GIANT CELL TUMOURS OF THE SACRUM

T S Liang, C T Tan, B K Tay, S Krishnamoorthy

ABSTRACT

Giant Cell Tumours (GCT) of the Bone is one of the commoner primary bone tumours. Although considered a benign tumour, it does occasionally metastasize to the lungs. Treatment modalities vary according to the surgical staging and the site of the tumour. Treatment is further complicated when the tumour occurs in difficult locations like the sacrum. The paper includes a review of literature into treatment options and the presentation of 3 patients with sacral GCTs, one of which also has multiple pulmonary metastases from a "benign" giant cell tumour.

Keywords: Giant Cell Tumour, Sacral Tumours

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INTRODUCTION

A lesion of bone, possibly a giant cell tumour was first described by Cooper and Travers in 1818 but it was not till a hundred years later that the name of Giant Cell Tumour was introduced by Bloodgood in 1919. Stewart in 1922 noticed the giant cells which he believed were derived from osteoclasts and proposed the name osteoclast sarcoma from which osteoclastoma was derived. Many lesions were confused with Giant Cell Tumour because of a similar radiographic picture and the histological presence of giant cells. Lesions previously confused with giant cell tumours included aneurysmal bone cysts, chondroblastomas, non-ossifying fibromas and hyperparathyroid bone lesions. Jaffe, Lichtenstein and Portis in 1940 attempted to define what is to be regarded as a proper giant cell tumour.

Giant Cell Tumours are widely regarded as benign tumours of bone with some unusual behavioural characteristics. It should be differentiated from giant cell sarcomas which are malignant and to be managed differently. Giant Cell Tumours differ from other benign tumours in that occasional metastases (usually to the lung) are seen. Contrary to other tumours which metastasize it has been found that patients with metastases in giant cell tumours still have good prognoses (Bertoni)[1].

The clinical presentation (age, sex, location) of giant cell tumours with pulmonary metastases was similar to that of ordinary giant cell tumours. The histology of the primary lesion in patients with metastases was identical to cases that did not metastasize. In fact, benign giant cell tumour of bone remains a difficult and challenging management problem because there are no absolute clinical, radiographic or histologic parameters that accurately predict the tendency of any single lesion to recur or metastasize.

The mainstay of treatment is surgery[2] but the surgeon is often faced with the dilemma of joint preservation following surgical excision as it is the tendency of this tumour to occur peri-articularly. Surgical management is also complicated when the occurrence of the tumour is in the spine (8%)[3]. Enneking's surgical staging is helpful in planning the initial surgical treatment (see Table I)[4]. The ideal aim of management is to eradicate the tumour and preserve the joint. Table I also shows the frequency which Giant Cell Tumours present as Stage 1, 2 or 3 tumours. Preservation of the joint usually implies a currettage procedure and this gives different recurrence rates when applied to the various stages of presentation. A Stage 1 tumour when curretted gives a less than 10% recurrence rate and a Stage 3 tumour a 60-70% recurrence rate (see Table II). Fortunately surgical adjuncts are available which will half the recurrence rates. They may be divided into thermal eg liquid nitrogen, bone cement or chemical eg phenol.

Table I - Enneking's Surgical Staging of Benign Tumours As Applied To Giant Cell Tumours Of Bone.

Stage 1 :	Inactive Lesion	(10% of GCTs)
Stage 2:	Active Lesion but contained within bone.	(60% of GCTs)
Stage 3:	Aggressive Lesion extending outside bony confines.	(30% of GCTs)

Table II - Results After Treatment With Currettage Alone.

Stage 1 :	10% Recurrence Rate
Stage 2:	30% Recurrence Rate
Stage 3:	60-70% Recurrence Rate

Adding an adjuvant halves the recurrent rate.

Radiation therapy is a secondary method of treatment and has its major role in the treatment of giant cell tumours of the spine that are not amenable to complete surgical resection[5].

With this background on the difficult problems one can face with giant cell tumours, we wish to present the management of 3 cases of giant cell tumours in a difficult location, the sacrum. One of the patients had multiple pulmonary metastases at the time of diagnosis. These patients were treated in our Department from 1989 and have between 12 - 24 months of follow-up.

