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# A Case of Sacrococcygeal Chordoma Diagnosed by Fine Needle Aspiration Biopsy Cytology

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A case of sacrococcygeal chordoma diagnosed by fine needle aspiration is presented. This is a case of a 54-year-old woman who came with coccygeal pain of 5-6 months duration. Aspiration biopsy cytology revealed many nests of cells having abundant bubbly cytoplasm and round to oval variably sized nuclei. The cells had indistinct cytoplasmic borders and many of the cells had cytoplasmic vacuoles. The nuclei had thin regular nuclear membranes, finely granular chromatin and one or two small nucleoli. The cells were generally monotonous, but focally pleomorphic with giant cell formation. Mitotic figures were scanty. The background of the aspirate contained abundant mucinous material. These findings were typical of those of recorded chordoma cases and the diagnosis was confirmed by a following open biopsy. The patient received 4,000 rads of neutron radiotherapy and has been well till March '88.

**Key Words**: Chordoma, Aspiration cytology, Sacrococcyx

## INTRODUCTION

Chordoma, a rare tumor, probably arises from remnants of the notochord along the spinal axis. In adults the tumors arise primarily in the sacrococcygeal (50 percent) and spheno-occipital (35 percent) regions. Clinically, chordomas are slow-growing neoplasms that produce first pain and then symptoms of spinal cord compression. There is usually bony destruction radiographically and a palpable mass on physical examination[1].

Since surgical removal of these neoplasms is the treatment of choice[2], preoperative diagnosis would be helpful in planning the procedure. Fine needle aspiration (FNA) of these lesions under radiologic guidance can greatly facilitate planning of optimal surgical treatment.

This paper reports one case of a chordoma of the sacrococcygeal vertebrae diagnosed by FNA. Due to the rarity of this neoplasm, there have been few reported cases of its appearance in fine needle aspirates[3~7].

## CASE REPORT

A 54-year-old woman first noticed coccygeal pain 5-6 months prior to the hospital visit. This problem was slowly progressive. Physical examination revealed no neurologic abnormalities. A computed tomographic (CT) scan showed a large destructive dumbbell-shaped mass in and out of the spinal canal in S3-5 vertebrae. An angiogram showed avascular mass shadow.

A 22-gauge spinal needle was directed into the sacrococcygeal area under radiographic guidance, with negative pressure applied with a syringe. Several smears were made directly from aspirated

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material and syringe-washing sample was made for cell block. All smears were immediately fixed in 95 % ethanol. The ethanol-fixed smears were stained with a conventional papanicolaou stain. Cell block was processed routinely and stained with hematoxylin-eosin, PAS, D-PAS, mucicarmine and several immunohistochemical stainings (EMA, CEA, Cytokeratin, Vimentin and S-100).

Examination of the aspirate revealed findings diagnostic of chordoma. Histologic evaluation of the tissue removed confirmed the presence of a typical chordoma. The patient was recommended surgical treatment, but she refused surgery due to the risk of neurological complication postoperatively. She received 4,000 rads of neutron radiotherapy instead of surgery and has been well till March '88.

