CASE REPORT

DEDIFFERENTIATED CHORDOMA WITH A SARCOMATOUS COMPONENT: AN OVERLOOKED DIAGNOSIS

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A 60 year old male presented with unremitting complaints of acute urinary retention and constipation. Computerized Tomography of the abdomen and pelvis confirmed a sacral mass. The biopsy diagnosed dedifferentiated sacral chordoma; an extremely rare entity, seldom reported and frequently misdiagnosed. We present a unique case report after an extensive review of retrospective literature on dedifferentiated chordomas.

Keywords: Chordomas, Dedifferentiated chordomas, Sacral chordomas, Malignant fibrous histiocytoma

INTRODUCTION

Chordomas are aggressively slow growing, malignant tumours; derived from the notochordal remnants along the neuraxis. The notochord is an ectodermic derivative that defines the midline of the chordate embryo. The growth of the notochord expands at the site of the future nucleus palposus regressing to leave an acellular sheath at the cranial and caudal ends. A negligible quantity of cells, persist in at these sites; which have the highest incidence of chordoma. Despite the fact that the neoplastic change is explicit, chordomal cells resemble notochordal cells morphologically and immunohistochemically such as staining positively for epithelial membrane antigen, S-100 protein positively or cytokeratin.[1,2]

CASE REPORT

A 60 year old male presented at Combined Military Hospital, Rawalpindi with unremitting complaints of acute urinary retention and constipation. There was no prior history of trauma, fever or recent weight loss. Physical examination revealed subtle signs of pallor, the patient being normal otherwise. An X-ray of the pelvis showed a mass in the sacral region. The Ultrasonic investigations showed normal echo-texture of the abdominal viscera.

Computerized Tomography of the abdomen and pelvis confirmed a sacral mass measuring 12.5×8.8 cm, extending through the lesser sciatic notch involving the gluteal muscles of either side and posterior wall of the rectum. Trivial enlargement of the prostate was an asymptomatic incidental finding, unlikely to account for urinary retention (Figure-1).

The biopsy specimen had gelatinous haemorrhagic and bony hard areas on its cut surface. The histology reports diagnosed a dedifferentiated sacral chordoma. A surgical resection of the tumour along the margins of its extent was carried out (Figure-2). A permanent colostomy was done. The patient was post operatively stable and treated with radiotherapy.

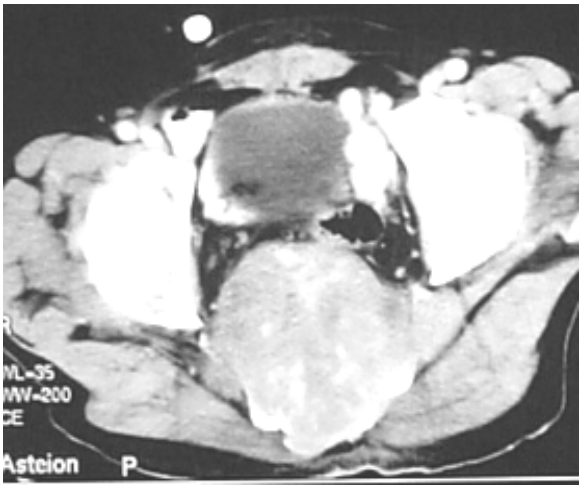

Figure-1: CT scan of the pelvis showing trivial enlargement of prostate and chordoma.

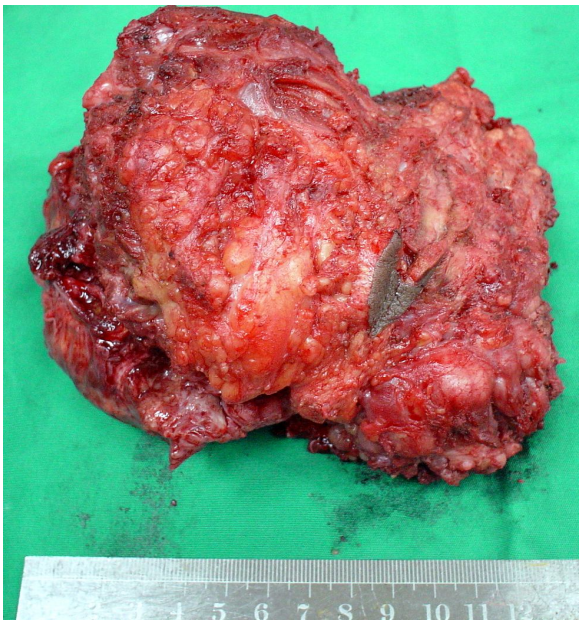

Figure-2: Mass after resection showing measuring scale for size.

DISCUSSION

Chordomas are aggressively slow growing, malignant tumours; derived from the embryonic remnants of the fibrous notochord. They occur spontaneously and have no known aetiology. The clinical presentation of chordomas with the symptom of low back ache is very common but our patient unusually presented with the symptoms of constipation and detrusor areflexia; yet to be reported in medical literature.[1,2]

Chordoma microscopically revealed lobules of atypical cells varying in size and nuclear appearance. Some cells had a vacuolated cytoplasm and physalifrous nuclei while others were inconspicuous. The tumour showed a sarcomatous component comprising of phenomorphic spindle shaped cells forming a vague stariform pattern with frequent mitosis and areas of necrosis and calcification. There was very scanty stromal tissue. Conventional chordomas contain a dominant spindle shaped cell component; hence they are differentially diagnosed as dedifferentiated chordomas or other spindle cell tumours.

Dedifferentiated chordomas with a sarcomatous component are rare bone tumours that have been reported occasionally in the previous literature.[2–4] A retrospective analysis of such reports shows a predisposition for males over 50 years of age, with an increasing occurrence in the sacrococcygeal region.[4] The microscopic features of the tumours mimics malignant fibrous histiocytoma, despite typical areas of conventional form of chordoma in the sacrum. More than one third of the cases of chordoma are often misdiagnosed as fibrous histiocytoma. Dedifferentiated chordomas have been described familiar to osteosarcoma, fibro sarcoma or high grade chondrosarcoma. Inconclusive evidence has linked conventional histological variants transforming to a sarcomatous form. This is common in diagnosed chordoma cases that undergo radiotherapy to regress the tumour. Hence the role of vigilant histopathological diagnoses can not be over emphasized.

The general consensus on the surgical treatment of sacral chordomas resides in surgical resection with wide margins, avoiding as much of functional loss as possible. The major approaches used, include the posterior approach, combined antero-posterior approach, laparoscopic mobilization of anterior structures and an anterior open approach. The posterior approach is commonly used for the lesions occurring at S_3 and inferiorly, while for lesions occurring superior to S_3, the combined antero-posterior approach is considered more practical. Rectus abdominis Flap is used in closing the wound posteriorly, in the anterior open approach. The most frequent complications include local recurrences. The combined antero-posterior approach can be very valuable in obtaining a wider margin and might become an even more accepted approach in the future.[5–7]

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