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A diagnostic dilemma: A dorsal spine chondromyxoid fibroma turned out to be chondrosarcoma

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Abstract
Introduction: Chondrosarcoma is the most common malignant primary tumor of the chest wall. Only a few cases were reported to arise posteriorly from the paravertebral component.
Case presentation: In this case report, we present a case of chondrosarcoma, a rare bony tumor which was misdiagnosed with chondromyxoid fibroma in 50 years old male patient. The patient presented with left loin pain with no radiation. The patient underwent radiological imaging and computed tomography guided biopsy, results showed chondromyxoid fibroma, which turned out to be chondrosarcoma on post-operative histopathological examination of the excised tumor. The patient underwent surgical excision of the lesion, with improvement of his symptoms.
Conclusion: Despite its rarity, this tumor can be misdiagnosed with chondromyxoid fibroma. Overlapping radiological and histopathological features can make diagnosis difficult with unsatisfactory surgical excision margins. Hence, an extensive pre-operative evaluation and adequate tissue sampling can help avoiding this confusion.

Keywords: chondromyxoid fibroma, chondrosarcoma, rib tumors

Introduction
Chondrosarcoma is the most common malignant primary tumor of the chest wall. It accounts for more than 30% of all primary chest wall tumors, of which 19% arises in the ribs [1, 2]. Chondrosarcoma of the ribs typically originate from the anterior chest wall near the costochondral junction particularly in the superior five ribs, hence presents with an enlarging painful anterior chest mass. On the contrary, only a few cases were reported to arise posteriorly from the paravertebral component [2, 3]. Computed tomography (CT) scan with intravenous contrast is the gold standard for both the diagnosis and operative planning [1]. Given that these tumors are well recognized for their radio-resistant and chemo-resistant characteristics, complete local excision with widely negative margins offers the best therapeutic results [1, 3, 4].
On rare instances, both chondrosarcoma and chondromyxoid fibroma had been confused for each other due to the similarities in the radiological and histological findings [5-7]. In this report, we present a rare case of chondrosarcoma arising from the head of the 10th rib which was initially misinterpreted as chondromyxoid fibroma.

Case Presentation
History and physical examination
A 50-year-old male, not known to have any medical illnesses, with no known history of malignancy presented to our spine clinic complaining of left-sided back pain persistent for the past 6-months. The pain was gradual in onset, progressive in nature with no radiation. The patient denied any history of trauma or other neurological symptoms. Clinical examination revealed no palpable masses. A thorough neurological examination detected no abnormalities.

Imaging studies
Preoperative high-resolution chest CT scan showed an expansile mass at the dorsal aspect of the left 10th rib, measuring approximately 5 x 3.2 cm in axial dimension with associated cortical erosive changes of the corresponding vertebral body. Further evaluation by dorsal and lumbosacral spinal MRI with contrast demonstrated an expansile multiloculated lesion with peripheral enhancement arising from the head of the left 10th rib and causing erosion of the surrounding vertebra (Figure 1).

Histopathology

A CT-scan guided true cut biopsy under sterile technique and general anesthesia was done. The initial histopathological examination revealed osteochondroid and fibrovascular fragments with few foci of spindle cell lesions. The spindle cells were bland with no significant pleomorphism, atypia, or mitotic activity. Scattered bone and chondroid trabeculae were also seen. Immunohistochemical stains revealed only immunoreactivity for Vimentin marker. Based on the histopathological and immunohistochemical findings, a provisional diagnosis of either benign or low-grade spindle cell neoplasm such as chondromyxoid fibroma was proposed.

Management

The patient underwent a left-sided posterolateral thoracotomy performed by a multidisciplinary team consisted of cardiothoracic and spine surgeons. Under C-arm guidance, T10-T12 ribs were identified. The skin incised alongside the course of the 10th rib, extending from posterior to anterior. The ribs were separated by a retractor with direct exposure and visualization of the tumor and the involved surrounding tissue. Intra-operatively, almost all of the tumor tissue was excised with macroscopically negative rib margins. Moreover, the medial end of the 10th rib was excised along with surrounding fibrous reactive tissue. The tumor was excised in fragments due to the anatomical restraint and the critical tumor site, which made complete tumor excision difficult and risky.

The excised specimens were sent for histopathological examination. Grossly, the specimen consisted of whitish multiple fragments, part of which attached to fragments of the 10th rib measuring in aggregate 8x10x2 cm (Figure 2 & 3). Based on the histopathology findings, the tumor was labeled as chondrosarcoma, moderately differentiated, high grade (G2) with invasion to the bone marrow, and the surrounding soft tissue. Mitotic rate was 6/10 high-power fields with both macroscopic and microscopic necrosis of about 40% (Figure 4).

