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Giant-cell tumour in the cervical spine: a case report

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A forty-six-year-old man presented with a two-month history of increasing neck pain of insidious onset. He received treatment from his familty doctor and chiropractor consisting of analgesics and manipulation respectively, both of which did not offer relief. The patient presented to University Hospital where plain radiographs and CT showed a pathological fracture of the C4 vertebral body. A neoplasm was suspected and surgical excision revealed a giant-cell tumour of bone. This type of neoplasm is rare in the spine and difficult to manage in this site. This case highlights some of the problems encountered in the treatment of giant-cell tumour of the spine. (JCCA 1989; 33(2): 76–81)

KEY WORDS: giant-cell tumour, cervical spine, chiropractic, manipulation.

Un homme de quarante-six ans présente depuis deux mois une douleur croissante au cou d'origine insidieuse. Il s'est fait traiter par son médecin de famille et son chiropraticien et le traitement a consisté respectivement en analgésiques et en manipulation, dont ni l'un ni l'autre n'a permis de soulagement. Le malade s'est présenté à l'Hôpital universitaire où des radiographies simples et la tomographie axiale ont révélé une fracture d'ordre pathologique du corps vertébral C4. On a soupçonné la présence d'un néoplasme et l'excision chirurgicale a révélé une tumeur à myéloplaxes osseux. Ce genre de néoplasme est rare à l'épine dorsale et difficile à traiter dans ce foyer. Ce cas permet de souligner certains des problèmes inhérents au traitement d'une tumeur à myéloplaxes à l'épime dorsale. (JCCA 1989; 33(2): 76–81)

MOTS CLEFS: tumeur à myéloplaxes, épine cervicale, chiropraxie, manipulation.

Introduction

Giant-cell tumour of bone is defined by the World Health Organization as an aggressive tumour characterized by richly vascularized tissue containing many round, ovoid, or spindle-shaped, mononuclear stromal cells and fewer multinucleated giant-cells of unknown origin.[1] The incidence of this tumour is low and it very rarely affects the spine above the sacrum. There is considerable controversy as to the histogenesis, clinicopathologic diagnosis, and treatment of this tumour. It is also very difficult to predict whether or not a giant-cell tumour will undergo malignant degeneration or metastasize.

Historically, there has been difficulty separating true giant-cell tumour of bone from a group of bone lesions with giant-cells in them (Table 1). Definitive diagnosis requires radiographic and histologic confirmation. Diagnosis is often delayed since the presenting complaints may be minimal. It is not uncommon for these patients to be seen by several practitioners prior to diagnosis. The following case illustrates these points.

Table 1 Giant-cell tumour variants that are commonly misdiagnosed as giant-cell tumours

Former variant designation	Present classification
Chondromatous	Benign chondroblastoma Epiphyseal chondroblastoma Codman's tumour Epiphyseal chondromatous giant-cell tumour
Myxomatous	Chondromyxoid fibroma Fibromyxoid chondroma
Xanthomatous	Nonosteogenic fibroma Nonossifying fibroma Fibrous cortical defect Metaphyseal cortical defect Histiocytic xanthogranuloma Fibrous histiocytoma
Subperiosteal	Aneurysmal bone cyst Multilocular hematic bone cyst

(modified from Schajowicz)[1]

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Case presentation

N.P. is a forty-six-year-old foreman who presented to the Emergency Department at the University Hospital with neck pain of two months' duration. The onset was insidious and the pain was worsening. The pain was located in the mid-cervical spine with radiation into both shoulders. Treatment with analgesics from his family doctor and manipulation by his chiropractor offered no relief. The morning prior to presentation, the patient asked his family doctor to send him for radiographs because the pain had become quite severe.

On physical examination, the cervical spine range of motion was limited and painful in all directions. There was no neurological deficit in the upper limbs. Palpation revealed marked tenderness in the mid-cervical spine.

Plain radiographs showed marked destruction of the C4 vertebral body (figure 1). CT (figure 2) and tomograms defined the lesion and showed possible involvement of the right neural arch of C4 and the inferior end plate of the C3 vertebral body. A bone scan demonstrated slightly increased uptake at the C4 level with no other lesion noted elsewhere.

