A CASE OF SACRO-COCCYGEAL CHORDOMA MIMICKING A PILONIDAL SINUS

Mehmud Aurangzeb and Farsheed Sartaj
Department of Surgery,
Khyber Teaching Hospital, Peshawar

SUMMARY

Chordoma is a rare, low grade malignant neoplasms thought to develop from embryonic remnants of the notochord. Pilonidal sinus disease, characterized by communicating granulating natal cleft tracts and abscesses, is a common condition. We report a case of a sacrococcygeal chordoma diagnosed incidentally on surgical exploration of a case of presumed recurrent pilonidal disease.

INTRODUCTION

Constituting less than 3% of primary malignant bone tumors, chordomas are very rare.1 The sacrococcygeal area is the most common site, and patients present with a mass, neuralgic symptoms, or pain.2

CASE REPORT

A 65 years old man was referred from Jalalabad, Afghanistan with a discharging sinus in the natal cleft region and increasing sacral pain. The referral chit mentioned that the patient had undergone an incision and drainage of the lesion, for what appeared to be a pilonidal abscess, 3 months back. No pus was expressed at the time and since the procedure, sinuses had developed at the site.

On examination, a scar and three sinus tracts were observed in the natal cleft region over an underlying mass (Fig-1). On exploration under general anesthesia, a grey, yellowish white firm mass, roughly 15x10 cms was found extending down to the sacral fascia, ischiorectal fossae, and around into the retro-rectal space (Fig-2). After a tedious dissection, the lesion was excised in toto and a primary suture was preformed. Subsequent histopathology revealed the diagnosis of chordoma. The patient made an uneventful recovery, and at one year follow up, there was no evidence of recurrence.

DISCUSSION

This low-grade malignant neoplasm arises in the sacrococcygeal or occipitocervical area and is thought to develop from embryonic remnants of the noto-
Fig. 1: Chordoma seen masquerading pilonidal sinus disease

Fig. 2: The Chordoma specimen after complete excision
As the lesion grows it pushes forward on the rectum, causing constipation, this is followed by sacral nerve sensory loss weakness of the musculature, impotence, saddle anesthesia, and finally loss of bladder and bowel control. The radiographic hallmark of the sacrococcygeal chondroma is its location, which by definition, must include the midline of the anterior body of the segments. Chordomas are always radio lucent and show cortical destruction and poor margination. X-ray may thus show expansion, destruction of bone, trabeculation and calcification. The MRI is the most useful means of defining the nature and extent of these lesion. Differential diagnosis includes plasmacytoma, chondrosarcoma, giant cell tumor and metastatic carcinoma. The tumour is composed of cords and nests of cells resembling chondrocytes, with typical highly vacuolated “basket” or physaliferous cells. The stroma consist of a basophilic mucoid, or myxoid ground substance. The lesion are generally not responsive to radiation or chemotherapy and surgical resection is the treatment of choice. The loca-tion makes wide resection difficult and causes significant morbidity, but without treatment the lesion is uniformly fatal, with late pul-monary metastasis. Some recent evidence suggests that this tumor may be somewhat responsive to proton beam irradiation.

Pre operative radiotherapy followed by local resective surgery (hoping to spare sufficient sacral roots to allow ambulation without support and bladder and bowel function) may reduce the incidence of local recurrence should tumor spill or close margins occur during the surgery.

Pre operative radiotherapy aims to make the tumor cells less implantable, although the incidence of wound complication is increased. Unfortunately most of these lesions are recognized late, treated improperly (many times diagnosed by a transrectal biopsy, necessitating resection of the rectum with the tumor), and the usual course is one of several partial resections, palliative radiation, and finally (quite late in the course) pulmonary metastasis and death. The current five year disease free survival is between 30% and 50%.

This case illustrates and example of a chordoma mimicking pilonidal sinus disease. Malignant degeneration of pilonidal sinus resulting in squamous carcinoma has been reported, but is extremely rare. This case report emphasizes that, albeit rare, alternative pathology should be kept in mind while dealing with pilonidal sinus disease.

REFERENCES