tumors of the sternum and scapula [4,5] and mostly located anteriorly within the chest wall, arising from the sternum or costochondral arches.[6] Occasionally, tumors arising from the ribs manifest as intrathoracic masses with only minimal osseous involvement.[7] The classic chondrosarcomas are peripheral (arising from an osteochondroma) or central (arising from the surface of a bone).[1,8,9]

The clinical presentation varies from no symptoms to referral pain, local pain or symptoms due to invasion of adjacent organs. Most chest wall chondrosarcomas (80%) manifest clinically as masses that can be painless or painful.[10]

Case Presentation

A 44-year-old man was referred to our institute because of a large destructive mass lesion in the posterior aspect of his right hemithorax detected on CT scan. His first symptom was pain in the right chest wall since 4 months prior to admission. Physical and neurologic examinations revealed normal. Patient had a medical history of delusional disorders and had been taking clonazepam, respiridone and trihexiphenidyl since 10 years ago. No history of malignancy was found in his family. Routine laboratory test results were normal. Conventional chest radiography revealed a large posterior basilar lung mass on the right side with foci of dense calcifications and adjacent rib destruction. (Figure 1) Right hemidiaphragm was elevated. On CT scan, a mixed-density mass with a lobulated margin and internal calcification was noted.

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The costovertebral junction was destroyed and the tumor had passed through the neural foramen into the spinal canal. (Figure 2) The tumor caused cord compression and erosive widening of the right T8-T9 neural foramens producing a dumb-bell shaped mass. At this stage, our first differential diagnosis was a bone tumor; hence, CT-guided fine needle biopsy was carried out. However, pathological examination of the tissue specimen was unremarkable. On MRI, a large, well defined, heterogeneous mass with low signal intensity on T1 and high signal intensity on T2 was seen in the right thoracic paraspinal region (Figures 3A and 3B). The tumor extended to both the nearby soft tissue and neural foramens. Proximal rib destruction with distal bone marrow edema could be seen. There was no evidence of either sclerotic rim or periosteal new bone formation. Tumor showed enhancement after administration of contrast medium on MR imaging. Finally, the patient had a surgery. A large tumor at the inferior portion of the right hemithorax arising from the posterior portion of the 8th rib was seen. Tumor was situated over the diaphragm; however, it could be isolated easily from underlying diaphragm and the lung without resistance. Destruction of three ribs and some minimal destruction of the 8th thoracic vertebral body were noted. Tumor passing through the neural foramen had produced a dumb-bell shaped mass. Posterolateral thoracotomy was performed along with resection of the chest wall with removal of three ribs (7th, 8th, and 9th) and half of the 8th vertebral body was carried out. The tumor had apparently invaded the spinal canal and epidural space.