The patient underwent anterior decompressive surgery to remove the tumour and a fusion between C3 and C5 with iliac bone graft. He was fitted with a halo-vest postoperatively. The excised tissue, which was sent for pathological examination, showed a giant-cell tumour of bone (figures 3 and 4). Adjunctive radiotherapy was planned to start after removal of the halo-vest.

When the halo-vest was removed two months later, the patient developed severe neck pain. Plain radiographs showed progressive destruction of the graft, as well as the C3 and C5 vertebral bodies (figure 5). Within 48 hours of removal of the halo-vest, while waiting for radiotherapy, the patient developed profound quadriparesis with bowel and bladder dysfunction over-night. Emergency anterior decompressive surgery with

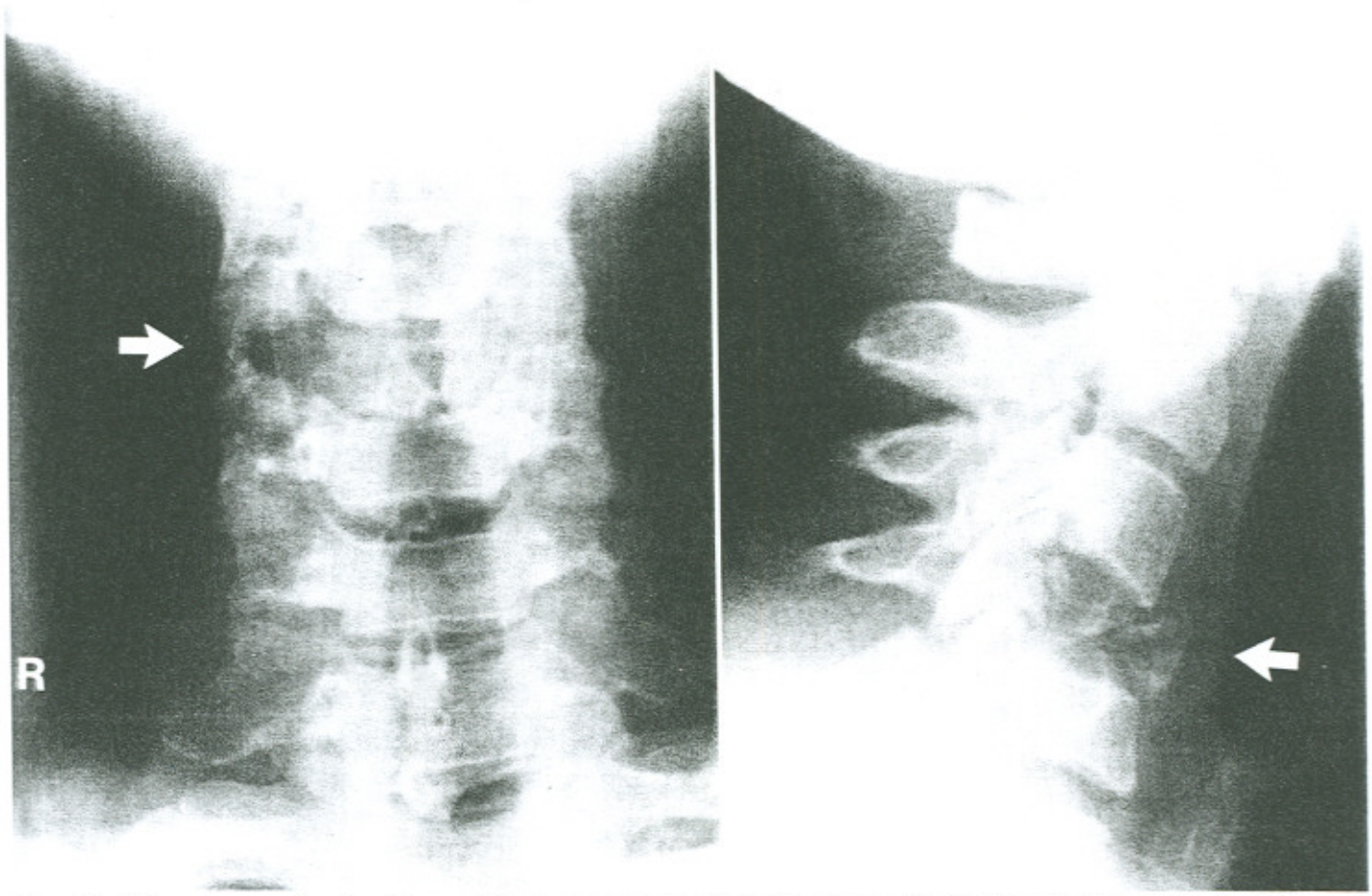

Figure 1 (a) An anteroposterior view of the cervical spine shows extensive destruction of the vertebral body and the right neural arch of C4 (arrow). (b) A lateral radiograph of the cervical spine shows marked destruction of the C4 vertebral body and a kyphotic deformity centred at the same level (arrow). There is an increased interspinous distance between C4 and C5.

