Review of Sacrococcygeal chordoma

Pandey S

Department of Orthopaedics, Chitwan Medical College Teaching Hospital , Bharatpur

ABSTRACT

Chordomas are very uncommon malignant bone tumours derived from the primitive notochord and preferentially located in the sacrococcygeal region, with slow but progressive infiltration9 of the nerve roots and neighbouring soft parts. **.**Local invasiveness and destructiveness are characteristic features of the disease. Clinical history and examination combined with imaging work up, mainly MRI, provides with the diagnosis most of the time. Incisional biopsy from posterior approach can confirm the diagnosis in case of doubt with other differential diagnosis, commonly Giant cell tumor. Surgical excision along with tumor free margin through posterior approach or combined anterior and posterior approach is the mainstay of treatment. Radiotherapy is supplemented whenever margin is positive or in doubt to increase the duration before recurrence. Due to its local invasiveness to adjacent soft tissue and neural structure, obtaining tumor free margin is difficult. This is the most important prognostic factor which decides local recurrence and survival.

Key words: Sacrococcygeal chordoma, malignant tumor, resection, radiation therapy, local recurrence

INTRODUCTION

Chordomas are very uncommon malignant bone tumours derived from the primitive notochord and preferentially located in the sacrococcygeal region, with slow but progressive infiltration of the nerve roots and neighbouring soft parts. Because of their location they are difficult to manage. In 1857, Virchow originally described chordomas when he named them ecchondrosis physaliphora, believing they were cartilaginous in origin. It is an uncommon tumour of the human skeleton, and has an incidence of 1% of all malignant bone neoplasms.[1] Approximately 50% of chordomas are sacrococcygeal in origin. Chordomas account for more than 40% of all sacral tumours and should be included in the differential diagnosis of sacral tumour, particularly in the older age group. Men and boys are affected more frequently than women and girls, with a ratio of about two to one. The tumor occurs predominantly in the fifth to seventh decade of life, although it has been reported in infants and in the elderly also. A younger mean age in female patients has been reported.[2]

Chordomas are considered low-grade malignant lesions and are slow to metastasize. They are malignant by position, in that their proximity to the cauda equina makes them extremely difficult to treat effectively. Local invasiveness and destructiveness are characteristic features of the disease. The indolent nature and unpredictable behaviour of sacrococcygeal chordomas make early detection and treatment difficult. By the time the diagnosis is established, the tumour is usually very large. Complete surgical excision is the only and mainstay of treatment. Local recurrence results in tissue destruction and generally is the cause of death. Metastases are recognized but are uncommon.

Symptoms are often vague and indolent and can be confused with those of a variety of other conditions, leading to a delay in diagnosis. Sacral chordomas often reach a large size before diagnosis, and their poor margination and the complex pelvic anatomy make treatment difficult. Achieving long-term survival remains a challenge and is probably influenced by local tumor control.[3,4] Still sometimes, chordomas are excised with curettage or are biopsied through the

rectal wall, procedures that compromise outcome. Chordomas are insensitive to chemotherapy, and complete surgical excision is the mainstay of treatment. [5] However, neurogenic dysfunction and pelvic instability are frequent consequences of adequate surgical removal and these complications correlate with the level of resection. A radical surgical approach therefore remains controversial.

APPLIED ANATOMY

The pelvic surface of the sacrum gives attachment to the piriformis muscles with the first three sacral ventral rami running along the anterior surface. The gluteus maximus muscles are inserted into the dorsal surfaces of the lower sacrum and the coccygeal body. The dorsal surface of the sacrum gives attachment to the erector spinae muscles. Situated at the root of the sigmoid mesentery and on either side of the midline are the paired internal iliac arteries and veins. In axial sections through the second sacral segment, the iliac vessels are medial to the iliopsoas muscles and anterior to the lumbosacral trunk. Identification and clear definition of these structures is important, as the iliac arteries are ligated in the course of accessing the sacral tumour through a lower anterior abdominal surgical approach.

