Chondrosarcoma of right 1st rib presenting as neurogenic thoracic outlet syndrome; A 13th case report in world literature

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Abstract Thoracic outlet syndrome [TOS] caused by a tumor of the rib is rare and has been reported only 12 times in the literature over the past one and one-half centuries, with the majority of cases due to osteochondroma. We report a case of chondrosarcoma of right 1st rib causing neurogenic TOS that was resected via posterolateral thoracotomy and biopsy confirmed a grade I chondrosarcoma. In the treatment of chondrosarcoma, chemotherapy and radiotherapy are less effective, and appropriate surgery is needed.

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Introduction

Thoracic outlet syndrome [TOS] refers to symptoms that arise from the compression of the subclavian artery, vein, or brachial plexus as they traverse the thoracic outlet [1]. Tumors of the chest wall are uncommon, and those causing TOS are even more so. Chondrosarcomas often arise in the pelvis or bones of the trunk, but primary chest wall chondrosarcomas are relatively rare. We report the thirteenth case to our knowledge of tumor of the first rib causing TOS and describe a successful thoracotomy approach for rib resection with resolution of neurologic symptoms.

Case report

A 55 year old man came to our hospital with complaint of progressively increasing constant pain and paraesthesias in right arm and hand for the last 3 months. He also had diffuse discomfort in the right side of neck and anterior chest wall. Physical examination revealed a fixed, nontender, palpable mass in the right infraclavicular fossa. Adson’s test results were negative, and peripheral pulse examination results were normal. Laboratory studies were unremarkable. Respiratory function testing was within normal limits for his age. A chest X-ray revealed a lesion measuring approximately 6 cm on the anterior chest wall side of the right first rib (Fig. 1), and a chest CT showed a lobulated tumor arising from right first rib showing coarse matrix calcification with associated enhancing soft tissue protruding into the thoracic cavity (Figs. 2 and 3).
Core-needle biopsy [CNB] under CT guidance diagnosed the tumor to be grade I chondrosarcoma. Patient was taken up for right posterolateral thoracotomy through 4th intercostal space. After freeing the tumor (Fig. 4) circumferentially achieving good dissection plane between right subclavian vessels and brachial plexus the first rib along with the tumor was excised in toto. Frozen section of the margins of the tissue revealed no obvious tumor microscopically. Defect was not too large as the clavicle, scapula and remaining ribs were

Figure 1 Chest X-ray showing tumor in right side from first rib.

Figure 2 CT chest demonstrating frontal view of the tumor arising from 1st rib with good plane all around.

Figure 3 CT chest saggital view showing dimensions of the tumor.

Figure 4 Intraoperative photograph of the tumor being removed.
spared of resection, so no need of chest wall reconstruction was there. Examination of the resected specimen showed tumor diameter to be $5 \times 6$ cm, and when sliced, the internal surface was slightly uneven and white, but capsular infiltration into the pleura was absent. On histopathological examination, increased tumor cell cellularity and tumor cell nucleus irregularity in both size and morphology occurred, and enlarged images confirmed tumor cell nucleus irregularity and morphology, as well as binucleated tumor cells. S100 antibody was negative. Therefore grade 1 chondrosarcoma was diagnosed. The surgical margin was free of malignant cells, and the patient was given no postoperative adjuvant therapy. The patient now shows no sign of recurrence (1 years and 9 months after surgery).

Discussion

There are three subtypes of TOS, classified on the basis of the particular portion of the neurovascular bundle that is involved [1]. The surgical goal of treatment is decompression of these structures in the cervicoaxillary canal to restore normal neurovascular function to the upper extremity. Neurogenic TOS is the most common subtype, manifesting as neuropraxia of the affected upper extremity from the compression of the brachial plexus between the anterior and middle scalene muscles. Venous TOS leads to effort thrombosis of the axillary or subclavian veins as they are compressed between the anterior scalene and the first rib and clavicle. Arterial TOS, often associated with a cervical rib or other skeletal abnormality, can lead to stenosis, arterial thrombosis, thromboembolism, and upper extremity ischemia. Chondrosarcoma is a malignant tumor exhibiting neoplastic chondrogenesis. It often arises in the pelvis or long bones, and it is relatively rare for chondrosarcoma to arise in the rib. However, of the various primary malignant bone tumors, the incidence of chondrosarcoma is relatively high [2–4]. Prior to therapy, it is important to confirm the pathological diagnosis because treatment differs between benign and malignant tumors and between primary and metastatic tumors; even with chondrosarcoma, the prognosis depends on the grade [5]. The accuracy of fine needle aspiration cytology [FNAC] as a diagnostic method is low, and histological diagnosis by CNB is recommended [6]. As in the present patient, when a tumor protrudes and proliferates into the thoracic cavity, CT-guided biopsy is useful for preventing such complications as pneumothorax. With respect to treatment, chondrosarcoma is not responsive to chemotherapy or radiotherapy, and extensive resection with a sufficient margin (3–4 cm) is considered first-line therapy [7]. King and colleagues [2] compared various resection methods and reported that prognoses were favorable for patients having extensive resection. Depending on the onset location, however, it may be difficult to have a sufficient safety margin. TOS caused by a tumor of the rib is rare and has been reported only 12 times in the literature over the past one and one-half centuries, with the majority of cases due to osteochondroma [8]. A variety of operative approaches have been used, including the interscapulovertebral route for posterior tumors and the thoracotomy, supraclavicular, and transaxillary approaches for anterior tumors.

Conclusion

We reported a case of chondrosarcoma of right first rib causing neurogenic thoracic outlet syndrome which in itself is a very rare occurrence, 13th in the world literature. In such cases fine needle biopsy with CT guidance helps to be very useful in preoperative tissue diagnosis. CT chest should be obtained to look for any tissue plane loss between subclavian vasculature and brachial plexus. Also infiltration into lung tissue should be excluded before planning the surgery. Wide surgical excision is the main stay of the treatment for chondrosarcoma of the rib. Adjuvant therapies are having very minimal role. The case should be followed up very closely for any tumor recurrence.

Conflict of interest

Authors and co authors declare no conflict of interest of any kind to any person, institute, or organisation.

References