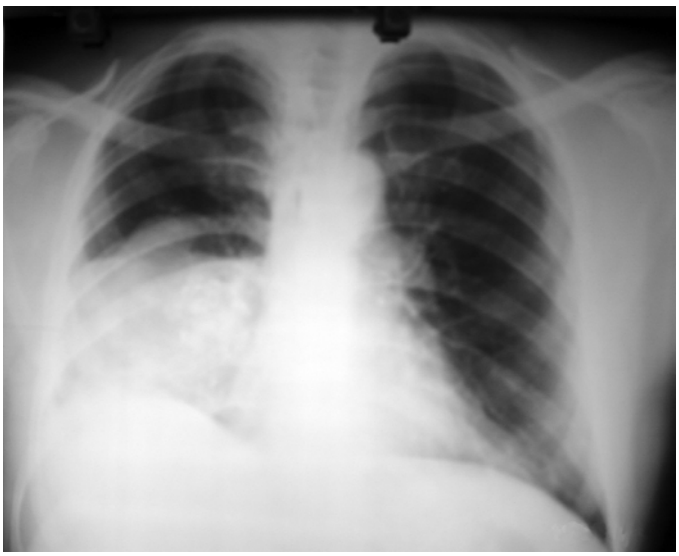


Figure 1: Large Posterior basilar lung mass consolidation

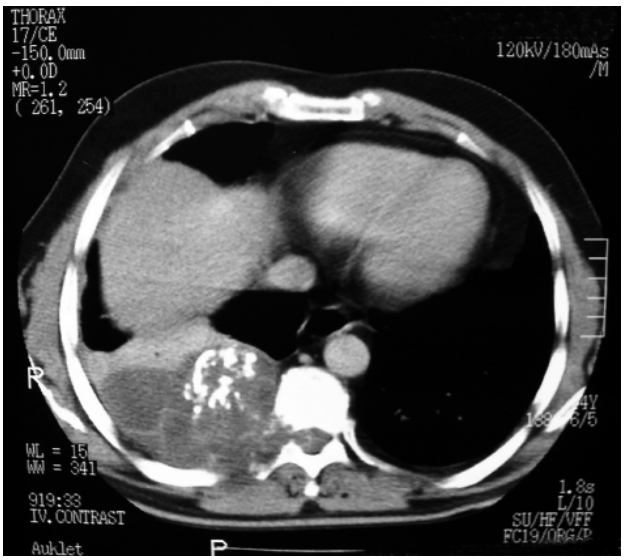
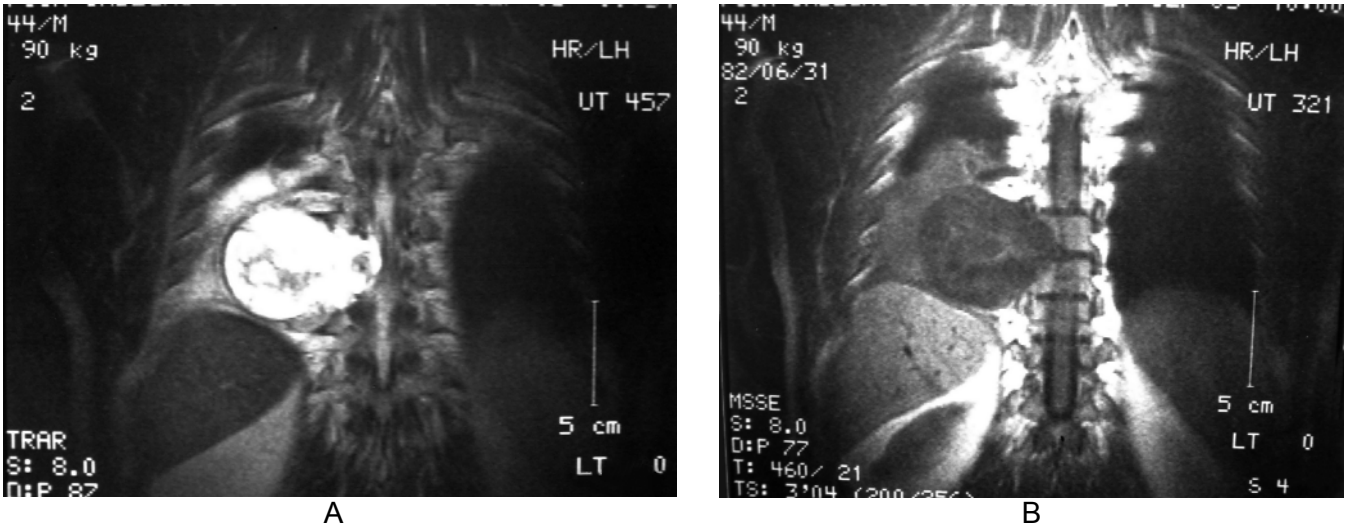


Figure 2: Lobulated margin, mixed density mass with internal calcification



A B

Figure 3: large well defined border, heterogeneous low signal intensity mass on T1 (A) and high signal intensity on T2 (B) was seen in right paraspinal portion of thorax.