The presacral fascia is the caudal continuation of the anterior longitudinal ligament. The presacral fascia is described by Yonemoto et al as a strong structure,resistant to tumour invasion. The low signal of the fascia is due to its increased fibrous content. The periosteum and presacral fascia are tough membranes that resist transgression by tumor, so the tumor rarely invades the rectal wall. It has been pointed out that one of the cardinal errors in management is a transrectal biopsy, a procedure that violates these containing membranes and the rectal wall and thus often makes it necessary to resect the rectum at the time of the operation.

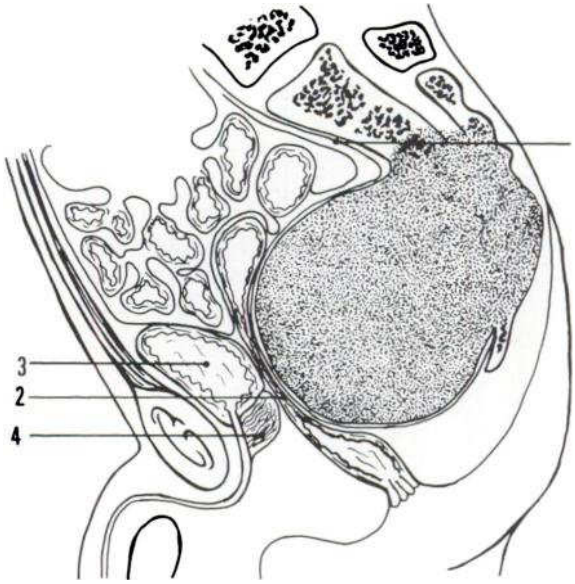

Fig. I
To show the relation between a typical chordoma and the surrounding viscera. 1, presacral lamina ; 2, rectum ; 3, bladder; 4, prostate.

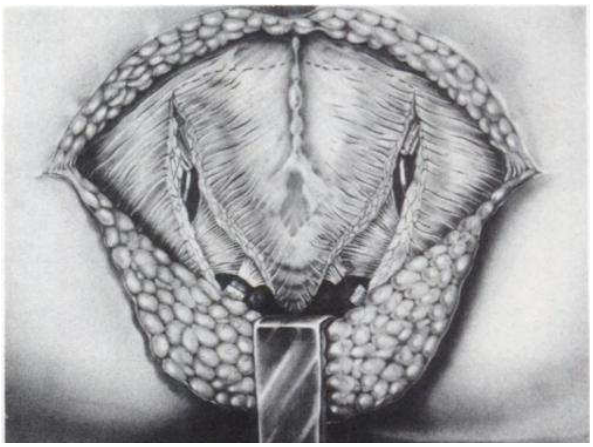

Fig II.
The appearance of the sacrum after division of gluteus maximus and the sacrotuberous ligaments; the sacrospinous ligaments are seen; above and lateral to them are the S2 nerve roots

PATHOLOGY

Macroscopically, sacrococcygeal chordomas are usually well-demarcated by a pseudocapsule and range from 3 cm to 20 cm in diameter (median 8 cm).[4] Virtually all tumours involve bone, with extension into adjacent soft tissue and skeletal muscle. The cut surface of the tumour is characteristically soft, gelatinous, mucoid, and haemorrhagic. Occasional greyish-yellow, friable foci are observed, probably secondary to necrosis.

Characteristic histological features include a mixture of epithelioid and physaliferous cells. The latter are typically large with vacuolated cytoplasm and prominent vesicular nuclei. A sea of muco-myxoid material surrounds these cells. The tumour cells are frequently arranged into lobules, separated by fibrous tissues. Mitotic figures are scant or absent. Areas of cartilage and bone may be present.[4] These histopathological features form the basis of the variable and heterogeneous appearances shown in T1- and T2-weighted MRI sequences.

CLINICAL PRESENTATION

The clinical presentation is entirely dependent on the location of the chordoma. At the sacrum, common presenting symptoms are back and/or lower extremity pain. Some present with bony prominence over the sacrum region. About one half of patients with chordomas have autonomic symptoms, particularly rectal dysfunction or urinary incontinence associated with motor and sensory impairment of lower limb over the invovlved nerve roots. About one half of patients with chordomas have a palpable sacral mass. Patients with tumors located along lower vertebrae may present with pain, bladder dysfunction, or lower extremity weakness. The time span from the onset of symptoms to diagnosis averages 10 months.

