# SACROCOCCYGEAL CHORDOMA: A CASE REPORT AND REVIEW OF THE LITERATURE

(Received January 2 1991)

O.Büyükgebiz, M.D.*** / Ö.Aktan M.D * / H.H.Döşlüoğlu M.D.***
T.Esemenli M.D.** / F.Söylemezoğlu, M.D.****

* *Associate Professor, Department of General Surgery, Faculty of Medicine,Marmara University, Istanbul, Turkey.*
** *Assistant Professor, Department of Orthopedics and Traumatology, Faculty of Medicine,Marmara University, Istanbul, Turkey.*
*** *Research Assistant, Department of General Surgery, Faculty of Medicine,Marmara University, Istanbul, Turkey.*
**** *Research Assistant, Department of Pathology, Faculty of Medicine,Marmara University, Istanbul, Turkey.*

## SUMMARY

Chordomas are slowly growing malignant tumors arising from notochordal rests. We report a case of sacrococcygeal chordoma treated by sacral resection through posterior approach.

**Key Words:** Chordoma, Sacrococcygeal Tumors, Bone Neoplasm.

## INTRODUCTION

Retrorectal tumors are uncommon lesions. Chordoma is a low grade retrorectal tumor arising from the rests of the foetal notochord (1). Their anatomical location may commonly lead to difficulty in diagnosis. In this paper, a case of sacrococcygeal chordoma is presented and the literature reviewed.

## CASE REPORT

A 45 years old female patient was admitted to Marmara University Hospital with a seven months' history of a growing mass in the sacrococcygeal area and lowback pain. There was no urinary or bowel dysfunction. Physical examination revealed a 6x6 cm palpable mass in the sacrococcygeal region with normal abdominal findings. On rectal examination the tumor was also palpable as an extrinsic mass behind the posterior rectal wall. Laboratory investigations showed a normal blood picture and hepatorenal function. Contrast-enhanced CT scan of the pelvis revealed a large midline soft tissue mass about 7 cm in diameter that destroyed the coccgeal and sacral bones up to S2 level (Fig.1). On exploration, using a midline sacral incision in the prone jack-knife position a well-circumscribed, encapsulated mass was found. The gluteus maximus muscle was infiltrated with tumor tissue on the right side and resected with the tumor and then the sacrum was transected at the level of the S2-S3 interspace. After direct repair of the gluteal muscles skin was closed with a closed suction drainage. There was no blood loss requiring blood transfusion. The patient was discharged on the eighth day after an uneventful postoperative period. Postoperatively 5000 cGy radiotherapy was given to the patient in divided doses.

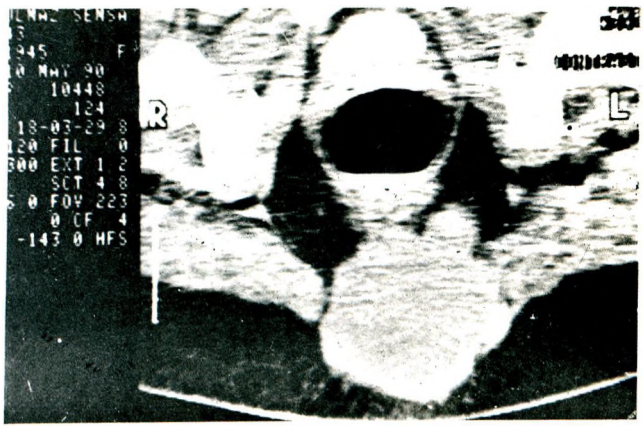

Figure 1 : Computerized tomographic appearence of the sacrococcygeal chordoma

On histopathologic examination, grossly, the specimen was gelatinous, soft and encapsulated, the cut surface revealed areas of hemorrhage and necrosis. Histologic sections were stained with H.E., Alcian Blue pH 2.5 and periodic acid-schift. Microscopically, the tumor was composed of cell cords and lobules seperated by an extensive amount of myxoid intercellular matrix. Some of the tumor cells were large with vacuolated cytoplasm and prominent vesicular nucleus (known as physaliferous cells) where as other tumor cells are small, with small nuclei and no visible nucleoli (Fig. 2).

## DISCUSSION

While some authors consider this a primary bone tumor, chordoma usually is classified as a congenital tumor (2, 3). They occur most commonly in adults (50 to 60 years old) and are mainly located in sacrococcygeal or spheno-occipital regions (85%). Another main localisation is the cervical spine (1). The most common symptom is pain which characteristically aggravetes by sitting or symptoms due to compression of the surrounding viscera may be encountered (3, 4).