Follow-up

Patient returned back to our spine clinic after 6 weeks of the surgery. The patient reported significant improvement of his back pain. Follow up MRI was done and showed residual tumor at the left exit neural canal of T 10 – 11, measuring 2.8 x 1.5 cm in dimensions compared to 4 x 3 x 5 pre-operatively (Figure 1). From our point of perspective, as patient is currently stable, off symptoms, our plan of management is close observation and follow-up of lesion for progression or reemergence of symptoms.

Discussion

Though primary chest wall bone and cartilaginous tumors are rare, a wide variety of benign and malignant tumors have been reported. The majority are malignant and the most commonly identified subtype is chondrosarcoma [3]. Chondrosarcoma is a rare malignant neoplasm that originates from the cartilage-producing neoplastic mesenchymal cells and appears in both the appendicular and atlantoaxial skeleton [3, 8]. It commonly arises in the shoulder, pelvis, hips, and chest wall. In the ribs, most cases arise in the anterior costochondral junction and only a few cases reported to arise from the posterior costotransverse junction as we report in this case [9, 10]. Table 1 presents previous reported dorsal paravertebral chondrosarcoma cases.

Chondrosarcoma is the third most common primary bone malignancy following multiple myeloma and osteosarcoma [9]. Most cases arise de novo (primary) within the medullary cavity, however, some cases may result from malignant transformation (secondary) of preexisting benign cartilaginous tumors [1]. Chondrosarcoma is more common in the elderly from the sixth decade of life with slight male predominance [11]. A previous history of local trauma or radiotherapy is not uncommon and can be associated with thoracic chondrosarcoma [1, 11], however, in our patient, there was no evidence of such events.

In several previous reports, both chondrosarcoma and chondromyxoid fibroma had been confused for each other due to the similarities in the radiological and histological findings. On CT scan, both tumors are locally aggressive and might demonstrate intra-lesion calcifications, yet, chondrosarcoma is more likely to show calcifications [5]. The presence of intra-lesion calcifications is the radiographic hallmark of chondrosarcoma [7, 12]. In our patient, the absence of calcifications on the initial CT scan contributed to the initial misinterpretation of the tumor as a chondromyxoid fibroma. On histopathology, both tumors can be easily misdiagnosed for each other particularly in the case of limited tissue sampling [12, 13]. Given the critical anatomical location of the tumor in our case, the true cut biopsy resulted in fragmented specimen which made specific characteristics difficult, moreover, the presence of osteochondroid and fibrovascular fragments with the absence of necrosis, atypia, or mitotic activity favored the diagnosis of benign condition over chondrosarcoma.

Complete surgical excision, preferably wide en-bloc excision offers the best therapeutic outcomes [1, 3]. With the pre-operative provisional diagnosis of either benign or low-grade spindle cell neoplasm and the close proximity of the tumor location to the descending aorta, we tried to remove as much tumor tissue and to dissect as far as we can to achieve maximal tumor tissue excision with minimal morbidity to the patient.

Chondrosarcoma of the bone generally has a good prognosis when early diagnosed and appropriately treated by a multidisciplinary team [4]. It is estimated that the 10-year survival rate for patients underwent wide surgical resection about 92% compared to 47% for patients with intralesional resection [9]. The most important factors influencing the survival rate are the grade of the tumor and complete surgical resection [3]. Given the possibility of local and systematic recurrence, patients should undergo routine lifelong surveillance with physical examination and radiological imaging (chest x-ray or CT-scan) every 3-6 months for the first 5 years then annually for at least 10 years [1, 10].

