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Chondromyxoid fibroma of the ala of the sacrum presenting as a cause of lumbar pain in an adolescent

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Abstract We report a case of chondromyxoid fibroma of the ala of the sacrum: its presentation, diagnosis, treatment, and resolution. Although this tumor is admittedly rare, our case demonstrates the need for careful evaluation of back pain in an adolescent.

Key words Chondromyxoid fibroma · CT · Sacrum · Adolescent

Introduction

Persistent, severe lumbar pain in teenagers is relatively unusual. This case report provides an example of the need for complete and thorough evaluation in an attempt to find the true etiology of the patient's symptoms.

In 1948, in a report of eight cases, Jaffe and Lichtenstein were the first to describe chondromyxoid fibroma, a slowly growing benign bone tumor composed of varying amounts of chondroid, myxoid, and fibrous elements [10]. They recognized it as being distinct from chondrosarcoma and histologically resembling the myxoma of bone described earlier by Bloodgood [9, 10]. This distinction was important since the tumor was easily mistaken for a sarcoma and subsequent treatment could be much too aggressive [2, 5, 9, 17]. In recent times, this tumor has been

well described and differentiated from myxoma, myxosarcoma, fibromyxoma, chondrosarcoma, and chondromyxosarcoma.

Chondromyxoid fibroma is a distinctly uncommon tumor. In most large series, it has comprised fewer than 1% of the documented bone tumors [2, 4, 6, 14, 15]. To date, only about 400 cases have been reported [1–15, 17–20].

The tumor most often involves long tubular bones, especially the tibia (about one-third of the reported cases), the femur, and the fibula. Less commonly affected are the bones of the feet, upper extremities, and pelvic girdle, with occasional examples arising in the scapula, ribs, vertebrae, sternum, jaw, and skull bones. A review of the literature reveals only three cases of chondromyxoid fibroma involving the sacrum [2, 12, 18]. Our report describes one additional case, involving the ala of the sacrum.

Case report

A 17-year-old healthy white girl, L.F., was referred to a pediatric orthopedic spinal consultant after 6 months of unremitting lumbar and right sacroiliac pain with radiation to the right buttock. The pain developed while the patient was participating in athletics and, although not associated with a specific injury, was diagnosed as muscular strain. Extension exercises of the lumbar spine and buttock were prescribed and, over the next 3–4 months, there was some improvement.

Two months prior to referral, the patient returned to school and began participating in a vigorous high school soccer program. The pain increased and was exacerbated by extension of the lumbar spine; athletics were eliminated. There was no radiation of pain or numbness to the lower leg and no history of bowel or bladder changes. Subjective weakness was not present.

On physical examination, the patient stood with a very slight right lumbar scoliosis. There was no lumbar rotation on forward bending. Forward flexion of the lumbar spine was to 70° with extension to neutral. Right lateral bending of the lumbar area to 15° caused right sacroiliac pain. Straight leg raising was to 70° on the right and Patrick's test reproduced the patient's pain. The area of the right sacroiliac joint and the posterior-superior iliac spine were tender to palpation. There was 1 cm of atrophy of both the right thigh and calf when measured 12.5 cm above the superior pole of the patella and 12.5 cm below the tibial tubercle. Muscular strength, sensation, and deep tendon reflexes were normal and symmetrical in both lower extremities.

A radiograph of the lumbar spine and sacrum revealed a lytic lesion in the right ala of the sacrum with a well-defined sclerotic border and some cortical expansion (Fig. 1). CT confirmed the presence of a lytic area with a stated density of 25 HU (Fig. 2). This is higher than usually seen in a unicameral bone cyst. A bone scan did not reveal increased uptake in the area of the sacroiliac joint surrounding the lesion. An EMG, nerve conduction studies, an H-reflex stimulation were carried out in the lower extremities as well as along the paravertebral muscles; all of these were unremarkable.

A biopsy and/or curettage and bone grafting was planned. The right sacroiliac joint was approached through an oblique incision. Exposure of the ala of the sacrum found the inferior and lateral aspect to be softened. The lesion had perforated the cortex of the sacrum, but the periosteum remained intact. The mass measured 4 × 3 × 3 cc in size and contained gelatinous, brownish-gray, fibrous but gritty material. A frozen section revealed only multiple giant cells and, since there was no evidence of malignancy, the tumor was completely removed by curettage. The lesion extended close to the articular cartilage of the sacroiliac joint, but did not appear to enter the synchondrosis. Local bone was taken from the right ilium and grafted into the curettage bed.

The gross specimen consisted of multiple fragments of soft, papillary, white to pink translucent tissue. Microscopically, this lesion showed characteristic lobules separated by more cellular areas containing oval to spindle-shaped stromal cells and abundant osteoclast-like giant cells (Fig. 3). Most histologic fields consisted of hypocellular myxoid and chondroid areas.

Two years after curettage the patient developed aching pain in the region of her right sacroiliac joint. Radiographs were remark-

Fig. 1 Plain radiograph of the lumbar spine and sacrum demonstrating a lytic lesion of the right ala of the sacrum

Fig. 2 A CT scan confirmed the presence of this lesion

Fig. 3 The lesion showing characteristics lobules separated by more cellular areas containing oval to spindle-shaped stromal cells (× 100)

Fig. 4 Follow-up CT scan after 8 years showed no evidence of tumor recurrence

able only for degenerative changes in the right sacroiliac joint, but the possibility of a recurrence could not be discounted. Thus, the patient underwent reexploration and biopsy; no tumor was found, but the right sacroiliac joint showed degenerative changes and was fused in situ with autogenous cancellous graft.

Eight years have now passed since the patient's sacroiliac fusion. She has been fully active in recreational athletics and is pain free. She is able to participate in aerobics and other vigorous types of lumbar and pelvic motion. Her only stated complaint is occasional stiffness. Recent follow-up CT of the sacrum and pelvis revealed no evidence of recurrence of the tumor and complete fusion of the right sacroiliac joint (Fig. 4). The patient is functioning well and carrying out normal activities without limitation.

Discussion

There has been little previous experience with a tumor of this type involving the sacrum. Indeed, only three other cases have been reported [2, 12, 18]. Pain is the most common presenting complaint, although local swelling may be present if the overlying tissues are not thick enough to obscure the developing mass [2, 4, 7, 11, 13, 14, 20]. Occasionally the tumor is asymptomatic and physical exam unrevealing. These facts, combined with the lesion's rarity, can lead to delay in diagnosis when the tumor presents in an unusual location.

The radiologic, pathologic, and histologic findings reported in our case are characteristic of chondromyxoid fibroma in general and have been detailed on numerous occasions [1-15, 17-20].

Treatment can be by en bloc resection for bones of minor functional demands or by curettage and grafting for large weight-bearing bones. Curettage alone is associated with a higher recurrence rate, but if performed thoroughly, curettage combined with bone grafting closely approaches the nearly zero recurrence rate of en bloc resection [2, 3, 6, 8, 10, 11, 13, 19]. Sarcomatous transformation is exceedingly rare, with only two confirmed cases reported in the literature, one of these arising in an irradiated bed [2].

Our report represents the first long-term follow-up of a sacral chondromyxoid fibroma. (As with many rare tumors of the sacrum, some pain after resection is found [16]. In this patient discomfort may have resulted from irregularity of the sacroiliac joint, since degeneration in this area seems to have occurred as the patient aged and since fusion of the affected joint relieved the symptoms.)

Back pain in a child or adolescent must be evaluated carefully. A detailed history and physical examination will direct the examiner to specific studies such as radioisotope imaging, CT, and MRI should these be necessary to assess rare pathologic processes.

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