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CASE REPORT

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Chondromyxoid fibroma of the lumbar spine

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Abstract A bulky tumor of the first lumbar vertebra is described. The case is the 21st to be reported. The tumor resembled an aneurysmal bone cyst radiologically. It was resected without incident. The previously reported cases are reviewed and the literature discussed

Key words Chondroyxoid fibroma of spine · Benign cartilage tumor · Spinal tumor · CT of spine

Case report

A young married woman of 19 years of age noted a lump in her lumbar region some 3 years prior to referral to the National Cancer Institute (INCA) in Rio de Janeiro. The lump had been slowly increasing in size, but had not troubed her until the development of pain over the 2 months prior to her referral. The pain had progressively increased in intensity during this perioid. It radiated into her left thigh and ankle and was aggravated by walking and movement of her head. There were no other symptoms. On physical examination she was found to have a large mass, approximately 12 cm in its largest diameter, that was not tender. It was situated in the upper lumbar region in a left paravertebral position. It was of bony consistency, and attempts to move the mass over the underlying tissues elicited pain in her left leg. The patient was in good general health, but had absent patellar and ankle reflexes. No other abnormalities were noted.

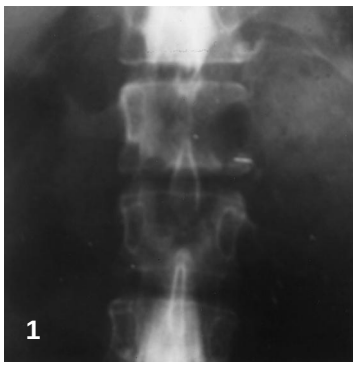

Fig. 1 AP myelogram. Note complete block at the upper end of the L1 vertebra. There is extensive destruction of the left half of L1 including the pedicle and transverse process. Note expansion and erosion of the inferior aspect of the spinous process

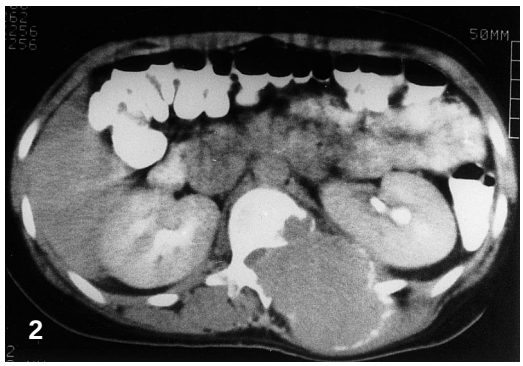

Fig. 2 CT scan (with contrast). Extensive bony destruction with vertebral extent is clearly evident. Note destruction of the spinous process and also the bony fragments around the margin of the mass. The kidney dislocation is clearly visible

She underwent myelography (Fig. 1) and was found to have a complete block at the the upper level of L1. There was destruction of the left half of L1 including the pedicle. A large

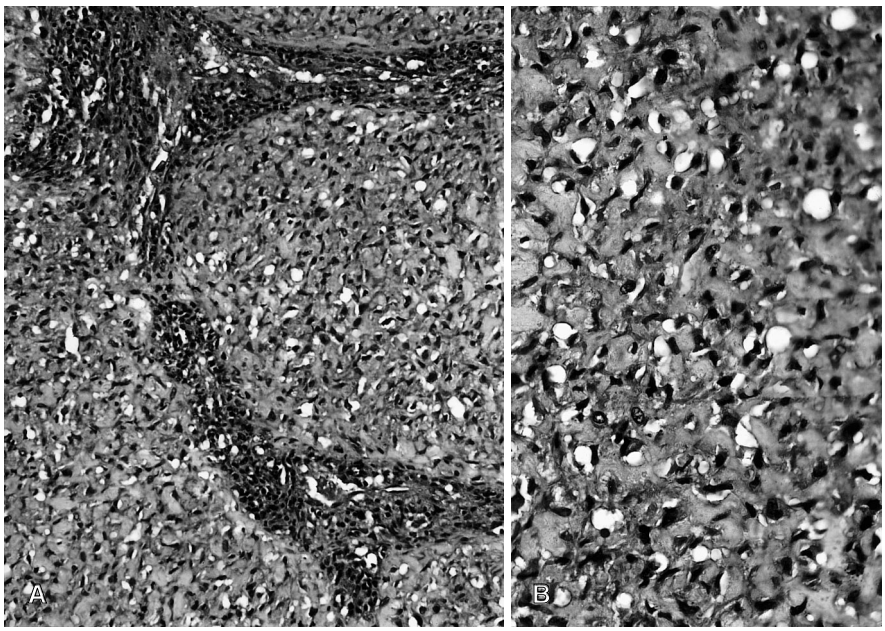

Fig. 3 **A** Photomicrograph taken from the operative specimens demonstrates lobulated structures in which there were rounded cells in a myxoid stroma. The lobules are demarcated by spindle-cell proliferation with osteoclast-like giant cells present (H&E ×50. **B** Elongated and rounded cells with irregular nuclei with fine chromatin and eosinophilic cytoplasm in a rich myxoid background (H&E ×200)

soft tissue mass was also noted. CT with oral and intravenous contrast, as well as without contrast, (Fig. 2) demonstrated a bulky mass destroying the left pedicle and transverse process. It extended into the pararenal space posteriorly and infiltrated the soft tissue in the posterior paravertebral space.