CASE REPORTS

Case 1:

This was a 31-year-old female presenting with low back pain with right sciatica for 1.5 years. She was found to have a

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Fig 1a - Case 1 Sacral radiograph showing a radioluscent lesion destroying the body of the sacrum and the adjacent right ilium.

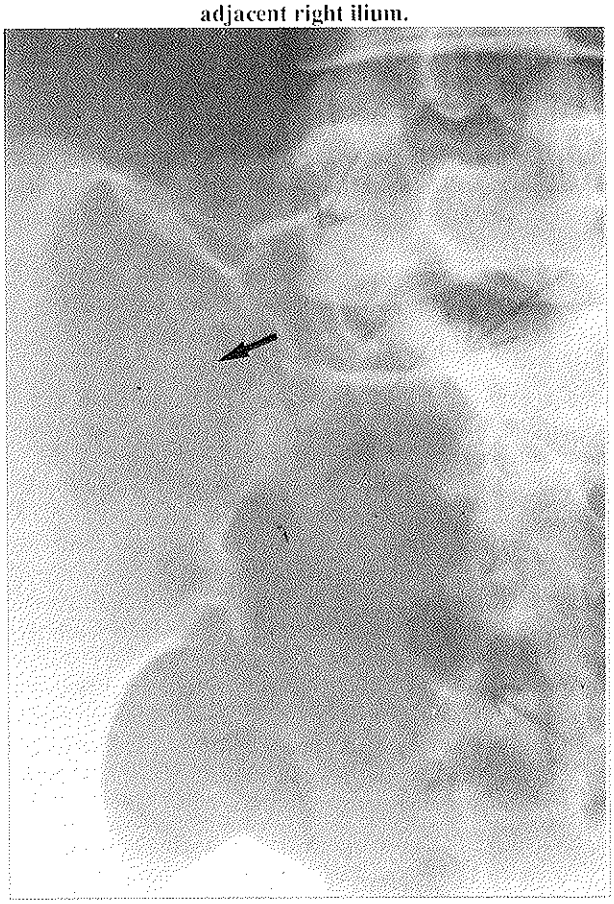
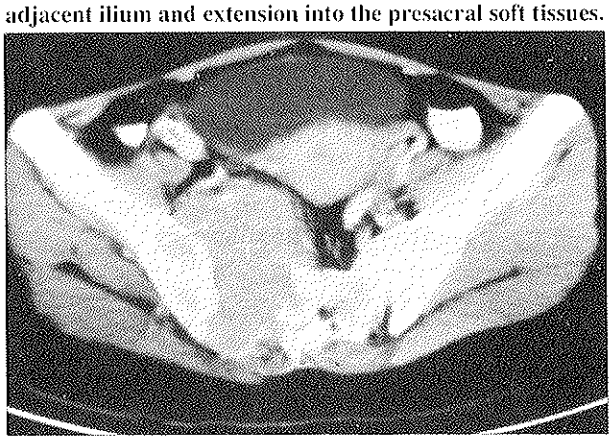


Fig 1b - CT Scan of the sacrum showing a Stage 3 tumour destroying the right side of the sacral vertebra, the adjacent ilium and extension into the presacral soft tissues.



limited straight leg raising test. Plain X-rays showed a destructive lesion in the right side of the body of the upper 3 sacral vertebra with extension across the sacro-iliac joint to the right ilium as well as to the L5 vertebral body (Fig 1a). Staging studies included chest X-rays, blood biochemistry, CT Scan of the lumbo-sacral region and the lungs, a MRI Scan of the lesion and angiography. Blood biochemistry was normal as was the chest X-ray. The imaging studies showed the anatomical extent of the lesion which had a large presacral soft tissue component as well as involving the sacral nerve roots within the spinal canal (Fig 1b, Fig 1c). CT Scan of the lungs showed multiple bilateral pulmonary lesions (Fig 1d).