## CYTOLOGIC FINDINGS

The aspirates contained many groups of cells with

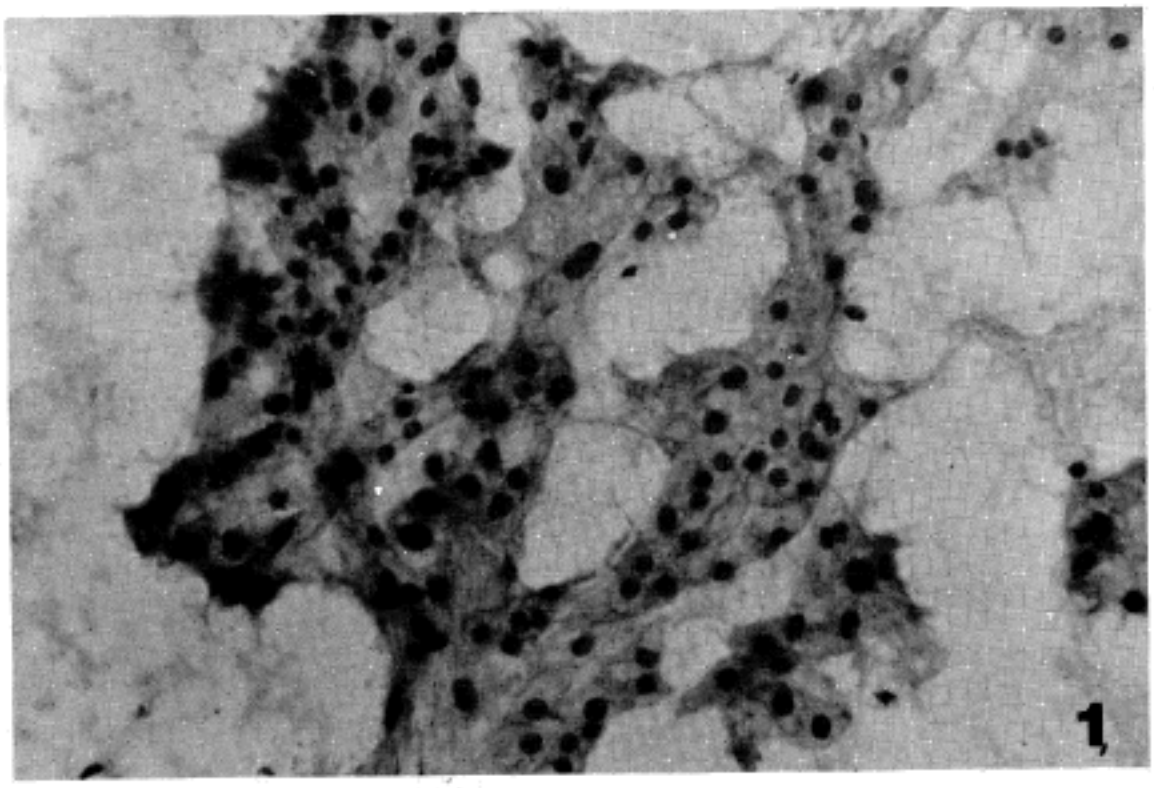

**Fig. 1.** Cluster of cells show indistinct cytoplasmic borders but numerous cytoplasmic vacuoles. A slight nuclear size variability is present. (Papanicolaou stain, X 96).

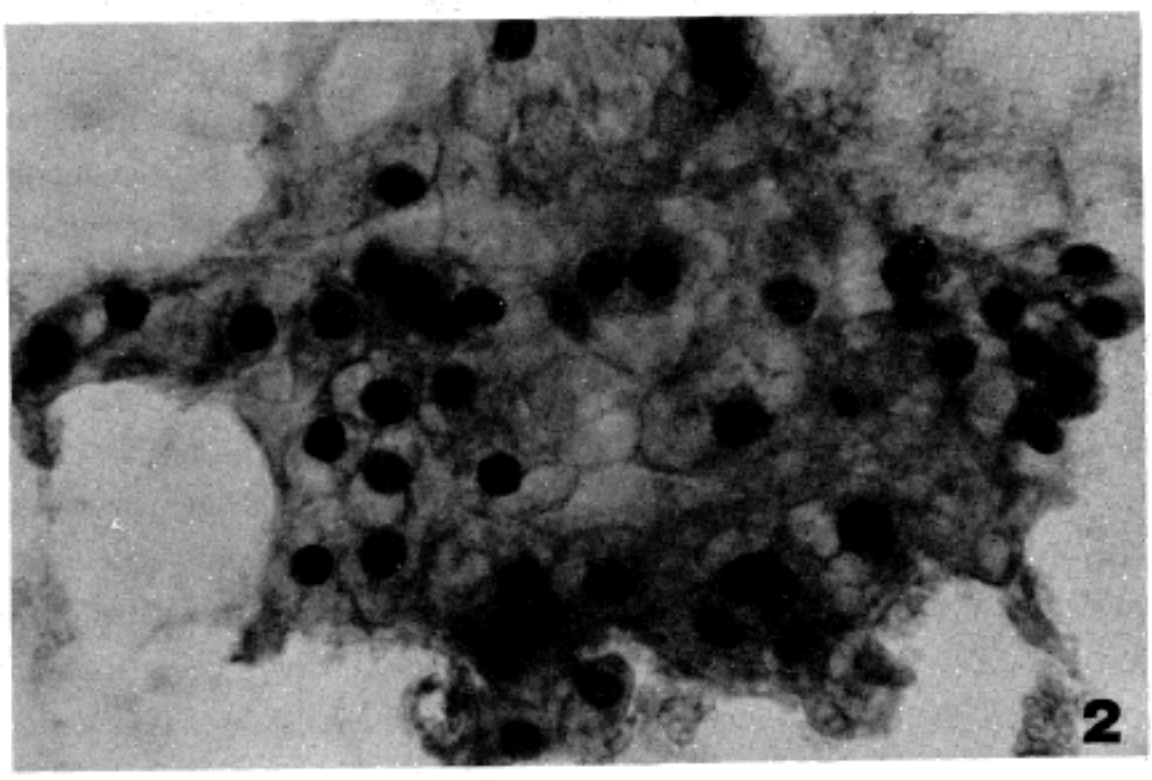

**Fig. 2.** Several physaliferous cells are seen. (Papanicolaou stain, X160).

abundant cytoplasm and oval, variably sized nuclei (Figure 1). The cells in these groupings had indistinct cytoplasmic borders, and many of the cells had a bubbly cytoplasm. The nuclei had a smooth, thin, regular nuclear envelope and a uniformly finely granular chromatin with a single small nucleolus (Figure 2). Many of the individual cells showed similar cytologic features. The background of the aspirates contained abundant mucinous material.

