

Sacrococcygeal Chondroid Chordoma: FNAC Diagnosis: A Case Report with Histological and Radiological Correlation

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Abstract: We report a case of 30 year old female patient who presented with complain of gradually progressive swelling at the lower back for 8-months duration. Fine needle aspiration cytology (FNAC) of swelling revealed characteristic physaliferous cells seen as in chordoma and, radiological and histological findings are favoured the diagnosis of chondroid chordoma. Chondroid chordoma is a controversial and confusing entity and it is diagnosis on FNAC is rarely described.

Keywords: chondroid chordoma, FNAC, chordoma, physaliferous cell, sacrococcygeal

1. Introduction

Chordoma is a rare malignant tumour arising from notochordal remnants. In human remnants of notochord are found within the vertebral bodies and intervertebral discs and rarely in presacral soft tissue. Chordomas accounts 1-4% of all primary bone tumours.[1] It occurs mostly in adults with a median age of 60 years whereas chondroid chordoma occurs in slightly younger age group patients (mean age 40 years).[2] We report a case of sacrococcygeal tumour, which was diagnosed as chondroid chordoma on FNAC and, confirmed with radiological and histological correlation.

2. Case Presentation

A 30 year old lady presented with history of swelling and pain in the left lower back of 8 months duration. On examination, there was firm palpable swelling of size 8×6 cm in the left sacrococcygeal region and, aspiration of mass revealed mucoid and jelly like material. Fine needle aspiration cytology from the swelling showed large cells with vacuolated cytoplasm and prominent nucleoli-physaliferous cells also called soap bubble cells along with some medium sized cells with vesicular cytoplasm and centrally placed round nuclei with inconspicuous nucleoli - chordoma cells entrapped within abundant chondromyxoid stroma [Figure 1a and 1b].

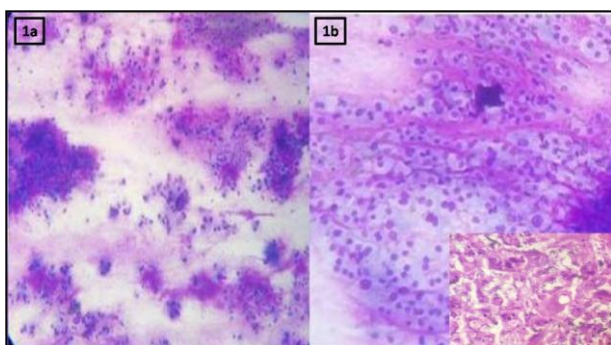


Figure1: FNAC and HPE images

MRI images showed abnormal signals involving sacrococcygeal vertebrae associated with presacral soft tissue component causing indentation on cervix and vagina and abnormal signals in the left gluteal and perianal regions [Figure 2a-c].

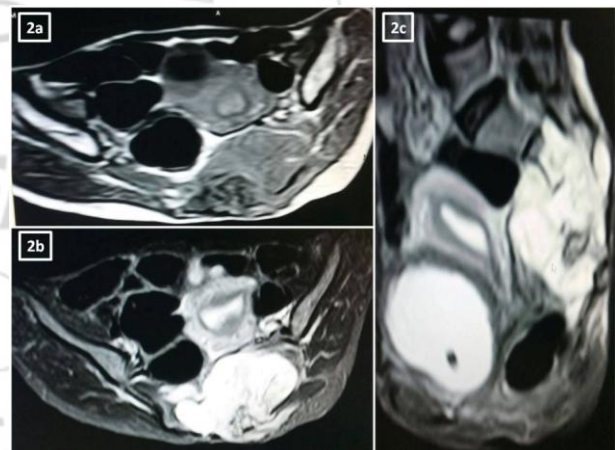


Figure 2: MRI axial T1W1, T2W1 and saggital T2W1

FNAC finding and radiological impression were suggestive of chondroid chordoma, and patient prepared for surgery. Near total excision of mass was done. Intraoperatively, the tumour was soft, lobulated, gelatinous and bluish-gray, with areas of haemorrhage. Histopathological examination of specimen showed classic arrangement of tumour cells in cords and lobules separated by a variable but extensive amount of mucoid intercellular tissue and fibrous septa. Tumour cells showed variable size of nuclei having prominent nucleoli with granular vacuolated cytoplasm (Physaliferous cells). Areas of cartilages were also present. Other cells are small with inconspicuous nuclei and no visible nucleoli. Mitotic figures were absent. Based on this, a diagnosis of chondroid chordoma was made [Figure 1b inset]. Post-operatively patient was referred to medical-oncology department, where local radiotherapy given. She showed a good clinical outcome with adjuvant radiotherapy.