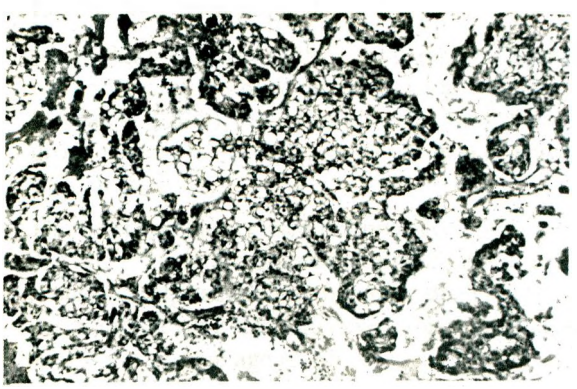

**Figure 2:** Tumor cells with large vacuolated cytoplasm (physaliferous cells) are seen within myxoid matrix. H.E.x 80

Radiologically direct x-rays may display the characteristic signs associated with chordoma namely osteolysis and soft tissiue opacity. In the preoperative staging radiodiagnostic techniques including barium enema and urography are not reliable for definition of margins and soft tissue involvement. Computed tomography (CT) or nuclear magnetic resonance (NMR) is indicated when radical surgery is planned. As in our patient CT may reveal muscle infiltration but usually the rectum is preserved. Plaza et al. (5), emphasized the importance of fine needle aspiration biopsy (FNA) which excludes the other more common neoplasms arising from this area (e.g., chondrosarcomas, liposarcomas). Because of the similar cytologic appearences of these tumors and the brevity of the FNA biopsy experience with chordomas, Jao et al. (4) pointed out that preoperative biopsy is unnecessary in patients about to undergo resection. During surgery the sacrum must be sectioned en block with the tumor based on the preoperative investigations. The division of the muscles must be performed on a safe distance, as chordoma has a tendency of invasion. Sacrococcygeal chordoma is considered operable when the cranial margin of the lesion is contained under or through the second sacral vertebra (6). Resections as high as S1-S2 interspace have been done successfully without any urinary or stool incontinence. Care should be given to preserve the S1 and S2 roots for a full continence. In our patient full urinary and stool continence was achieved after resection at the S2-S3 interspace. Special care was given to resect the infiltrated gluteus maximus muscle on the right side.

On macroscopic examination, the tumoral tissue has mucoid a appearence and histologic examination generally reveals lobules of epithelial appearing cells surrounded by acid mucosubstances. They are surrounded by argyrophylicrim due to pericellular condensation of intercellular matrix , well viewed on electron microscopy. Cells contain glycogen and neutral mucosubstances. Chordoma cells express epithelial differentiation antigens (low mollecular weight cytoceratins, EMA, CAM 52, HFM 62 even CEA), vimentin and S-100 protein. This triple positivity allow differentiation between chordomas and numerous other tumors (7, 8). Chordomas are relatively chemoresistant but cisdichlorodiammineplatinum, vinblastin, bleomycin (PVB) regimen has been found to be beneficial (6). The role of radiotherapy (RT) is still controversial (9) because of the absence of well controlled studies but it seems to be effective as an adjuvant or palliative measure. While studies indicate reduced local recurrance rates after RT our patient received 5000 cGy postoperatively.

The prognosis for chordoma varies greatly. Moreover, cytohistologic features have not yet proved to have any prognostic value. Gray et al. (10) noted that

avarage survival was 5.7 years, while Azzarelli et al. (6) gives 11 years with a possible survival up to 15 years. Local recurrences occur frequently. A chordoma patient should be followed up to 10 years since late local recurrences are not uncommon (11). Our patient is free of any symptoms or signs of recurrence in the first postoperative year.

## REFERENCES


1. *Charpentier Y, Belleqıh S, Boisnic S, Roy Camille R. Chordomes. Ann Path 1988; 8: 25-32.*
2. *Cody HS, Turnbull AD, Fortner JG, Hadju SI, The continuing challenge of retroperitoneal sarcomas. Cancer 1981: 47: 2147-2152.*
3. *Gennarı L, Azzarelli A, Guagliuolo V.A post. approach for the exicision of sacral chordoma. J Bone Jt Surg 1987; 69-B: 565-568.*
4. *Jao SW, Beart RW, Spencer RJ, Reiman HM, Ilstrup DM. Retrorectal tumors. Dis Colon Rect 1985; 28:644-652.*
5. *Plaza et al., Cyto., Cytoch., Ultrast., Diag. of A Sacrococ. Chordoma In A FNA Biop. Rega., ACTA Cyto., Jan. -Feb., 33, 89-92.*
6. *Azzarelli A. et al., Chordoma, Natural History And Treatment. Jour Surg Onco 1988; 37:185-191.*
7. *Ekfors TO. Immunohistochemical differential diagnosis of Chordoma. J Path 1986; 148: 255.*
8. *Salisbury JR, Isaacson PG. Distinguishing chordoma from chondrosarcoma by Histochem. Tech J Path 148 - 351-252.*
9. *Tewfix HH, McGinnis WL, Nordstrom DG, Lotourette HB. Chordoma, Evaluation of clinical behaviour and treatment modalities. Int J Radi Onco Biol Phy 1977; 2: 952-959.*
10. *Gray SW, Singhabhandu B, Smith RA, Skandalakis JE. Sacrococcygeal Chordoma. Surgery 1975; 78: 573-577.*
11. *Michel LA, De Cloedt P. Sync. abdo. and transacral app. for excision of sacrococcygeal chordoma. ACTA Chir Belg 1989; 89: 316-319.*