The staging studies were followed by an incisional biopsy of the sacral lesion and a fine needle aspiration biopsy of the pulmonary lesion both of which showed a giant cell tumour. (This makes the lesion a Stage 3 tumour). She underwent a

Fig 1c - Axial MRI showing the soft tissue extension of the sacral tumour

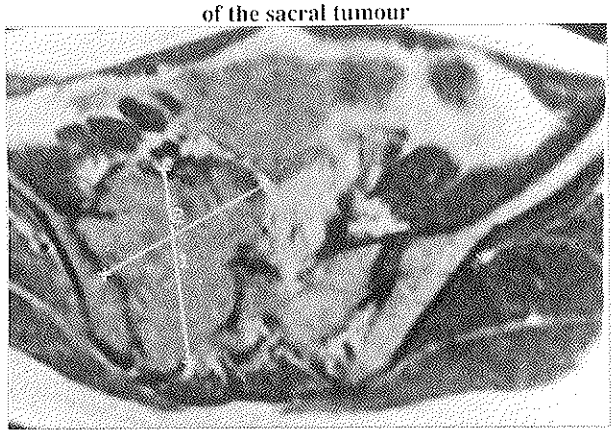


Fig 1d - CT Scan of the lungs showing multiple metastatic lesions in the right lung

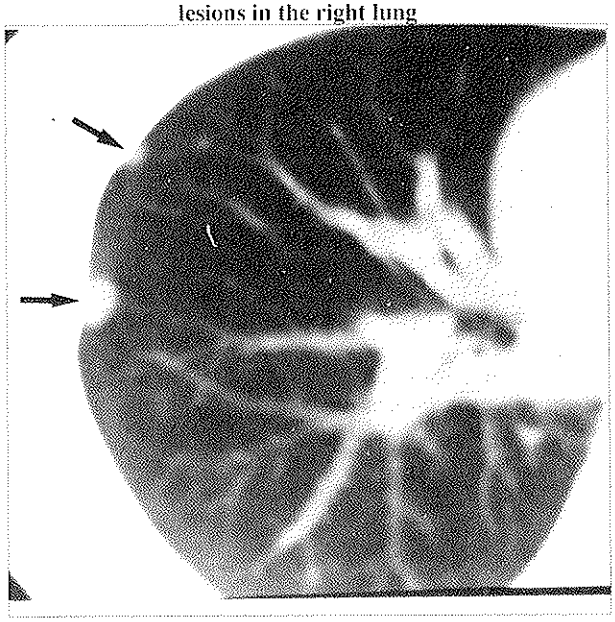
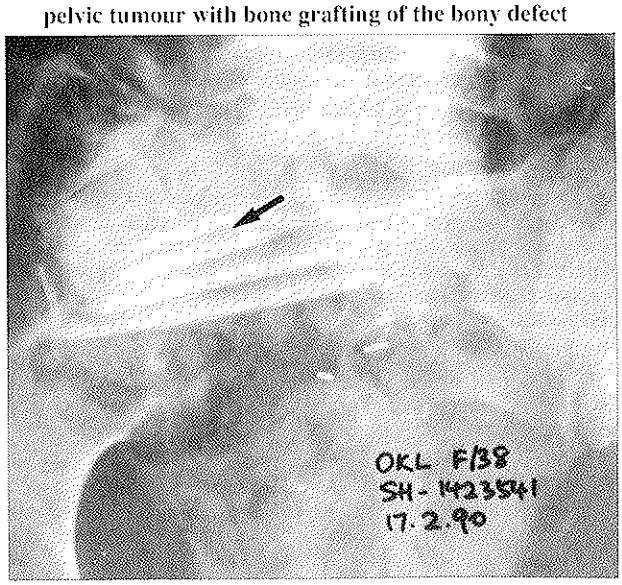


Fig 1e - Pelvic radiograph following currettage of the pelvic tumour with bone grafting of the bony defect



currettage and bone grafting procedure via a transperitoneal approach. No adjuvant therapy was used (Fig 1e). She was later referred for a thoracotomy to attempt to remove the pulmonary lesions. This was abandoned because of the numerous lesions encountered. She was then placed on chemotherapy with 6 courses of epirubicin and cis-platinum. Twelve months

Fig 1f - CT Scan demonstrating a recurrent lesion in the ilium associated with a para-psoas soft tissue lesion

