Giant cell tumor of the sacrum

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Giant cell tumor (GCT) is a locally highly aggressive tumor of bone comprising 5 to 10% of all benign bone tumors. The sacrum is the third most common site of involvement. Patients with sacral GCTs present with localized pain in the lower back that may radiate to one or both lower limbs. Vague abdominal complaints and bowel and bladder symptoms may also be present. Neuroimaging workup should include advanced modalities, preferably magnetic resonance imaging, prior to obtaining a biopsy specimen. Giant cell tumor has a 1 to 5% rate of metastasizing to the lung and may convert to a fulminate malignant variant, which has a very poor prognosis. The standard treatment for GCT is curettage combined with adjuvant bone grafting or cement-augmented stabilization. In appropriately selected cases, sacral resection is a valuable procedure to effect local tumor control and overall survival. Embolization may also prove palliative and/or curative in cases in which the tumor is unresectable or refractory to treatment.

KEY WORDS • sacrum • giant cell tumor • sarcoma

SACRAL GIANT CELL TUMORS

Presenting Signs and Symptoms

When GCTs are involved with the sacrum, patients present with localized low-back pain that may radiate to one or both lower limbs. Neurological symptoms, if present, are often subtle. Vague abdominal discomfort, early satiety, and a change in bowel/bladder habits are possible. The onset of symptoms is generally insidious; the patient most frequently complains of a slowly progressive problem over several months.

Diagnostic Workup

After obtaining a thorough patient history, the physician should conduct a complete physical examination including abdominal, neurological, spine, and rectal vault assessment. Plain radiography frequently fails to reveal the true extent of the disease (Fig. 1 upper). Axial CT scanning (Fig. 1 center) or, preferably, magnetic resonance imaging (Fig. 1 lower) is necessary for a full evaluation of the anatomical characteristic of the tumor and its invasion into surrounding structures. Bone scintigraphy demonstrates nonspecific increased activity but is recommended in the workup and staging of GCT as well as other aggressive sacral tumors to evaluate noncontiguous disease. Multilevel spinal involvement has been reported in cases of GCT. Furthermore, these lesions have been reported as a purely soft-tissue mass involving only the sacral canal without osseous involvement.

A staged biopsy procedure is appropriate. Pathological evaluation of the tissue specimen will reveal a large number of osteoclastic giant cells, admixed intimately with clusters and a scattered population of mononuclear cells. Mitotic figures are generally absent (Fig. 2).
Behavioral Features

Giant cell tumor has a 1 to 5% incidence of metastasizing to the lung. Recurrent tumors are at a significantly increased risk. Accordingly, pulmonary staging is an important component in the initial and follow-up evaluation of GCT of bone. Generally, lung metastases are late onset; their mean interval to progression is 4 years. The prognosis for survival when lung metastases develop is favorable in more than 70% of patients. Although surgical management of the metastatic lesions is the mainstay of treatment, some metastatic foci may resolve spontaneously. Nevertheless, in such cases approximately 15% of patients with metastatic disease may die (Fig. 3).13

Giant cell tumor can convert to a fulminate malignant variant associated with a very poor prognosis.2 In such cases the disease can become widely metastatic, spreading to locations beyond the lung, including bone, liver, spleen, and other viscera. Some authors have speculated that this is caused by radiotherapy. A conversion rate of 15 to 20% has been reported in patients in whom radiotherapy had involved greater than 3000-cGy doses of radiation, and conversion occurred 3 or more years after treatment. The conversion rate in patients who have not undergone irradiation is less than 5%. This has come into question with newer radiotherapy modalities.3

Treatment Options

The standard treatment for the GCT is curettage and placement of a bone graft or bone cement. Recurrence rates have been reported to be as high as 50% or greater when the intralesional resection is not carefully performed. Follow-up treatment for recurrent disease consists of an aggressive resection and reconstruction involving a large osteoarticular allograft, cementation, endoprosthesi- 

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Fig. 1. Imaging studies obtained in a patient with sacral GCT. Upper Left: Anteroposterior radiograph of the pelvis and sacrum. Upper Right: Lateral radiograph of the sacrum. Although a destructive mass can be identified in the sacrum, its extent is poorly demarcated. Center Left and Right: Axial CT scans demonstrating the cephalad extent (center left) and caudad extent (center right) of the disease. Lower Left and Right: Sagittal (lower left) and coronal (lower right) magnetic resonance images revealing the anterior soft-tissue extension.

Fig. 2. Photomicrograph showing a tissue specimen of sacral GCT. Note the omnipresent multinucleated giant cells and the absence of mitotic activity. H&E, original magnification ×10.

Fig. 3. Axial CT scan of the chest obtained in a patient with advanced metastatic involvement of the lung. Note the bilateral malignant pleural effusions. The patient soon died after this scan was acquired.
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followed by adjuvant phenol, hydrogen peroxide, liquid nitrogen, or argon beam therapy. Cementation may serve as a local adjuvant while also restoring mechanical integrity to a compromised region, facilitating spinal and/or pelvic fixation. In the sacrum, adjuvants must be used with caution to minimize inflicting trauma on the sacral nerve roots. Nerve root monitoring may be appropriate.

When GCT involves an expendable bone such as the proximal fibula or ilium, it should be primarily resected; however, this is generally not recommended for the sacrum. En bloc resection continues to be performed for multiple recurrent tumors, intensive soft-tissue involvement, or massively destructive lesions.

In appropriately selected patients, sacrectomy is a valuable procedure to effect local tumor control and overall patient survival, despite potential complications as well as neurological and sexual dysfunction (Fig. 4). If such an operation is undertaken, it may necessitate an interdisciplinary surgical team including orthopedic oncology, general surgery, and spine surgery depending on the extent of the resection. Preoperative embolization should be considered because these tumors are highly vascular (Fig. 5). A combined anterior–posterior approach is necessary for tumors with anterior extension and/or significant involvement of the S-1 and S-2 segments. Reconstruction involving segmental spinal instrumentation and an allograft or prosthesis may be necessary after total sacrectomy (Fig. 6). An alternative to a conventional transperitoneal anterior approach is the extensile ilioinguinal approach, which permit simultaneous visualization of the anterior and posterior sacrum when combined with a posterior midline approach.

Embolization may also prove palliative and/or curative in cases in which the lesion cannot be resected or in which the disease is refractory to other treatments; the risk of local recurrence is equal to 31% at 10 years and 43% at 15 and 20 years. In cases of advanced multiple recurrent or aggressive metastatic disease, investigators are developing experimental medical protocols. Close follow-up evaluation for locally recurrent disease and pulmonary involvement is critical. Surveillance should include radiographic examination of the chest every 6 to 12 months for at least the first 2 to 3 years.

Fig. 4. Partial sacrectomy at S-3. Upper: Intraoperative photograph. The bilateral S-2 nerve roots were spared, and the patient experienced minimal bowel and bladder dysfunction. Center: Intraoperative photograph showing a rectal patch in place to prevent rectal prolapse. Lower: Postoperative radiograph demonstrating the preservation of S-1 and S-2 segments.

Fig. 5. Preoperative angiograms demonstrating embolization of sacral GCT. Left: Tumor blush prior to embolization. Right: After embolization, flow is diminished but still present.

Fig. 6. Anteroposterior radiograph revealing reconstruction of region after total sacrectomy.
References


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