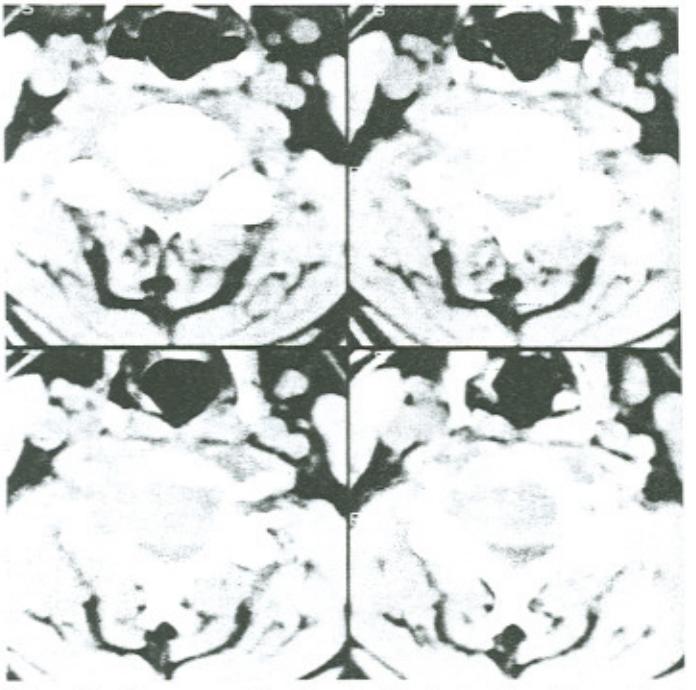

Figure 2 CT scans confirm the extensive destruction of the vertebral body and right neural arch of C4. A soft tissue mass can be seen within the spinal canal at the level of C4, particularly on the lower two scans.

anterior and posterior instrumentation and fusion were performed. Postoperatively, motor function began to improve almost immediately. The patient underwent radiotherapy for five weeks and was subsequently discharged.

When seen in review one year after the second operation, the patient was remarkably well considering the circumstances. He was pain-free and had returned to farming. Examination revealed only twenty-five percent of the normal range of motion in the cervical spine. Muscle strength in the upper limbs was 4+/5 and his gait was somewhat unsteady. He needed self-catheterization to void. Plain radiographs showed good alignment and no sign of recurrence of the giant-cell tumour (figure 6a, b). Overall, the patient seemed to have made a good recovery.

N.P. will be seen for follow-up periodically due to the high risk of recurrence of these tumours following intracapsular excision.

Discussion

Epidemiology

Giant-cell tumour is a relatively uncommon primary tumour of bone, representing five to eight percent of all primary bone tumours.[1,2] It has a slight predilection for females. The tumour occurs most commonly between twenty and forty years of age, with peak incidence in the third decade.

The most common location is in the epiphysis of a long bone.