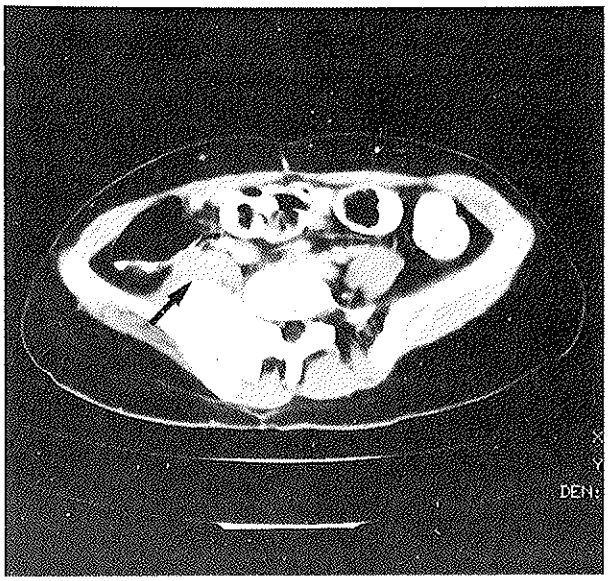


Fig 2a - Case 2 Pelvic radiograph showing the destruction of the upper sacrum with the adjacent L5 vertebrae and left ilium

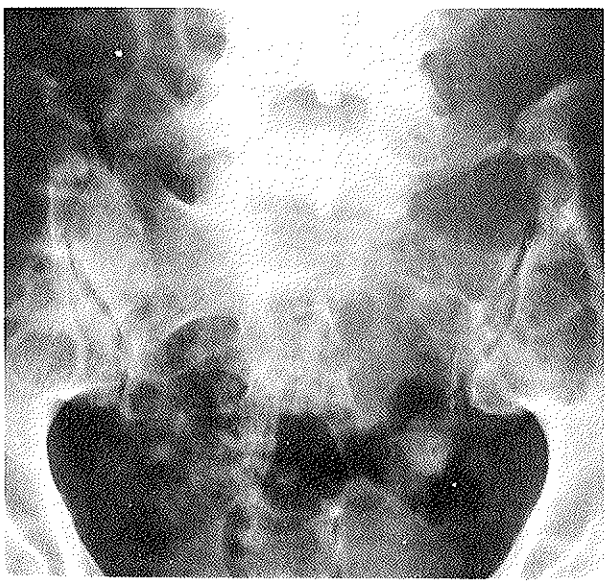


Fig 2b - Case 2 CT Scan showing the extent of tumour within the sacrum and displacement of the dura

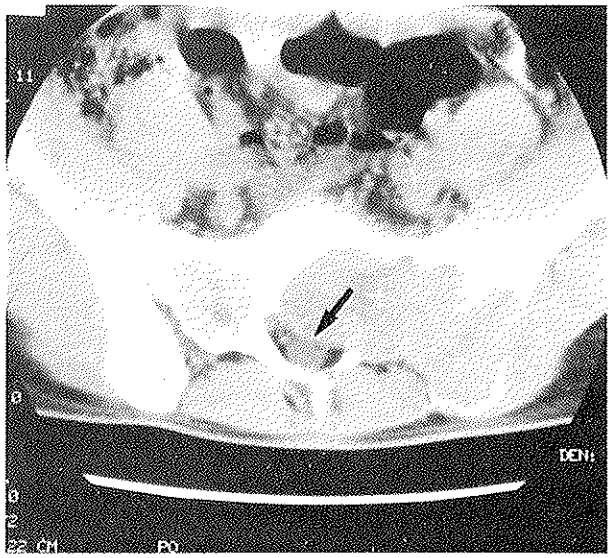
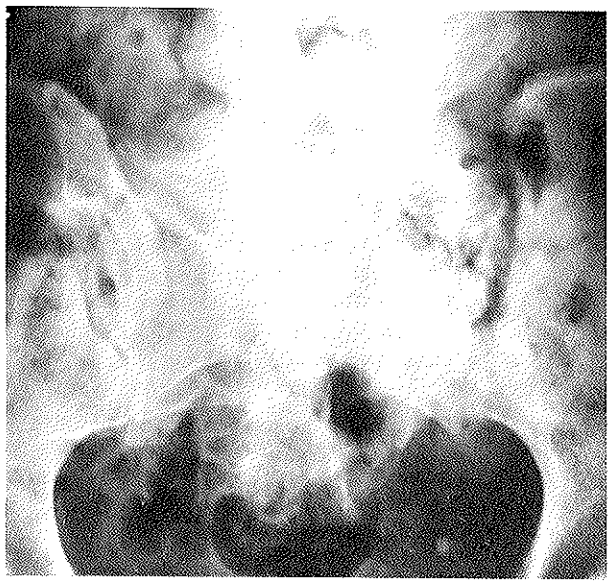