DIAGNOSTIC WORK UP

IMAGING STUDIES

Plain radiographs will show the amount of bone involvement. Plain-film may show an ill-defined endosteal margin or a bulky mass in the soft tissue. The lesions also may be lytic. The mass

appears as a destructive well-demarcated lesion. The discovery of these features can better clarify the diagnosis of chordomas in the differential of bony lesions.

MRI is better than CT scan in evaluating tumor extent into soft tissue and is choice of imaging modality. Knowledge of the extent of the tumor is important in planning the optimal surgical approach. With CT scans, chordomas at any site appear as single or multiple areas of decreased attenuation vertebrae, or sacrum. Fingers of low density radiate throughout the mass and into the adjacent tissues. If the chordoma has a significant chondroid component, focal regions of hyperdensity may be present. The lesions are expansile with destructive or lytic lesions in the bone. On MRI, the appearance of a chordoma is similar to the appearance on CT scan, with better resolution of the soft-tissue component, resulting in better anatomical definition.

The classical features of sacrococcygeal chordoma are that of a soft tissue mass with aggressive destruction of bone, and invasion of adjacent soft tissues and neurovascular structures in the pelvic cavity.[1,2] These features are pronounced in recurrent disease. Intratumoural calcifications are observed on CT n almost 90% of cases.[5] On MRI, the lesion is of intermediate to low signal intensity in T1-weighted images and high intensity in T2-weighted images. In T2-weighted sequences the lesions are usually of heterogeneous intensity caused by the presence of internal septations.

DIAGNOSTIC PROCEDURES

Biopsies of chordomas are useful only when other bone lesions remain in the differential diagnosis after imaging studies are performed. In this instance, tissue diagnosis by biopsy can enable optimal planning for surgical resection of the tumor. Fine needle aspiration (FNA) is the preferred method for establishing the preoperative morphologic diagnosis of chordoma and has been reported to lower local recurrence rates when compared with open biopsy.[2] The diagnostic criteria for chordoma in FNA include the presence of physaliphorous cells with round nuclei, bland chromatin and distinct cytoplasmic borders in a background of abundant myxoid ground substance.

HISTOLOGIC FINDINGS

Microscopically, chordomas are composed of uniform cells with small oval or round eccentric nuclei and dense chromatin. The hallmark microscopic features of chordomas are the numerous, variably sized vacuoles located in the tumor cell cytoplasm, the physaliphorous cells. Some tumor cells may have more solid or eosinophilic cytoplasm.
Various histologic growth patterns can be seen in chordomas. The cells may be arranged in a diffuse or lobular pattern, or they may be clustered in groups or islands in a sheetlike pattern. Areas of tumor cells may be seen in a solid, perivascular, or even ribbonlike pattern. Between the cells or clusters, an abundant basophilic-to-metachromatic mucinous matrix exists. Mitoses, foci of pleomorphic cells, or focal hemorrhage rarely can be seen but are not prominent features.
Fibrous tissue surrounds the neoplasm and extends projections into the tumor, usually without forming a true capsule.

With specialized histochemistry, chordoma tumor cells tend to be periodic acid-Schiff (PAS) positive. The matrix stains diffusely with mucicarmine and Alcian blue, and it stains metachromatically with toluidine blue; it is negative with Sudan black.

DIFFERENTIAL DIAGNOSIS

Giant cell tumor is the most common differential diagnosis of bony tumor arising from sacrococcygeal region next to chordoma. Other lesions that can give rise to similar pictures are

chondrosarcoma, ependymoma, plasmacytoma, and a solitary metastatic deposit.9 On MRI, these neoplasms may have similar signal characteristics to chordoma. However, calcification on CT, origin at the sacrococcygeal junction, and the presence of internal septations are features that distinguish chordoma from other sacral neoplasms.9

STAGING

Chordomas, like other bone tumors, have been subject to staging methods. Studies analyzing the prognosis and outcome in comparison to stage have not proven to be very useful. The local extent and degree of resection are much more important to the prognosis of a chordoma.