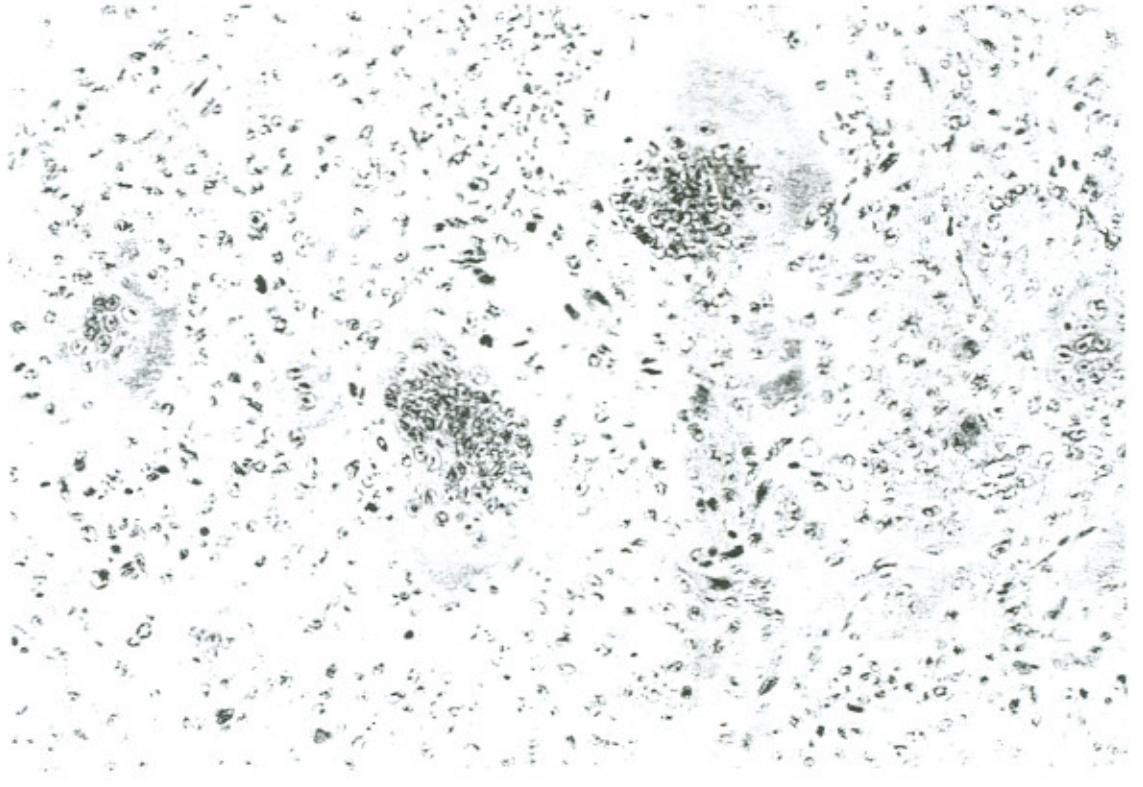

Figure 3 This pathological section shows a neoplasm composed of stromal cells with indistinct cell boundaries and oval to spindle-shaped nuclei. Numerous multinucleated giant cells with bland cytoplasm are present. (H&E X240)

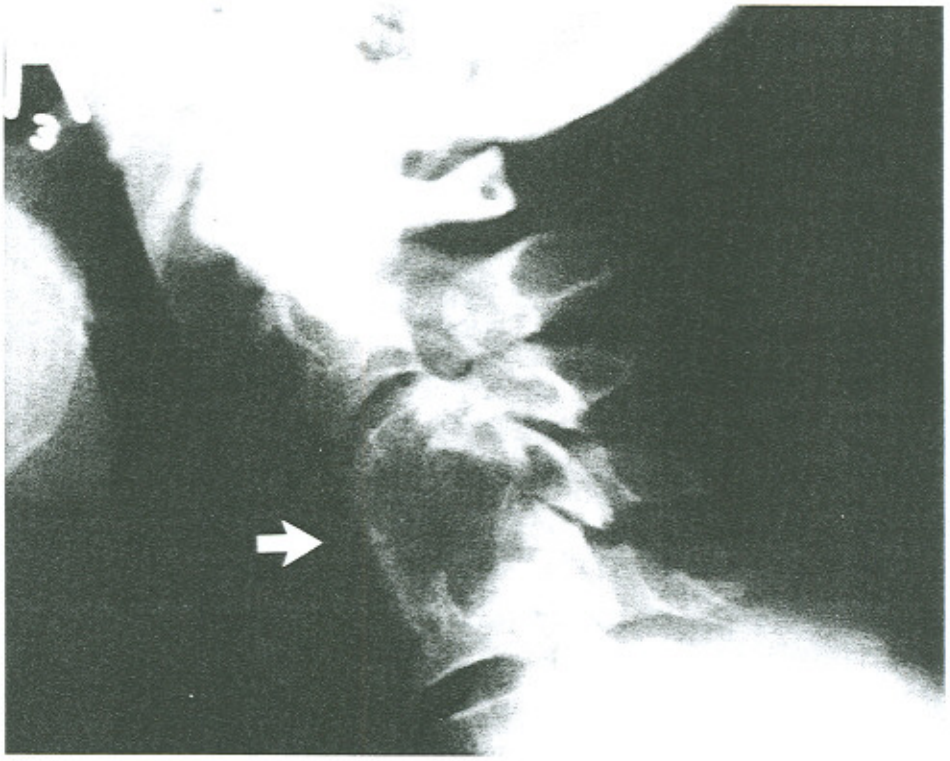

Figure 5 A lateral radiograph of the cervical spine taken two months after first surgery shows marked destruction of the bone graft and the vertebral bodies of C3 and C5 (arrow).