Fig 2c - Pelvic radiograph following curretage of the tumour, liquid N_2 installation and bone grafting



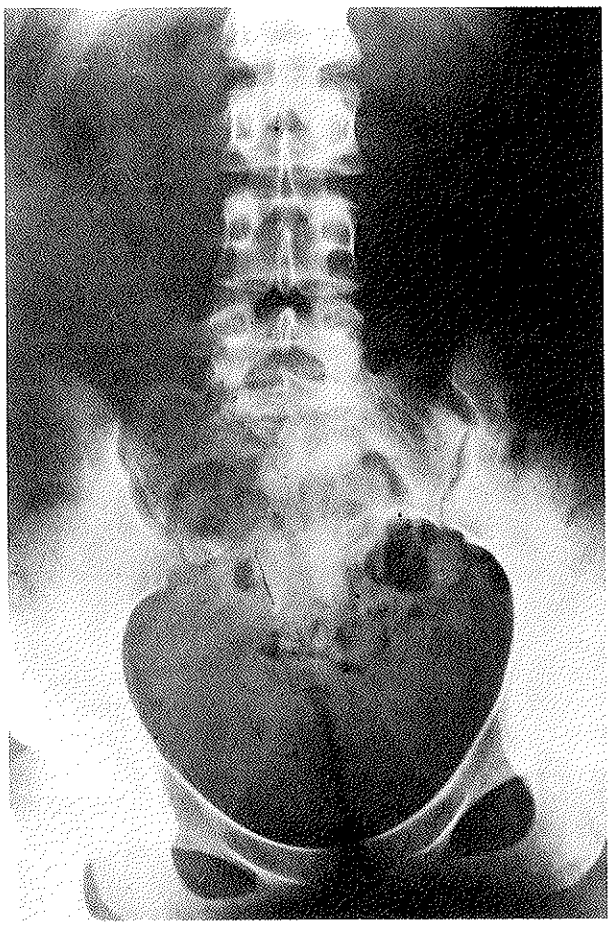
after her currettage procedure, an enlarging lesion was noted in the right ilium. Repeated staging studies were indicative of a recurrence in the ilium with a para-psoas extension (Fig 1f). Following an incisional biopsy which confirmed the presence of giant cell tumour, she had a wide psoas excision with an extended currettage procedure to the ilium and sacrum supplemented by liquid nitrogen instillation and allograft bone grafting. She is also being considered for adjunctive radiation therapy to reduce the likelihood of local recurrence. There has been no progression of her metastatic lung lesions on re-staging studies.

Case 2:

This was a 31-year-old male pilot who presented with a worsening long standing back pain associated with left sciatica. He had a limited straight leg raising test but no neurological deficits. Repeat X-rays showed a destructive lesion affecting the left side of the upper sacral vertebra (Fig 2a). Staging studies included CT Scans (Fig 2b), Bone Scan and angiography. Needle Biopsy showed a giant cell tumour. Surgery first involved ligation of the internal iliac vessels via a retroperitoneal approach followed by a posterior approach to the sacrum. The tumour was curretted out and the sacral nerve roots dissected free. Liquid nitrogen was used with 3 freeze-thaw cycles. This was followed by bone grafting with autogenous and allograft cancellous bone (Fig 2c).

He made a good recovery with mild hyposthesia over the left leg and foot. An additional bone grafting procedure to augment the bone fusion was necessary as well as a biopsy of a suspicious area on re-staging studies which was negative for tumour. Thus, he remains disease-free 2 years following surgery and has returned to flying.

Fig 3a - Case 3 lumbo-sacral radiograph showing upper sacral destruction of the tumour



Case 3:

This was a 17-year-old Malaysian female who presented with gait abnormality and right thigh tightness. She had a X-ray done in her home town which showed a sacral tumour (Fig 3a). Following biopsy, a diagnosis of GCT versus Aneurysmal Bone Cyst was made. She was then referred here for treatment. Examination revealed no neurological deficits. CT Scan and MRI Scan showed involvement of the entire upper sacral vertebra (Fig 3b). CT chest was normal. Following pre-operative embolization of the feeding vessels, a transperitoneal approach was used to currette out the tumour. The upper sacral nerve roots were dissected out from the tumour. Liquid nitrogen was used as an adjuvant.