Table 1: Literature review of previous chondrosarcoma cases

In-text Citation	Case Description	Presentation	Location	Management	Outcome	Notes
(Benson, 1955)	Chondromyxoid fibroma of the thoracic spine.	A 34-year-old woman with burning pains in her feet and distal legs of 8 months duration. Associated with weakness, spasticity, involuntary twitching of muscles of the left leg and foot + and right distal leg and foot, spasticity of the muscles of the lower abdomen.	T2 vertebral body	Curettage and postoperative radiation therapy	Improved rapidly. Able to walk long distance within 15 months. Sensory function almost completely returned within 6 months. Deep reflexes remained depressed.	Destruction of spinout processes of T2 and T3. Extension into spinal canal but dura was not involved. Extended superiorly within the canal until C7.
(Ramani, 1974)	Chondromyxoid fibroma of the thoracic spine.	A 44-year-old man with right leg weakness of 2 months duration. Associated with cold right leg below knee, genitourinary complaints, sensory/motor deficits, impotency, inability to ejaculate, hesitancy of micturition, nocturia, incontinence, lack of sensation when bowel was full, fecal incontinence, numbness of the scrotum & penis & buttocks (months before the weakness developed).	T12 – L1 vertebral bodies.	Lumbodorsal laminectomy from T11 to L2.	Patient could pass urine. Muscle power improved. Neurologically normal.	Flattening of the pedicles of T12 and L1 with increase in interpedicular distance and erosion of the undersurface of the right 12th rib (corticated margin). There is also an extraspinal tumor involving the 12th rib.
(Nunez, 1982)	Chondromyxoid fibroma of the thoracic spine.	A 38-year-old woman with midback pain of 2 years radiating bilaterally to the front of the chest. Associated with leg numbness, difficulty walking, trouble initiating urination.	T5 vertebral body.	Anterior transthoracic decompression of T5.	Pain disappeared, Sensory and motor functions recovered rapidly and completely.	Destruction of central portion of the vertebra and pedicles. Compression fracture at T5.
(Tsuchiya, 1992)	Chondromyxoid fibroma of the thoracic spine.	A 19-year-old woman with gait disturbance, paraplegia, and back pain of 2 weeks duration.	T2 vertebral body and neuralarch.	En-bloc total vertebrectomy on T2 following Laminectomies from C7 to T3. Ceramic spacer implantation with methlymethacrylate and Lug segmental sublaminar wiring from C4 to T6.	Patient recovered completely from paraplegia within 1 week.	Cortical expansion of lamina. Anterior and posterior spinal cord compression.
(Bruder, 1999)	Chondromyxoid fibroma of the thoracic spine.	A 27-year-old woman with interscapular pain of 9 months duration and right thoracic radiation only at rest.	T5 vertebral body expand nginto T4.	Posterior left-sided costotransversectomy with tumor resection and instrumented Posterolatera fusion from T2 to T7. After 14 days → anterior vertebrectomy of T4 and T5 by a right side thoracotomy. Anterior vertebral column was reconstructe with autologous rib graft.	Patient recovered.	Osteolysis with wedge deformity of T5. (Left pedicle and body destruction).
(Kikuchi, 2001)	Recurrence of a chondromyxoid fibroma of the thoracic spine 30 years after excision.	Stage 1: An 11-year-old boy with a Chondromyxoid fibroma at T6 (excised). Stage 2: A 41-year-old man with shoulder pain and leg weakness.	Posterior elements of T6 vertebral body.	-	-	Spinal cord was displaced to the right and slightly compressed at T6.
(Shimada, 2013)	Chondromyxoid fibroma of the thoracic spine with aneurysmal bone cyst.	A 25-year-old man with left upper back pain of 1-year duration.	T3 – T4 vertebral bodies.	Cystectomy.	Patient-recovered.	Large aneurysmal bone cyst projecting into the thoracic cavity.