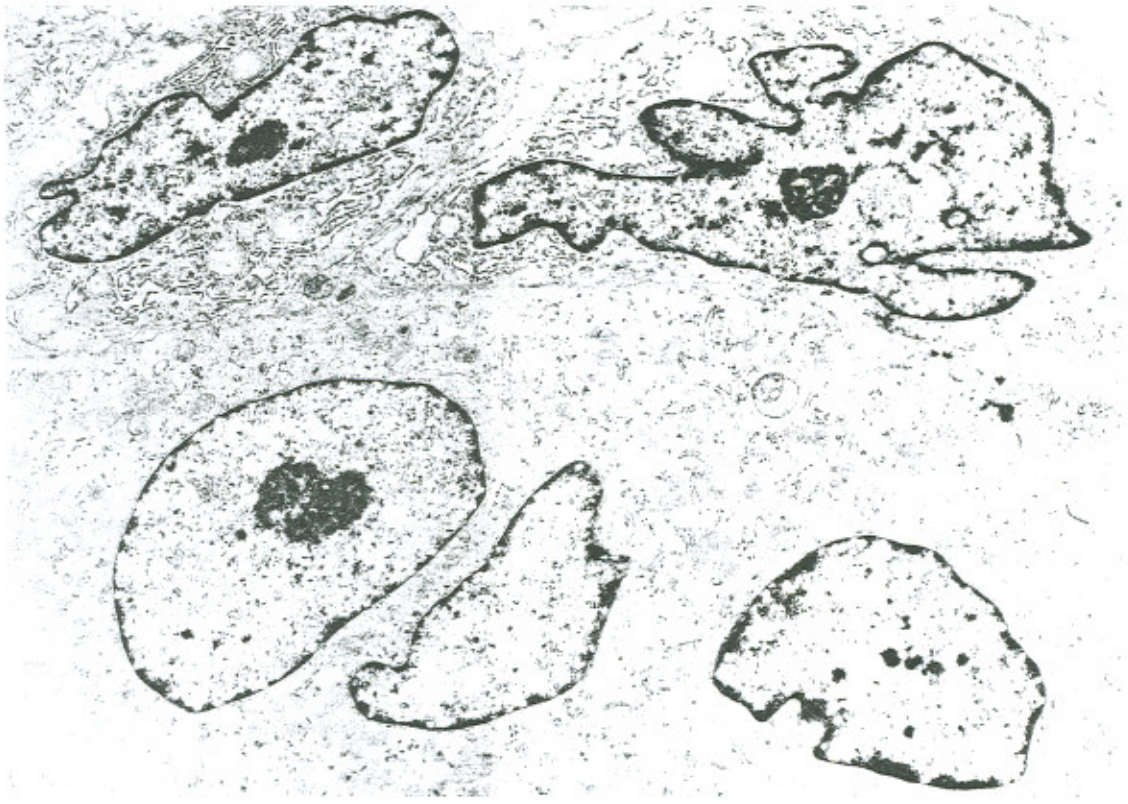

Figure 4 An electron micrograph of the tumour shows the interface between a multinucleated giant-cell (below) and two spindle-shaped stromal cells (above). The giant-cell cytoplasm contains numerous mitochondria and sparse rough endoplasmic reticulum. Three ovoid giant-cell nuclei are included in the micro-graph. The two stromal cells have abundant and slightly dilated rough endoplasmic reticulum in their cytoplasm and irregular spindle-shaped nuclei. (X 7400)