TREATMENT

MEDICAL THERAPY

Clinical trials are underway to study the effectiveness of imatinib mesylate in the treatment of chordoma (Casali et al., 2004). Imatinib mesylate is a tyrosine kinase inhibitor targeting several enzymes including platelet-derived growth factor receptor (PDGFRB), which can be expressed in chordomas. This drug has been shown to have antitumor activity in chordomas; however, research is ongoing and surgery remains the standard treatment for chordoma. Adjuvant radiation therapy is used in cases where incomplete resection is suspected. Chemotherapy has not been shown to be effective.

SURGERY

This is the mainstay of treatment for sacrococcygeal chordoma**.** In general, a more complete removal with wide excision delays the time interval between surgery and eventual recurrence. The natural history and the effectiveness of different kinds of therapy are not well understood in chordomas because of their rare incidence and slow-growing nature. Radical resections of tumors with clean margins are associated with a longer disease-free interval. If subtotal excision is the only option (generally due to location and proximity to delicate anatomy), the addition of radiation therapy can lengthen the interval to recurrence.

Removal should aim to be extensive and complete because, if not, recurrence is certain. The ideal location for sacral dissection, whenever possible, is the S2-S3 junction (preserving the S2 roots) in order to avoid incontinence. If both S2 roots are dissected, incontinence is unavoidable. When complete excision is not possible, high doses of adjuvant radiation therapy must be administered postoperatively to reduce the chance of local recurrence. Chance of local recurrence depends on respectability of the tumor and histology, those with chondroid differentiation having a better prognosis than those with a dedifferentiated pattern.

APPROACH

There are different techniques to approach this tumor. Commonly practiced approach are posterior and combined anterior and posterior. Some surgeons prefer posterior approach only if the mass is predominantly posterior bulge and combined approach if the tumor is predominantly anterior. There are different studies which have shown variable results of each methods but posterior approach is more easy, less time consuming and likely to be associated with lesser complication rate than the combined approach.
Posterior approach also involves different incision technique like transverse incision with upward arch so that inferior based flap is designed, midline longitudinal associated with more risk of wound infection and anal sphincture damage.

Intraoperatively the evaluation of tumor margins and identification of nerve roots with special care to preserve S2 is essential to assess the status of the resection as the resection proceeds. Knowledge of the completeness of the tumor resection helps predict patient outcome in terms of the length of the disease-free interval and assists in determination of the need for adjunctive therapy such as
radiation.

POSTOPERATIVE PHASE EVALUATION

General postoperative complications relevant to this or any surgery include flap necrosis, wound infection or infection of the operative bed (abscess), shock, pulmonary complications (respiratory failure, atelectasis, infection), and bladder infection or urinary retention. Morbidity from surgery can be very mild or severe following tumor resection. With the resection of sacrococcygeal chordomas, bowel and bladder dysfunction are the most frequent complications. Neurological status should be examined and documented to compare with the preoperative status. Rehabilitation is instituted early to make patient ambulatory with or without support depending upon the neurological status. Bowel and bladder training/care is initiated as per the postoperative functional status of bowel and bladder. Radiotherpy is advised as adjuvant therapy whenever in doubt about the adequacy of surgical margin to reduce the chance of local recurrence and to lengthen duration before recurrence.

FOLLOW-UP AND PROGNOSIS

Frequent and regular follow-up helps to diagnose the condition early as there is high rate of recurrence. Tumor recurrence identified early is easier to treat. The average interval to recurrence is 3.8 years for radically resected tumors, 2.1 years for subtotal resection followed by radiation therapy, and 8 months for subtotal excision without adjuvant therapy. The interval of follow-up, including repeat MRI or CT scans, depends on the completeness of the resection. Because residual tumor drastically shortens the recurrence time, patients with known or suspected residual tumor need to be evaluated more frequently. Complications occur at a higher rate after radical resections than with subtotal resections and depend somewhat on the location of the tumor.

Chordomas are relatively benign-appearing neoplasms; however, because of their tendency to erode bone and invade soft tissues, they usually display malignant behavior. In addition, the location of the tumor influences the ability to achieve complete resection. Chordomas often grow in inaccessible sites, and their margins within soft tissue often are not well defined. As a result, complete excision of chordomas is difficult at best. This is the most important determining factor for poor outcome resulting in high chance of local recurrence. The 5-year survival rate is estimated to be 51%, and the 10-year survival is estimated to be 35%. Factors that may improve prognosis are young age, complete resection, and the addition of radiation therapy in incompletely resected tumors.