Autogenous and allograft bone were used to fill the bony defect (Fig 3c). Post-operatively, she had transient bowel and bladder dysfunction till the time of her discharge from hospital 3 weeks later. She had a wound haematoma requiring surgical evacuation. She also had superficial skin necrosis as a result of thermal insult from the liquid nitrogen. She is being followed up by her local physician and has apparently recovered sphincteric function and is disease-free 2 years following surgery.

DISCUSSION

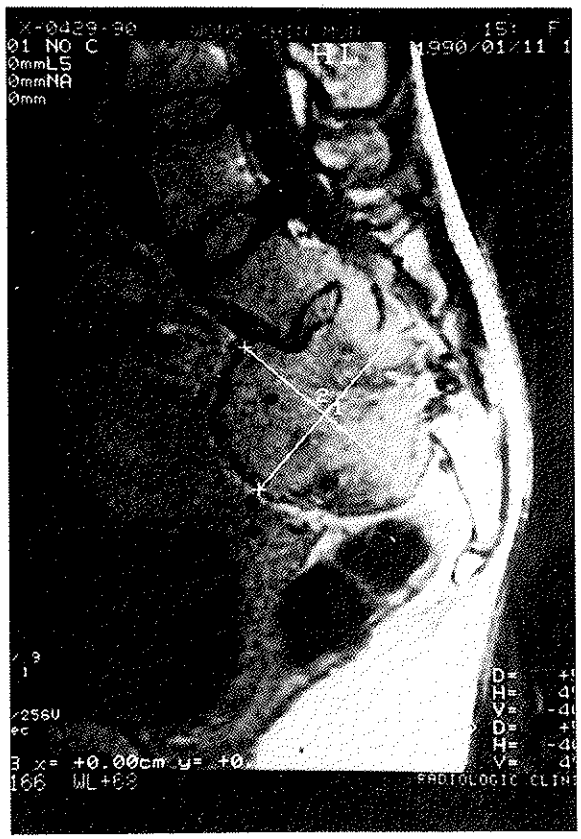
The surgeon treating giant cell tumours of the sacrum is faced with the following problems:

1. Establishing the diagnosis,
2. Complete removal of the tumour,
3. Protection/preservation of the sacral nerve roots and sphincteric function,
4. Stabilization of the vertebral column.

Establishing the diagnosis

Although the sacrum can be affected by almost any tumour, the common lesions to rule out would be giant cell tumour, chordomas, aneurysmal bone cysts, myelomas, bone sarcomas and secondaries to the bone[6]. Proper staging studies should be

Fig 3b- Sagittal MRI of Case 3 showing the large presacral extension of the tumour



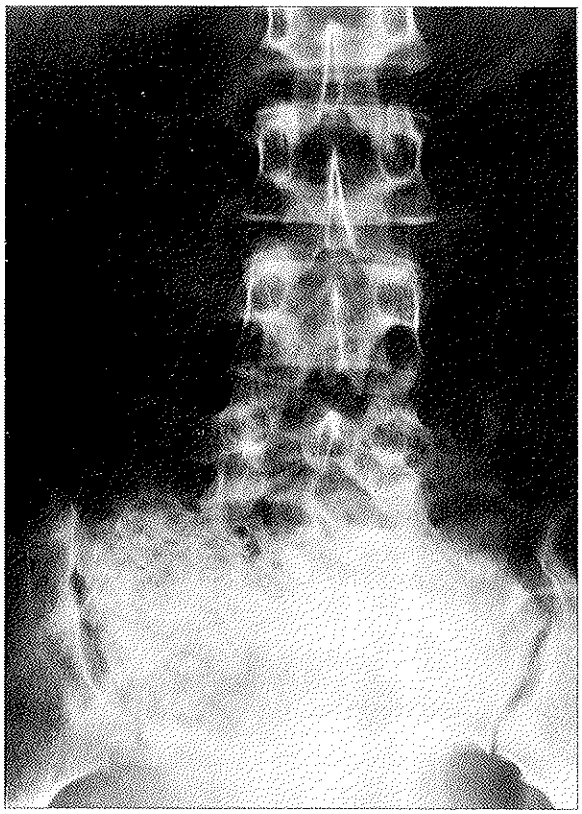
performed following clinical evaluation as the methods of treatment vary tremendously. Minimal investigations would include plain radiographs, chest X-rays, bone scans, CT Scans and blood biochemistry to rule out hyperparathyroid disorders and myeloma. CT Scan may be done with bowel contrast to delineate the position of the tumour with respect to the rectosigmoid colon. Of late, the MRI Scan has been used with increasing effectiveness in visualizing the soft tissue extension of the tumour. A needle biopsy under X-ray/CT control is useful, failing which a carefully planned incisional biopsy is performed. It should be the aim of the surgeon to excise the biopsy scar during the definitive surgery, hence the need of planning the biopsy portal[7].