Accordingly, compression of the spinal cord and involvement of the posterior longitudinal ligament were seen and the neurosurgeon decided to resect it since it was possible. Pathological diagnosis revealed grade 1, well-differentiated chondrosarcoma. The Patient was discharged from hospital 10 days after surgery without any complication

Discussion

To our knowledge, there are very rare reports of dumb-bell shaped chondrosarcoma of the rib in the posterior mediastinum.[11] The article reporting that one is in Japanese, so until now there has been no report of this type of tumor in English literature. Usually, chondrosarcomas have a specific behavior. They usually invade and destroy adjacent hard tissues such as ribs and vertebrae, and involve and envelop neighboring soft tissues. In 1998, Dr Noirhomme presented a case of thoracic chondrosarcoma which had invaded adjacent ribs and vertebral bodies and surrounded thoracic aorta in a horseshoe fashion. The esophagus and lung were intact.[12] In 1999, York et al published their findings in 21 patients with spinal chondrosarcoma.[13] In that article, there was a photograph of a pathological specimen showing thoracic chondrosarcoma invading and destroying adjacent rib and vertebral body and extending to epidural and paraspinal spaces. Although they did not explain this figure, it is clear that the tumor had first destroyed the vertebral bodies and then invaded the epidural spaces. Another example of such finding is a presentation from Yunten et al in 1997, representing a case of cervical dumb-bell shaped chondrosarcoma.[14] Their case had presented with a low-grade spinal chondrosarcoma developing from a preexisting osteochondroma in the neural arch of C6. Zibis et al in 2000 reported a similar case of chondrosarcoma passing through cervical neural foramen.[15]

Radiologically, there is some trouble in diagnosing this type of tumors. Various neoplastic, vascular, and developmental causes may lead to spinal neural foraminal widening, the most common of which being schwannomas and neurofibromas. Other slow-growing tumors such as chordomas, ependymomas, and meningiomas may occasionally cause neural foraminal widening.[16] Sometimes, there are certain radiological clues of the slowly growing osteochondroma such as some shapes (cauliflower-like), or calcification patterns. However, tumor enhancement after administration of contrast medium on MR imaging raise the possibility of a sarcomatous lesion. Chondrosarcomas of thorax usually manifest as large chest wall masses with bone destruction and soft tissue extension. Central chondrosarcomas have two distinct patterns.[17] One is a small well-defined lytic lesion with a narrow zone of transition and surrounding sclerosis with faint calcification. This is the most common malignant bone tumor that may appear radiographically benign. The second type has no sclerotic border and is difficult to localize. The signs of malignancy are endosteal scalloping, expansion with widening of the diameter of bone, followed by cortical destruction and soft tissue extension. Peripheral chondrosarcoma is easily recognized as a large, calcified mass protruding from a bone.[18]

Scattered areas of calcification may occur in the chondroid matrix and are optimally detected with CT. CT is more effective in detecting and characterizing the pattern of mineralization, cortical destruction and soft tissue extension.[19] MR imaging may show the presence or absence of bone marrow edema and typically demonstrates heterogeneous internal architecture.[11] The masses are of intermediate or low signal intensity on T1-weighted MR images and are heterogeneous on T2-weighted ones, typically with scattered areas of high signal intensity.[20] Nevertheless, radiological and histological findings may be indistinguishable or only slightly different from those of benign lesions, leading all too often to underdiagnosis and inadequate treatment. Aggressive surgical resection is the treatment of choice for these tumors.[12] Chondrosarcoma readily implants into soft tissues, with such recurrences survival is dramatically reduced. Thus, resection must be radical, including a wide margin of normal tissue. Therefore, MRI is the method of choice because it can provide the appropriate preoperative estimation for a successful resection.[15] Gross-total resection of the chondrosarcoma provides the best chance for prolonging the disease-free interval.[19]