DISCUSSION

Chordoma of the sacrum presents a diagnostic and therapeutic challenge. The overall survival of these patients is still relatively poor considering the low-grade nature of this malignant lesion. [1] As noted in the study carried out by Bruno et al the long duration of symptoms prior to diagnosis, the delay in diagnosis, and the large volume of the tumor may contribute to the relatively poor prognosis. Previous studies have shown five and ten-year survival rates of 45% to 77% and 28% to 50%, respectively. [8,9,10] The long term survival rate of the patients in a study shown by Bruno et al compares favorably with the rates in other reports; however, the mortality was still high (>50% at fifteen years). The cumulative probability of local recurrence at five and ten years was

46% and 54%, respectively in his series. The development of metastatic disease has been found to be an indicator of a poor prognosis. Several studies have shown that the risk of metastatic disease ranges from 10% to 40%. [11] Samson et al. reported a ten-year cumulative probability of metastasis of 50%.[9]

Local recurrence is of great concern because of its inverse relationship with survival. [3] There is general agreement that complete surgical resection with wide, tumor-free margins is the treatment of choice for a chordoma. However, the extent of the resection directly correlates with the resulting functional deficits .[12] Therefore, meticulous preoperative planning and discussion of potential neurogenic dysfunction with the patient are essential.

Several surgical approaches can be utilized to remove a sacral chordoma, and it is unclear whether some approaches may increase the probability of obtaining a wide margin. The posterior approach was described as being most appropriate for lesions at S3 and below.[13] This approach offers the advantage of a single operation, shorter operative time, and less morbidity. Disadvantages include a potential for hemorrhage and possible violation of the pelvic viscera or ureters during removal of specimens. For lesions above S3, it is preferred to use the combined anteroposterior approach. The anterior approach allows exposure of the entire sacrum with mobilization of the rectum, ureter, and major vessels. Ligation of the internal iliac arteries may have helped reduce bleeding during mobilization of the specimen from posteriorly. In a study carried out by Bruno et al, the majority of patients (81%) in whom a wide margin was achieved had a combined anteroposterior approach. This finding may help surgeons to decide which approach to use, particularly for lesions at the S2 or S3 level.

Different studies have shown that the single most important predictor of survival and local recurrence was the margin obtained during the surgery. All patients with a wide margin tend to survive longer.

The role of radiation remains a subject of debate, and its value in controlling local disease is controversial. Although chordomas may be relatively resistant to adjuvant radiation, it may prolong the disease-free interval and decrease the symptoms of recurrent lesions.[13] Indications for radiation therapy may include surgically inaccessible lesions, contaminated surgical margins, or incomplete surgical excision of the tumor. Radiation therapy can also be used after removal of the primary tumor or when there is a local recurrence. Previous studies have demonstrated mixed results regarding the ability of early radiation therapy to improve the prognosis when the surgical margin is positive.[1] In the study conducted by Bruno et al, it showed that radiation did not improve survival or disease status. However, fewer than half of the patients in their series received radiation, and, of these, two-thirds received it only for recurrence. The authors of a recent promising report found carbon-ion radiotherapy to be an effective treatment for chordomas.[14]

CONCLUSION

Sacrococcygeal chordoma ,a rare slow growing malignant tumor, presents a difficult diagnostic and therapeutic problem, with a high rate of local recurrence. Earlier diagnosis of chordomas is important and requires a high index of suspicion. The surgical margin is the most important predictor of survival, and aggressive surgery, may help to achieve an adequate margin. Hopefully, one can expect more cures with earlier recognition and aggressive surgical treatment. Adjuvant radiotherapy is still recommended whenever there is inadequate surgical margin or disease is not amenable to surgical excision.

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Correspondence:
Dr Suresh Pandey
Assistant Professor
Department of Orthopedics
Chitwan Medical College Teaching Hospital, Bharatpur
Email: pandeys59@yahoo.com