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CYTOLOAGICAL DIAGNOSIS OF CHORDOMA: A RARE CASE

Pathology

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ABSTRACT

Chordoma is a rare slowly growing malignant tumor arising from notochord remnants . It is diagnosed in just 1 in 1 million people per year with a male dominance M:F =2:1 and accounts for 1-4 percent of all bone tumors and 20 percent of primary spinal tumors.They are the most common tumor of sacrum and cervical spine, commonly seen in 5th and 6th decades but can occur in all ages .Chordoma has a relatively indolent ,protracted course ,often with multiple recurrences . Metastasis occurs in a minority of cases. We report a case of 51 years old male presenting with large gluteal swelling diagnosed as chordoma on FNAC and later confirmed on histopathology .

KEYWORDS:

Chordoma , Fine needle aspiration cytology ,Histopathology

INTRODUCTION

Rudolf Virchow originally described chordomas in 1857, under the name of "ecchordosis physaliphora "which refers to small ,well circumscribed ,gelatanious mass adherent to brainstem .The tumors were later named by Ribbert in 1894 ,he correctly surmised the notochordal origin of chordomas .They account for 1-4%[1,2] of all primary bone tumors . 50% of tumors arise in sacrococcygeal area, 35% in sphenooccipital and remainder along the cervicotho racolumber spine[3]. Patient's can have symptoms for months to years before diagnosis. Low back pain is the most common presenting symptom but when the tumors enlarge ,they can invade pelvic organs or nerve roots, causing organ and lower limb dysfunction[4]. Microscopically, three variants have been described, conventional (classic), dedifferentiated and chondroid.Conventional chordomas are the most common[5]. Here we present a case of chordoma diagnosed on FNAC and later confirmed on histopathology.

Case Report

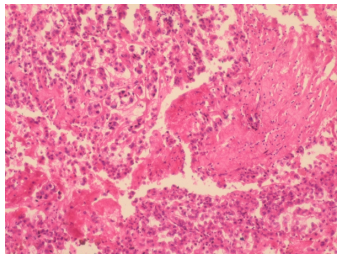
A 51 years old male presented in the orthopaedics department of GMC Jammu with a large swelling in the right gluteal region extending upto the left. A provisional diagnosis of soft tissue sarcoma was made and patient was advised to undergo FNAC in the cytology section of the deptt. of Pathology. On examination a large swelling (present since 1 year) measuring 10x7cm on right gluteal region extending on to the left was found. Swelling was soft to firm , slightly tender. FNAC was done yielded blood mixed aspirate, pap and giemsa stained slides were prepared. Smears were highly cellular, comprising of medium sized cells, some having eccentric nuclei with abundant basophilic cytoplasm entangled in a meshwork of purple chondromyxoid material . Large cells with abundant bubbly cytoplasm were also appreciated . Some cells depicted mild to moderate pleomorphism with prominent nucleoli.On the basis of above features, cytodiagnosis of Chordoma was extended.Excision with wide resection of the tumor was done and tissue was sent to histopathology section .Grossly the tissue was soft in consistency with areas of haemorrhage and necrosis .Microscopically, it showed tumor arranged in cords ,nests and lobules with large areas of necrosis. Individual cells, some large show vesicular nucleus and cytoplasmic vacuolation which at places show nuclear indentation and even a few signet ring like forms.These findings confirmed the cytological diagnosis of chordoma .

DISCUSSION

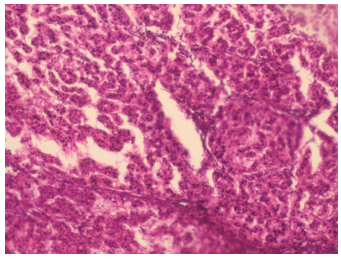
Chordomas are rare neoplasms composed of physaliphorous cells,occurring in the sites occupied by the embryonic notochord. These sites include the whole length of the spine from the skull base to the sacrum ,most commonly at one end or the other. Our patient presented with a huge swelling in the gluteal region, whose provisional diagnosis was high grade sarcoma ,but it displayed typical features of chordoma both on FNAC and histopathology. Crapanzano[6] also reported that chordomas have cytomorphological features which allow an accurate diagnosis to be rendered on FNAC. Radiographic features (CT scan) of the lesion showed a cystic and expansile mass in the sacral region.

Fine needle aspiration cytology is a safe , simple and quick method for early diagnosis of chordoma in presence of characteristic cytomorphological features (physaliphorous cells) and needs histopathological correlation which was done in our case to differentiate it from mimickers (Myxoid Chondrosarcoma, myx opapillary ependymoma and metastatic adenocarcinoma). The standard and the only curative treatment for chordoma is excision with a safe margin as recurrences are common[7]. Fuchs[8] reported 31 cases with wide resection ,only one of whom relapsed.Our patient was fortunate to receive an early and accurate diagnosis on FNAC which was later confirmed on histopathology.

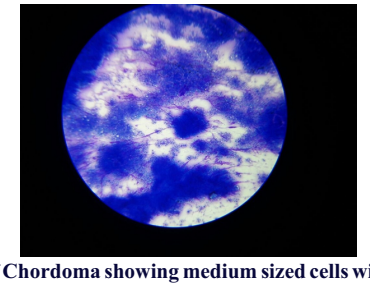
PHOTOS



HPE of Chordoma showing cells arranged in lobules and with areas of necrosis.



HPE of chordoma showing cells arranged in nests and chords with prominent cytoplasmic vacuolation. H&E (40X)



Cytology of Chordoma showing medium sized cells with abundant cytoplasm entangled in meshwork of myxoid material.

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