Complete Removal of the tumour and preservation of the sacral nerve function

It is the fundamental aim of surgery to achieve the former while trying to satisfy the latter.

Most authors feel that whenever surgical removal of a GCT is possible, it is the method of choice. As to the method, ie resection vs curretting, Eckardt summed up current thinking by saying that the bad reputation of currettage is undeserved and arose because of the indiscriminate application of this technique to lesions irrespective of their surgical stage. Good results may be expected when applied to Stage 1 or 2 lesions. Repetitive freezes with liquid nitrogen, though resulting in a lower recurrence rate, carry with them a not insignificant risk of local complications. However, Malawer had low local com-

Fig 3e - Lumbosacral radiograph following currettage of the lesion, liquid N_2 instillation and bone grafting



plications in paediatric patients with bone tumours treated with liquid nitrogen and he attributed it partly to careful attention to the technique of freezing[8].

Stener and Gunterberg have studied patients with radical surgery for tumours with resection of various sacral nerve roots. Their conclusion was that one can sacrifice all sacral nerve roots unilaterally and still preserve sphincteric function. Bilateral sacrifice of the S4 and S5 roots will also allow sphincteric function to be preserved[9,10].

Stener and Gunterberg have also studied pelvic strength following sacral amputations and found that amputation through the S1 canal weakens the posterior arch of the pelvis by 1/3 while amputation above the S1 root canal weakens the arch by 1/2. There is, however, sufficient strength for normal load bearing even after such amputations[11,12].

While such studies are available to guide surgeons, in our 3 patients with giant cell tumours, the extent of the tumour meant sacrifice of sphincteric innervation if surgical resection was performed. Moreover, in one patient, the presence of multiple metastases indicated a more conservative approach. Thus, the two patients with a localized tumour had currettage with liquid nitrogen treatment and remain disease-free. As GCTs are very vascular tumours, it is often prudent to either embolize the feeding vessels or to ligate the internal iliac vessels prior to surgical currettage of the lesions.

An alternative form of treatment is the use of radiation therapy in tumours that are not amenable to complete surgical resection. Proponents for radiation therapy quote excellent results with better than 85% local tumour control using radiation therapy alone[13].

Other authors recommend radiation therapy as a post-operative adjuvant (Eilers, Sung, Seider, Zhi)[14-17].

The opponents of radiation therapy will cite the risk of sarcomatous transformation following therapeutic doses of radiation (Senegas, Rock, Krebs)[18-20]. Such radiation induces sarcomas that are almost uniformly fatal. It is probably best to utilize radiation therapy on a case-to-case basis taking into account the patient's extent of disease, tumour biological behaviour, age group and personal wishes based upon informed consent.

Finally, the rare case of giant cell tumour presenting with pulmonary metastases remains a difficult management problem. Bertoni reviewed 39 cases of GCT with lung metastases from world literature and concluded that there is a better than 85% survival rate after an average of 8.2 years follow-up[1]. Patients were treated with various combinations of thoracotomies, chemotherapy and radiation therapy. It was recommended that lesions, if amenable, be treated surgically.

The histology yields no clues as to why some Giant Cell Tumours metastasize while others do not as the histology of the lesions that metastasize are identical to lesions that do not metastasize[3,4]. While chemotherapy is of no proven benefit to the bony lesions of GCT[20], its efficacy in patients with pulmonary metastases is too occasional to be more than anecdotal.