The patient was thought to have a benign tumor involving the vertebral body and posterior elements with a bulky extraosseous extension into the soft tissues. An aneurysmal bone cyst was thought to be more likely than an osteoblastoma. Because of the long history and the radiological appearance, a malignant tumor was thought unlikely. A malignant tumor of cartilage origin was felt to be very unlikely.

The patient had a large tumor removed together with the laminae and pedicles of the left half of L1 and L2. The excision extended from the paravertebral tissues up to the peritoneum, which was found to be intact.

The large specimen, which was removed in portions, showed pinkish-white areas with several areas that appeared to be cartilaginous. There were areas of residual bone. In aggregate the tissue measured 18×15×9 cm.

Microscopically the tissue revealed a lobulated structure in which there were elongated and rounded cells with irregular nuclei, with fine chromatin and eosinophilic cytoplasm lying in a richly myxoid background (Fig. 3). The lobules were of uniform size and sharply demarcated from each other by spindle-cell proliferation with multinucleated osteoclast-like giant cells present.

Despite nuclear irregularity, features of malignancy such as pleomorphism, mitotic activity, and necrosis were not present in this neoplastic process. There were scattered areas of dilated vessels, but these were not of malignant type.

Histologically the tumor was felt to be characteristic of chondromyxoid fibroma (CMF).

Discussion

CMF is one of the less common tumors of bone and few radiologists will encounter this benign tumor outside of large referral centers, and even then only rarely. Described by Jaffe and Lichtenstein in 1948 [1] it is composed of varying proportions of chondroid, fibrous, and myxoid material but, despite its name, is clearly a tumor of cartilage [2, 3, 4] and classified as such. A more appropriate name would be “fibromyxoid chondroma”, but attempts [5, 6] to change its name have been unsuccessful and its original name has stuck.

It is primarily a tumor of the long tubular bones and is particularly common in the metaphyseal regions around the knee joint. When seen in this location, it presents as a large eccentric, expanding lesion with a sclerotic rim and, unlike in other cartilaginous tumors, intrinsic calcification is rare. Periosteal reaction in the absence of fracture is also uncommon. Short tubular bones and flat bones may also be affected.

The tumor has been reported in the spine, but this is an uncommon site. The histological diagnosis of CMF of the spine is, therefore, nearly always a surprise, as it was in the present case. Table 1 summarizes the 20 cases previously reported [7–22] as well as our case. Fourteen of the cases were reported individually as case reports. Seven of the cases were abstracted from several large series. In three (cases 4, 5 and 6) no clinical information was presented [9, 10]. Four of the seven (cases 3, 16, 17 and 18) had some information but no radiographs and limited follow-up [5, 20].

The tumor is slow growing in the spine, as elsewhere in the skeleton. It may spread outside the spine and form bulky soft tissue masses, as in cases 1, 13, 14, and 21 [7, 17, 18]. However, its origin from the spine is virtually always apparent and its extension into the extradural space to cause either cord or nerve compression evident.

This is particularly so on cross-sectional imaging with either CT or MRI.

Table 1 Chondromyxoid fibroma of the spine and sacrum: review of the literature including the present case (*NED* no evidence of disease)

Author(s)	Case no.	Age (years) sex	Clinical presentation	Site	Radiographic findings	Treatment	Results follow-up
Benson and Bass 1955 [7]	1	34 F	Pain in legs and feet, weakness in legs (8 months), tender mass	T2	Destruction of posterior elements spinous process T2–T3, paravertebral mass	Curettage, radiation therapy (2100 rad)	NED 15 months
Gudscha 1968 [8]	2	23 F	Pain, weakness in legs	L3	Destruction of L3	Excision	NED 6 months
Schajowicz and Gallardo (5)	3	6 F	No clinical details (from study of 32 cases)	C3	Expanded spinous process	None available	
Spjut et al. 1971 [9]	4+ 5		Two cases included in study of 40 patients, no information available				
Rahimi et al. 1972 [10]	6		Case included in a study of 76 patients from the Mayo Clinic. No information available				
Raja-Reddy et al. 1973 [11]	7	34 M	Back pain, leg weakness (6 months)	T11	Destruction of posterior elements, extensive destruction of 10th and 11th ribs	Biopsy only, refused further treatment	
Ramani 1974 [12]	8	44 M	Paraparesis	T12	Erosion of posterior elements 12th rib eroded	Complete excision	NED 2 months
Tsuji et al. 1975 [13]	9	9 M	Lower back pain, back deformity (1 month)	L4	Destruction of posterior elements body of L4	Complete excision, bone graft	NED 4 years
Merli et al. 1978 [14]	10	11 M	Severe paraparesis	T10	Body T10 destroyed, enlarged spinous process, myelogram block	Complete excision	NED 20 months
Mayer 1978 [15]	11	23 M	Totally asymptomatic	L2	Expanding lesion of spinous process	Complete excision	Not stated
Nunez et al. 1982 [16]	12	38 F	Pain (2 years), worse (5 weeks), leg weakness and numbness	T5	Extensive destruction of body, compression fracture, posterior elements eroded	Complete excision 12 months	Recurrence at 2 years, re-excised, NED
Standefer et al. 1982 [17]		20 F	Neck pain radiating to left arm	C7	Bulky mass in the neck, destruction C5, body+ posterior elements, mottled calcification	1500 rad to neck, fusion C7-T1, soft tissue resection	No recurrence 15 months
Shulman et al. 1985 [18]		15 M	Lower back pain (1 year), recent limp	Sacrum	Expanding lesion of sacrum, large mass in buttock and pelvis	Combined resection (ant+post)	No following
Provelegios et al. 1988 [19]	15	32 M	Pain in left shoulder (1 year)	C4	Lytic lesion of posterior body, sclerotic margin. Spinal canal not involved (myclogram)	Curettage, bone graft	NED 2 years