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There was no evidence of recurrence of lesion at follow-up of 6 months.

3. Discussion

Chordoma described by Virchow in 1857, is a rare locally aggressive malignant tumour.[3] Its incidence is approximately 1 in a million population and 1-4% of all bone malignancies.[4],[5] In the axial skeleton, incidence of Chordoma is 50% in the sacral region, 35% in sphenoccipital region and 15% in vertebrae.[6],[7] In 1973, Heffelfinger et al., described chordomas at the base of the skull with prominent chondroid differentiation and it may occur at sphenoccipital and sacrococcygeal areas.[8]

Common presenting symptom is dull low backache which aggravates on movement, along with other pressure symptoms like neurological deficit, bladder and bowel involvement.[5] However clinical manifestations depend on location of tumour. In cranial region, they can cause cranial nerve palsies, hydrocephalus and torticollis.[6] The sacral lesions remain asymptomatic for a long time and presents with perineal pain, paraesthesia, constipation and bladder symptoms.

On gross examination, chordomas appear as soft, blue-gray, lobulated tumours. Histologically, there are 4 criteria used for diagnosis of chordoma: 1) a lobular arrangement of cells; 2) a tendency for the cells to grow in cords, irregular bands or pseudoacinar forms; 3) production of abundant intercellular mucinous matrix; and 4) the presence of large physaliferous cells.[9] Histomorphologically chordomas have been divided into three subtypes; conventional, chondroid and dedifferentiated. Conventional chordoma is the most common type which is slow growing malignant tumour. Chondroid chordoma is slower growing than conventional chordoma and shows foci of chondroid (cartilaginous) differentiation. It has a better prognosis than classic (non-chondroid) chordoma and, is indistinguishable from hyaline type chondrosarcoma.[8]

Dedifferentiated chordomas are rapidly growing tumours and are biphasic, with areas of high-grade sarcomatous changes, alongside conventional or chondroid chordoma. They represent 1-8% of all chordomas. Mostly these tumors arise from sarcomatous transformation of chordomas, and also occur after post-operative radiotherapy for conventional chordoma. Their prognosis is very poor with most patients succumbing due to tumour related complications within 1 year.[7]

The cartilaginous foci of chondroid chordomas resemble those of chondroma or low grade chondrosarcoma. Immunohistochemistry stain demonstrates that tumour cells of chondroid chordoma are reactive to epithelial markers like epithelial marker antigen (EMA) and cytokeratin (CK). Chondroma and chondrosarcomas are negative for CK and EMA and are positive for vimentin.[10] Therefore diffuse expression of CK and EMA in a tumour is diagnostic of chondroid chordoma in case of diagnostic dilemma.

Tumours detected and diagnosed early have a favourable prognosis if treated with a complete or en bloc excision. Unfortunately, many chordomas are large in size when initially evaluated and because of their locally aggressive behaviour have already infiltrated adjacent structures. Treatment options include low-dose or high-voltage radiation therapy, combined radiation and surgery and surgical excision alone. In some cases, debulking procedures are the only option. Use of photon radiation therapy may be beneficial especially for sphenoccipital lesions.[11] The 5 year and 10 year survival rates of conventional chordomas are approximately 50% and 25-30% respectively. Conversely, chondroid chordoma has 5 years 10 years survival rate of approximately 80% and 50% respectively.[12] Recurrent disease can be treated aggressively with surgery and adjuvant radiation therapy.[13] Dedifferentiated chordoma, metastasize early, while other two chordomas can metastasize latter in the course of disease, to skin, bone, lungs and lymph nodes. Vertebral body chordomas have a higher incidence of metastasis than the lesions at clivus or sacrum.[14] Therefore differentiation from other variants of chordomas and chondrosarcoma is important. High index of suspicion is required for early diagnosis and help to achieve adequate surgical margin for good outcome.

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