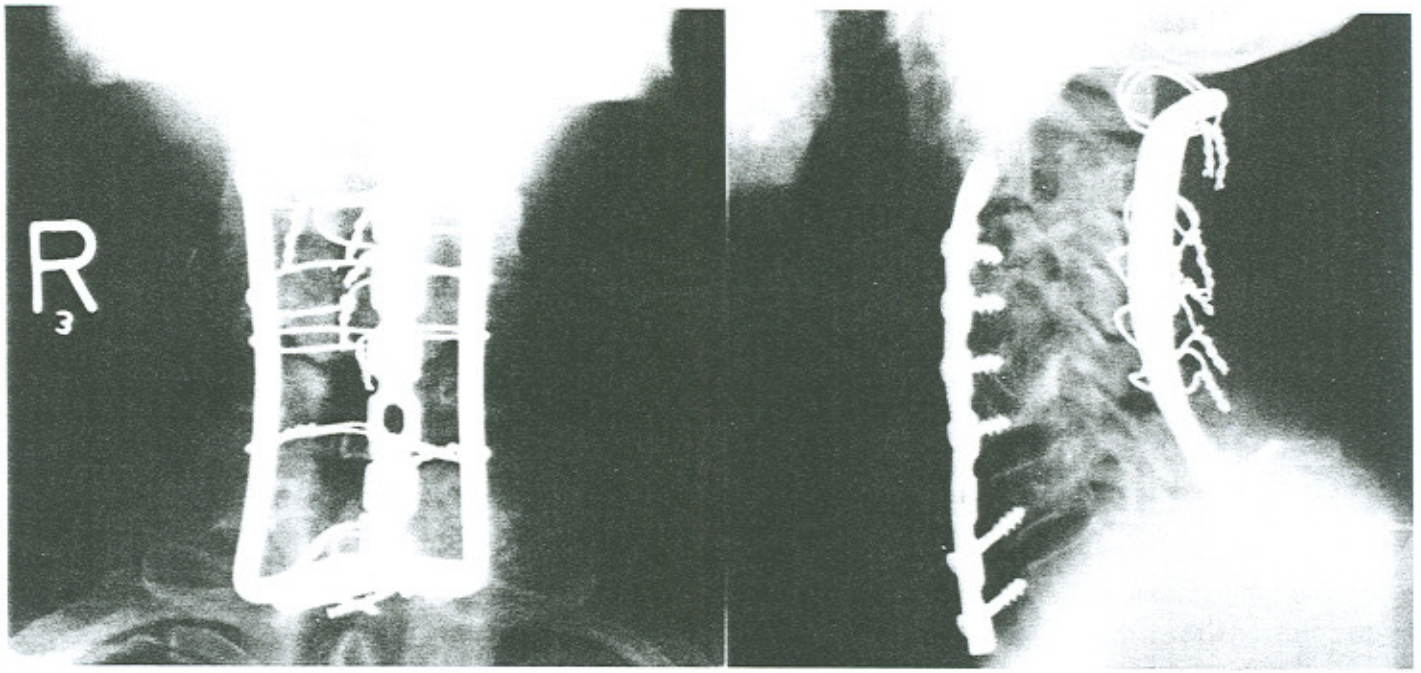

Figure 6 **(a & b)** An anteroposterior and lateral radiograph of the cervical spine taken after anterior and posterior instrumentation. The internal fixation appears stable and the alignment is satisfactory. There is no evidence of recurrence of the giant-cell tumour.

Almost fifty percent of the tumours occur about the knee in the distal femur and in the proximal tibia.[1,3] The distal radius, proximal humerus, and sacrum are less common sites. Involvement of the vertebrae above the sacrum, mandible, metatarsals, metacarpals, and phalanges is rare. Dahlin reported thirty-one cases of giant-cell tumour of the vertebrae above the sacrum.[4] Schajowicz reviewed Dahlin's published illustrations and suggested that the majority of the tumours were variants rather than true giant-cell tumours. Most of the lesions of the mandible are reparative giant-cell granulomas, a giant-cell variant.[1,3,5]

Multicentric giant-cell tumours are rare and they have a propensity for the bones of the hand.[2] Some authors feel that these lesions are more likely due to hyperparathyroidism or secondary bone involvement by localized nodular tenosynovitis.[5,6] It is also difficult to distinguish a multicentric giant-cell tumour from a primary giant-cell tumour with metastatic spread.[1,3] Giant-cell tumours associated with Paget's disease are extremely rare. Twenty-three cases have been reported in the literature.[7] The cause and effect relationship between the tumour and the disease is not clear.

Diagnosis

The most consistent symptom of giant-cell tumour is pain. There may be associated swelling of the affected area and limitation of motion of the adjacent joint. Occasionally, neurological deficit is present when the tumour affects the spine.