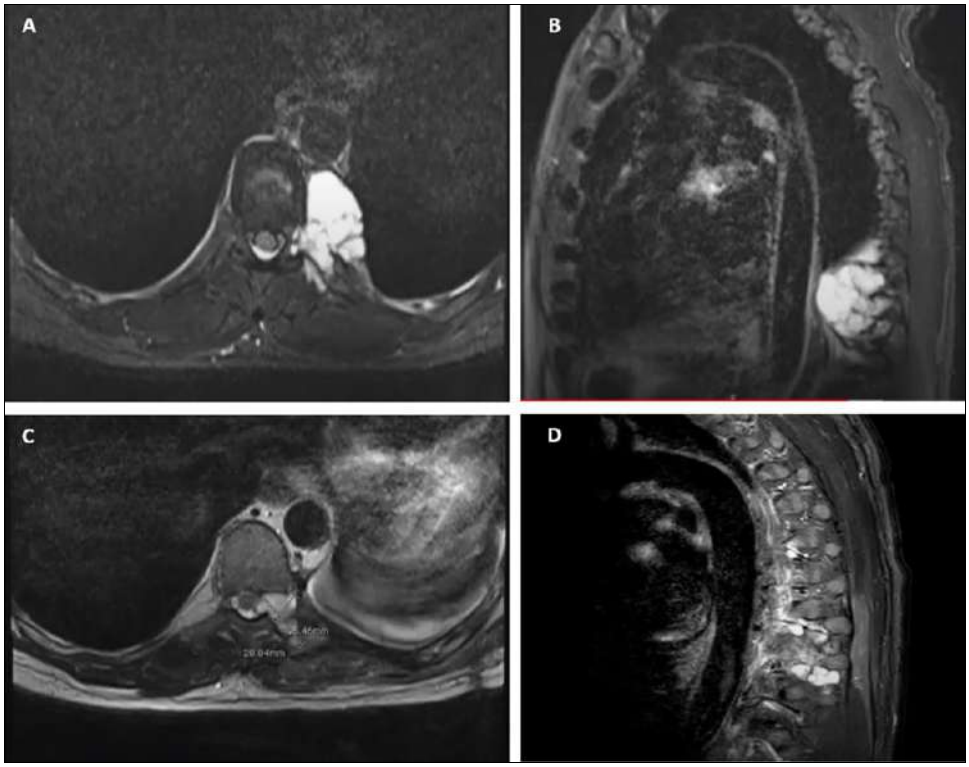

Fig 1: A, B, pre-operative MRI imaging, axial and sagittal respectively. C, D, post-operative MRI imaging, axial and sagittal respectively showing the residual tumor.

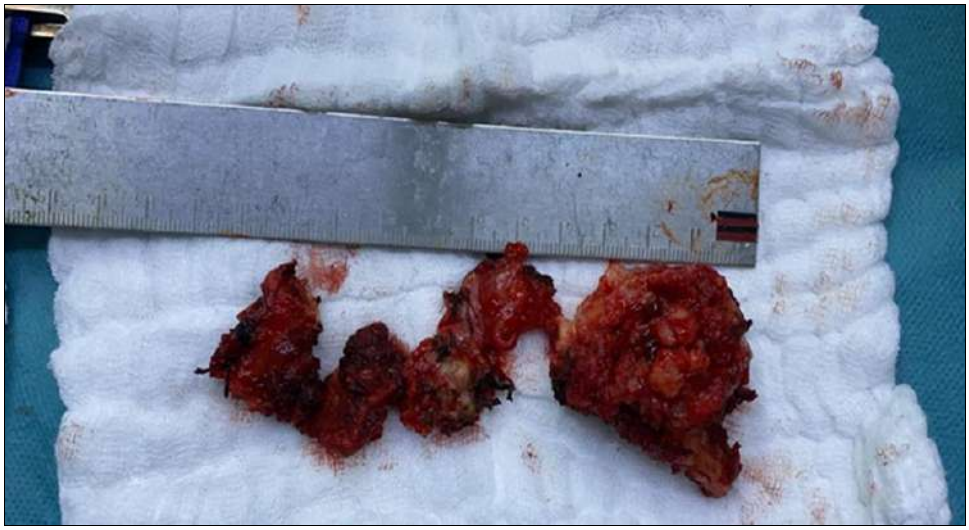

Fig 2: The whole excised surgical specimen in 4 pieces, the most right piece showing parts of the excised 10th rib.

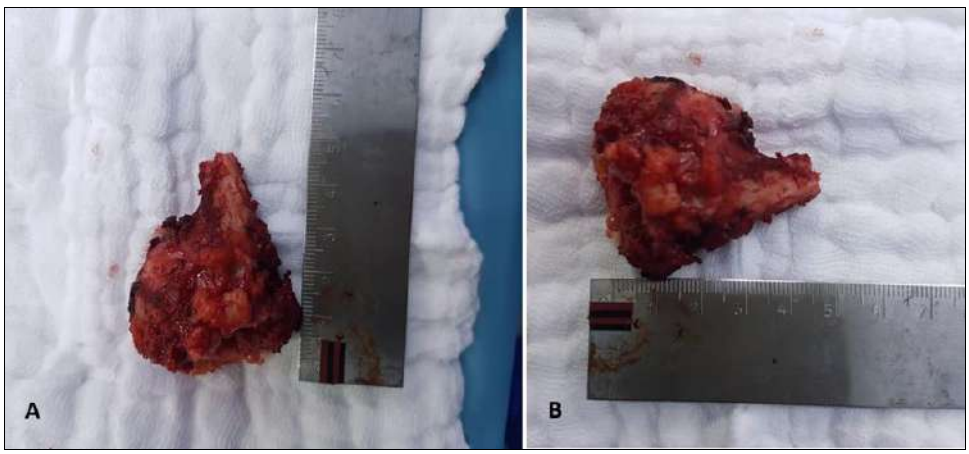

Fig 3: The largest excised fragment with part of the 10th rib, measuring about 5 cm in length.