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Author(s)	Case no.	Age (years) sex	Clinical presentation	Site	Radiographic findings	Treatment	Results follow-up
Zillmer and Dorfman 1989 [20]	16	20 F	Three patients included in study of 36 patients	C7	Body neural arch eroded, spinal canal involved		
	17	58 F	No clinical data	Sacrum	Extensive sacral destruction, dura infiltrated	Patient died	
	18		No clinical data	Thoracic spine	Marked involvement of spinous process		
Rivierez et al. 1991 [21]	19	41 F	Torticollis, upper limb pain (1 year)	C5	Destruction of part of body, posterior elements, peripheral sclerosis (CT scan, MR, arteriogram)	Resection in 2 stages	NED 10 months
Tsuchiya et al. 1992 [22]	20	19 F	Back pain, leg weakness (2 weeks), paraparesis	T2	Destruction of part of body posterior elements involved, soft tissue mass C6-T2 (bare scan, CT, MRI)	Vertebrectomy, wiring, cement, chemotherapy, radiation therapy	NED 2 years
Cabral et al.	21	19 F	Mass in upper back (3 years), pain (2 months)	L1	Extension and destruction of posterior elements, spinal canal invasion (myelogram, CT)	Extensive resection	NED 5 years

The posterior elements are characteristically involved with varying degrees of erosion of the vertebral body. When this is extensive, it may result in collapse of the body with secondary fracture. In the thoracic region a relatively silent tumor may thus cause an acute paraparesis. This was evident in cases 10 and 20 [14, 22].

Erosion of the vertebral body with a sclerotic margin was noted in two patients, cases 15 and 19 [19, 21]. In two patients the tumor had spread outside the vertebra to involve adjacent ribs. In case 7 [11] the destruction was extensive, involving the right 10th and 11th ribs. This was readily apparent on plain films. In case 8 [12] the rib erosion was of a much lower degree. Benign lesions such as giant cell tumor and aneurysmal bone cyst are well known to erode adjacent bony structures, so the rib involvement is not really surprising.

Of interest is the degree of involvement of the spinous process – either expansion or destruction. In two patients this was the only finding. In three others there was in addition extensive involvement of pedicles and laminae. In the one patient who was completely asymptomatic, case 11 [15], erosion of the spinous process was the only finding. Spinous process involvement may be more suggestive of CMF than either aneurysmal bone cyst or osteoblastoma, and extensive destruction of this process should raise the suspicion of CMF.

Hypervascularity is not a common feature of this tumor. Aneurysmal bone cyst may be engrafted on CMF, but this is uncommon [20]. Only three patients underwent CT; MRI was only performed in two patients.

Calcification of the tumor was thought to be present in only one patient, case 13 [18]. Feldman et al. [23] in a large series found macroscopic calcification in only 2%. Microscopic calcification may found in up to 20%.

In the 14 patients whose clinical details had been well documented, the thoracic spine was the most common site. Eight patients were under 25 years old. The oldest was 41. As a group, the younger patients had much shorter histories. There was no significant sex difference. The optimal treatment is enbloc resection. Simple curettage is unlikely to result in a cure.

Only one patient experienced a recurrence (case 12). Following re-excision there was no recurrence 12 months later. Follow-up in most cases in this collected series was quite short, and therefore the recurrence rate cannot be accurately assessed.

Radiation therapy, because of its oncogenic potential, is contraindicated. Nevertheless, four patients in this study were irradiated. In two cases this was used in an attempt to shrink very large tumors. In a third case (case 20) it was used because of a mistaken diagnosis of osteosarcoma on frozen section [22]. The fourth case, one of Zillmer and Dorfman's three cases, a woman aged 20 years had intralesional excision and irradiation. Seven years later she developed recurrent CMF and malignant fibrous histiocytoma.

A patient with a large sacral lesion with invasion of the dura died. A patient whose case was reported by Dahlin and Unni [24] developed a sarcoma following irradiation. These are unusual and rare cases. Spontaneous malignant change is extremely rare and has only been reported twice in the literature [20].

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Note added in proof Since acceptance of this paper, the patient has presented with a 5 cm bony hard mass in the scar. Surgical excision revealed this to be a simple recurrence.