Radiographic features include an expanding zone of radiolucency located eccentrically in the epiphyseal end of a long bone. It extends both towards the articular cartilage and towards the metaphyseal region. The cortex is generally thinned, expanded, and occasionally destroyed. However, a periosteal reaction is almost always absent. In the spinal levels above the sacrum, the vertebral body is most commonly involved. Radiographic appearance varies from a lytic defect to a pathologic fracture (figure 1). Most authors agree that it is difficult to predict the behaviour of the tumour from its radiographic appearance.

Pathology

The tumour tissue has a soft, friable appearance that varies from a gray to a red colour.[1,3] Thin septa of connective tissue extends throughout the tumour which is surrounded by a thin cortical shell of bone. Small cystic portions, indicating hemorrhage, are present in advanced tumours. These are often mistaken for aneurysmal bone cysts. In some advanced lesions, yellowish-gray necrotic areas typical of xanthomas are seen. Fibrosis and osteoid production may be present as a result of previous fracture or treatment.

The histologic structure of a giant-cell tumour is character-

ized by many multinucleated giant-cells that are distributed among mononuclear stromal cells (figure 3). The cytoplasm of the giant-cell has a granular appearance.[1] There are usually more than fifteen to twenty nuclei, sometimes even as many as one hundred, in a single giant-cell. The appearance of these nuclei is identical to that of the nuclei of the mononuclear stromal cells. The nuclei of the stromal cells show hypochromatism with few mitotic figures.[1,3] Silver staining shows a dense network of reticulin fibres that surround the giant-cells and stromal cells.

Ultrastructural features of the giant-cell includes a large number of mitochondria, a small amount of rough endoplasmic reticulum, and a variable number of lysosome-like bodies.[1,8] The stromal cells contain few mitochondria and a more abundant rough endoplasmic reticulum (figure 4). Histochemically, giant-cells differ from stromal cells in that they have a high content of acid phosphatase, beta glucuronidase, and succinic dehydrogenase. These histochemical and ultrastructural features are similar to those of osteoclasts, and the tumour is often referred to as an osteoclastoma.

The literature is divided as to the origin of the giant-cells. Most authors believe they arise by fusion of the mononuclear stromal cells while others advocate a mechanism of amitotic nuclear division. The stromal cells are undifferentiated mesenchymal cells with features suggestive of a fibroblastic origin.[1,5] The stromal cells are the neoplastic part of the tumour.

Although giant-cell tumours are often considered benign, seven to thirty percent of them undergo malignant transformation into fibrosarcoma or osteosarcoma.[1,2] It is almost impossible to distinguish histologically which tumour will stay benign and which one will undergo malignant degeneration. Most authors have observed that the majority of the malignant tumours were benign ones that had previously undergone surgery or radiotherapy. Nascimento et al. reported eight cases of primary malignant giant-cell tumour of bone that showed malignant behaviour from the very onset.[9] However, some authors argue against the existence of a true primary malignant giant-cell tumour and suggest that it is actually a sarcoma.[1]

Pulmonary metastasis may occur with both benign and malignant giant-cell tumours. There is a twenty percent incidence with a high recurrence rate. It is difficult to predict histologically which tumour will metastasize and thus, the grading of giant-cell tumours, both histologically and radiographically, has no predictive value as to the behaviour of the tumour.

Treatment

The treatment of choice for giant-cell tumour is excision. In difficult areas like the spine, curettage and bone grafting, with or without cauterization, is preferred. Large or aggressive tumours that involve the soft tissues are usually excised followed by bone graft, instrumentation, and fusion. Radiotherapy is reserved for lesions not amenable to total excision, as in the spine.[3] Radiotherapy is not recommended for tumours in the extremities because of the high risk of post-irradiation malignant transformation. It is used more as an adjunctive therapy after excision and curettage when there is incomplete removal of the tumour tissue.

Patients do well with surgery, but there is a fifty percent recurrence rate.[1] Long-term follow-up is essential. There have been reported cases of recurrence fifteen years after initial treatment of a giant-cell tumour.

Conclusion

In a patient that presents with progressively worsening neck pain of insidious onset, radiographs should be taken before commencement of therapy. Neoplasm must be considered in the initial differential diagnosis, especially when the pain is severe and progressive. Prompt referral to an orthopaedic surgeon for treatment can greatly improve the prognosis.

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