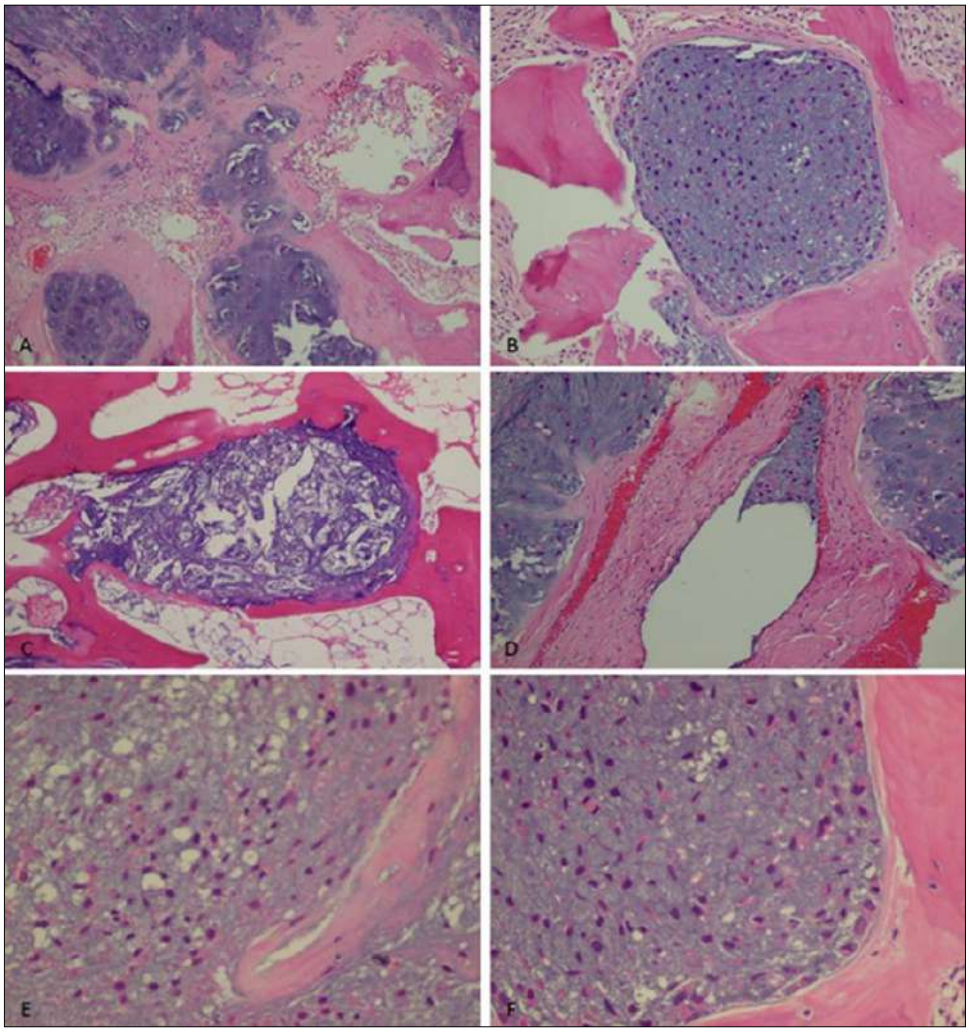

Fig 4: Sections from the mass demonstrate a cellular tumor composed of proliferating atypical chondrocytes embedded in a cartilaginous/myxoid matrix. The tumor cells permeate and entrap bone trabeculae (A,B) and they are seen within bone marrow (C) and the surrounding soft tissue. Lymphovascular invasion is present (D). The tumor cells are pleomorphic with hyperchromasia, nuclear enlargement and mitosis (E,F)

Conclusion

Chondrosarcoma is a rare bone tumor of the skeleton. Unfortunately, this rare tumor can be misdiagnosed with chondromyxoid fibroma. Overlapping radiological and histopathological features can make diagnosis difficult with unsatisfactory surgical excision margins. A thorough pre-operative imaging with adequate tissue sampling can help avoiding this issue.

Clinical Message

Chondrosarcoma and chondromyxoid fibroma have an overlapping histopathological and radiological feature. Orthopedics and spine surgeons should be aware of this diagnostic resemblance for appropriate pre-operative planning, and to ensure adequate tissue sampling in the pre-operative diagnostic phase. Even with the initial presumptive diagnosis of chondromyxoid fibroma. All efforts should be done to excise the entire lesion as much as possible to avoid unanticipated encounter of chondrosarcoma on later histopathological examination.

Consent

The patient included in this study voluntarily agreed on the inclusion of materials in this work by signing a written consent form. The informed consent explained the premise of this report, the state of his anonymity within the report and the confidentiality of his personal information.

Competing interests

The authors declare no conflict of interest.

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