References

1. Dahlin DC. Bone tumors: General aspects and data on 6221 cases, 3rd ed. Illinois, Springfield: Charles C Thomas Publisher, 1978, 11.
2. Lee FY, Mankin HJ, Fondren G ,Gebhardt MC, et al. Outcome analysis of chondrosarcoma. J Bone and Joint Surgery (Am) 1999; 81A: 326-338.
3. Marcove RC. The surgery of tumors of bone and cartilage, 2nd Ed. New York: Grune and Stratton, 1984.
4. Sanerkin NG. The diagnosis and grading of chondrosarcoma of bone. A combined cytologic and histologic approach. Cancer 1980; 45: 582.
5. Huvos AG. Bone tumors. Diagnosis, treatment and prognosis. Philadelphia: WB Saunders, 1979: 358-370.
6. Somers J, Faber LP. Chondroma and chondrosarcoma. Semin Thorac Cardiovasc Surg 1999; 11:270-277.
7. Gladish GW, Sabloff BM, Munden RF, et al. Primary thoracic sarcomas. RadioGraphics 2002; 22: 621-637.
8. Jaffe HL. Tumors and tumorous conditions of the bone and joints. Philadelphia: Lea Febiger, 1958: 117-141.
9. Shives TS, Wold LE, Dahlin DC, Beaubout JW. Chondrosarcoma and its variants. In: Sim FH, ed. Diagnosis and treatment of bone tumors a team approach. Mayo Clinic Monograph. Thorofare, NJ: Slack Inc., 1983:211.

10. Burt M. Primary malignant tumors of the chest wall: the Memorial Sloan-Kettering Cancer Center experience. Chest Surg Clin N Am 1994; 4: 137-154.
11. Horio H, Nomori H. A case of dumb-bell shaped chondrosarcoma of the rib in the posterior mediastinum. Nippon Kyobu Geka Gakkai Zasshi 1996; 44: 519-23.
12. Noirhomme P, D'udekem Y, Munting E, El Khoury G, Humblet Y, Dion RA. Resection of a Chest Chondrosarcoma Invading the Spine and the Aorta. Ann Thorac Surg 1998; 65:534–5.
13. York JE, Berk RH, Fuller GN, Rao JS, Abi Said D, Wildrick DM, Gokaslan Z. Chondrosarcoma of the spine: 1954 to 1997. J Neurosurg (Spine 1) 1999; 90: 73–78.
14. Yunten N, Calli C, Zileli M, Ustun EE, Sener RN. Chondrosarcoma causing cervical neural foramen widening. Eur Radiol 1997; 7: 1028-1030.
15. Zibis A.H, Markonis A, Karantanas AH. Unusual causes of spinal foraminal widening. Eur Radiol 2000; 10: 144-148.
16. Osborn AG. Diagnostic neuroradiology. St. Louis: Mosby, 1997: 876–918.
17. Edeiken J. Bone tumors and tumor-like conditions. In: Edeiken J, ed. Roentgen diagnosis and disease of bone, 3rd ed. Baltimore: Williams & Wilkins, 1981: 30.
18. DeVita VT Jr. Cancer: Principles and practice of Oncology, 6th ed. Baltimore: Lippincott Williams and Wilkins; 2001.
19. Stelzer P, Gay WA Jr. Tumors of the chest wall. Surg Clin North Am 1980; 60: 779-791.
20. Collins MS, Inwards CY, Koyama T, Swee RG. Clear cell chondrosarcoma: radiographic, computed tomographic, and magnetic resonance findings in 34 patients with pathologic correlation. Skeletal Radiol 2003; 32: 687–694.
21. Marcove RC, Huvos AG. Cartilaginous tumors of the ribs. Cancer 1971; 27: 794-801.
22. Gitellis S, Bertoni F, Chieti PP, et al. Chondrosarcoma of bone. J Bone Joint Surg (Am) 1981; 63: 1248.
23. Pritchard DJ, Lunke RJ, Taylor WF, et al. Chondrosarcoma: a clinicopathologic statistical analysis. Cancer 1980; 45: 149.