CONCLUSION

GCT is an enigmatic disease with a wide variation in presentation. Its classification as a benign tumour is sometimes muddled by multicentric presentations or the occasional metastasis. Its management thus requires the surgeon to consider several treatment options in dealing with the tumour.

Although our series is small, our patients all had a large presacral soft tissue component, extensive involvement of the sacrum, the adjacent ilium and in two patients the L5 vertebral body. This makes wide resection and the subsequent reconstruction very difficult. Our limited experience suggests that the use of currettage and adjunctive liquid nitrogen therapy can be considered as an alternative modality of treatment to wide resection surgery in the treatment of giant cell tumours of the sacrum.

References

1. Bertoni F, Present D, Sudanese A, Baldini N, Bacchini P, Campanacci M. Giant Cell Tumour of Bone with Pulmonary Metastases. CORR 1988; 237: 275-85.
2. Verbiest H. Giant Cell Tumours and Aneurysmal Bone Cysts of the Spine. J Bone Joint Surg 1965; 47B(4): 699-713.
3. Gunterberg B, Stener B. High Amputation of the sacrum for Extirpation of Tumours. Spine 1978;3(4):351 - 66.
4. Dahlin D, Unni K. Bone Tumours 2nd Edition. Illinois: Charles C Thomas 1986: 119-40, 337-45.
5. Enneking WF. Clinical Musculoskeletal Pathology. 3rd Revised Edition. University of Florida Press, JHM Health Science Center, 1990; 312-3, 451-66.
6. Eckardt JJ, Grogan JJ. Giant Cell Tumour of Bone. CORR 1986; 204: 45-58.
7. Enneking WF. Musculoskeletal Tumour Surgery 1st Edition. New York Churchill Livingstone. 1983: 1435-76
8. Heare T, Enneking W, Heare M. Staging Techniques and Biopsy of Bone Tumours. Orthop Clin North Am 1989;20(3):273-85.
9. Malawer MM, Dunham W. Cryosurgery and Acrylic cementation as Surgical Adjuncts in the treatment of Aggressive (Benign) Bone Tumours. CORR 1991; 262: 42-57.
10. Gunterberg B, Kewenter J, Petersen I, Stener B. Anorectal Function after major resections of the sacrum with bilateral and unilateral sacrifice of sacral nerves. Br J Surg 1976 63: 546-54.
11. Gunterberg B, Norlen L, Stener B, Sundin T. Neurologic Evaluation after Resection of the Sacrum. Investigation Urology 1975; 13(3): 183-7.
12. Gunterberg B, Romanus B, Stener B. Pelvic Strength After Major Amputation of the Sacrum. Acta Orthop Scand 1976; 47 635-42.
13. Schwartz LH, Okunieff PG, Rosenberg A, Sim HD. Radiation Therapy in the treatment of difficult giant cell tumours. Int J Radiat Oncol Biol Phys 1989; 12: 1085-8.
14. Eilers H, Habighorst LV, Albers P, Rebmann D: Radiation Therapy of an Inoperable Giant Cell Tumour. Strahlentherapie 1977; 153(2): 103-5.
15. Sung HW, Shu WP, Wang HM, Yuai SY, Tsai YB. Surgical Treatment of primary tumours of the Sacrum. Clin Orthop 1987; 215: 91-8.
16. Seider MJ, Rich TA, Ayala AG, Murray JA. Giant Cell Tumours of Bone: Treatment with Radiation Therapy. Radiology 1986; 161(2): 537-40.
17. Zhi XC, Da ZG, Zi HY et al. Radiation Therapy of Giant Cell Tumour of Bone: Analysis of 35 patients. Int J Radiat Oncol Biol Phys 1986; 12: 329-34.
18. Senegas J. Extensive Resection of primary malignant tumours of the sacrum. Neurochirurgie 1989, 35(5): 337-41, 353-4.
19. Rock M, Sim F, Unni K et al. Secondary Malignant Giant Cell Tumour of Bone. J Bone Joint Surg 1986; 68A(7): 1073-9.
20. Krebs B, Baca I. Tumours of the sacrococygeal region. Arch Orthop Trauma Surg 1979; 95(3): 187-97.