## IMMUNOHISTOCHEMICAL STUDIES

For the immunohistochemical studies, paraffin-embedded tissue was examined by the avidin-biotin-peroxidase complex (ABC) method for the demonstration of epithelial membrane antigen (EMA), cytokeratin, carcinoembryonic antigen (CEA), vimentin and S-100 protein. All primary antibodies were purchased from BioGenex Lab (California, USA). The tumor cells were positively stained for EMA and vimentin, the pattern was focal and weak. The staining pattern for cytokeratin and S-100 was more diffuse and intensive. CEA was negative.

## DISCUSSION

The morphology of chordomas as seen on FNA or touch preparations has been previously described. Cytologically, these lesions can mimic well-differentiated chondrosarcomas or adenocarcinomas, including mucinous adenocarcinomas and renal cell carcinomas.

Chondrosarcomas usually demonstrate definite cartilaginous differentiation, with an abundant matrix material in which the neoplastic cells are arranged in lobules. Chordomas may contain focal areas that are indistinguishable cytologically from chondrosarcomas. Chondrosarcomas do not contain the cells with bubbly cytoplasm (physaliferous cells) seen in chordomas, however. Multinucleated cells can be found in both chondrosarcomas and chordomas and are thus of little help in separating these two entities. The generalized intense staining of chordomas for cytokeratin and EMA obviously separates this tumor from chondrosarcoma.

The main cytologic and clinical differential diagnostic problem with a chordoma is metastatic well-differentiated mucinous adenocarcinoma, particularly those that form signet-ring cells. Chordomas may be differentiated cytologically from adenocarcinomas by several features. (1) While both chordomas and adenocarcinomas contain signet-ring cells, the cells in a chordoma have a flag of cytoplasm between the nucleus and the cell border opposite the large vacuole. This rim of cytoplasm may not be visible in large cell groupings due to the indistinct cytoplasmic borders; the large clear cytoplasmic vacuoles appear to be the cytoplasmic borders in these groups. (2) Chordomas contain cells with numerous cytoplasmic bubbles, the physaliferous cells. Adenocarcinomas may contain clear, mucinous cytoplasmic vacuoles, but, in general, they are not as numerous as in the physaliferous cells. In distinguishing a chordoma from a metastatic renal cell carcinoma, the vacuoles are bigger as well as mucin-positive in the former. The cytoplasmic vacuoles in renal cell carcinoma are small, fine and mucin-negative. (3) Chordoma nuclei have a smooth nuclear border. Even in well-differentiated adenocarcinomas, nuclear irregularities can be found. Overall, the nuclear fetures of a chordoma are benign, with size variation being the one feature suggesting malignancy. The immunohistochemical studies have not been useful for differential diagnosis, since adenocarcinomas show a pattern of staining very similar to that of some chordomas.

Bubbly cells or foamy cells can be found in xanthomas and in lipid-laden macrophages associated with fat necrosis. These cytoplasmic vacuoles are small and refractile on air-dried smears examined without a coverslip. Fat stains will be positive and mucin stains negative. Our case showed typical findings of those of recorded chordoma cases and the

diagnosis was confirmed by a following open biopsy.

The preoperative diagnosis of chordoma by FNA biopsy can greatly enhance optimal treatment. In this case, the patient received 4,000 rads of neutron radiotherapy and has been well till now.

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## ═국문초록═

### 척색종의 세침천자 세포학적 소견

원자력병원 해부병리과, 정형외과*

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저자들은 세침천자 세포학적 검사로 진단된 천미골 척색종 1예를 경험하였기에 보고하는 바이다. 환자는 54세 여자로 5, 6개월간 계속된 둔부의 동통을 주소로 내원하였다. 입원당시 CT 소견상 $L_5$에서 $S_5$에 이르는 척추의 파괴 양상과 연부조직 침윤이 덤벨상으로 보였다. 세침천자 도말상 풍부하고 포말상의 세포질을 가진 세포들이 군집으로 혹은 단독으로 산재해 있었다. 핵은 작고 난원형이거나 둥글었으며, 핵막은 얇고 염색질은 섬세하였다. 핵소체는 작았으며 하나 혹은 두개가 관찰되었다. 세포의 경계는 뚜렷지 않았고, 종양세포의 세포질은 풍부하였으며 특징적인 포말상의 공포가 관찰되었다. 부분적으로 불규칙한 세포크기를 보이기도 하였으나 비정형도는 미약하였다. 세포블록의 면역조직학적 염색상 EMA, Vimentin은 국소적으로 약한 양성반응을 Cytokeratin, S—100는 미만성으로 강한 양성반응을 보였으며, CEA는 음성이었다. 개방 생검술에 의해 천미골의 척색종임이 확인되었다. 환자는 4,000 rad의 중성자 치료를 받고 현재까지 잘 지내고 있다.