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Chordoma – FNAC Diagnosis: A Case Report and Review of Literature

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Abstract:

Chordoma is a rare slow growing malignant tumor which arises from primitive notochord remnants. A fifty year female patient attended outpatient complaining of swelling in the low back, gradually increasing over a six years period, attained present size of 3×4 cm. Fine needle aspiration cytology revealed characteristic physaliphorous cells seen in chordoma. Histopathology confirmed the diagnosis of chordoma.

Key words: Chordoma, Neoplasms, Nervous System Neoplasms, Notochord, Cytodiagnosis, Humans.

Introduction

Chordoma is a rare locally aggressive malignant tumor, described by Virchow in 1857 [1]. Incidence is approximately one in one million of population [2], and 1% to 4% of all bone malignancies [3]. It is commonly seen in 5th and 6th decades of life with male to female ratio of 3:1 [4,5]. Chordoma usually occurs in the axial skeleton [6] with an incidence of 50% in the sacral region, 35% in spheno-occipital region and 15% in vertebrae [5]. Common presenting symptom is dull pain in the low back which aggravates on sitting, along with other pressure symptoms like neurological, bladder and bowel symptoms [2].

Case Report

A 50 year old woman was admitted with the complaints of swelling and pain in the left low back for six years. On examination firm palpable swelling of size 3×4 cm in the left sacrococcygeal region was noticed. X-ray pelvis showed an ill-defined lytic lesion in the sacrococcygeal region with bone destruction and irregular amorphous calcifications anterior to the sacrum [Fig.1]. Ultrasound showed sacrococcygeal mass of size 7.6x6 cm. CT scan showed osteolysis of sacrum with soft tissue mass in the presacral region [Fig.2]. Routine investigations were within normal limits. Fine needle aspiration cytology from the swelling in left sacrococcygeal region showed large cells with vacuolated cytoplasm and prominent nucleoli-physaliphorous cells also called soap bubble cells. Some were medium sized cells with vesicular cytoplasm and prominent nucleoli-chordoma cells [Fig.3,4]. Background showed

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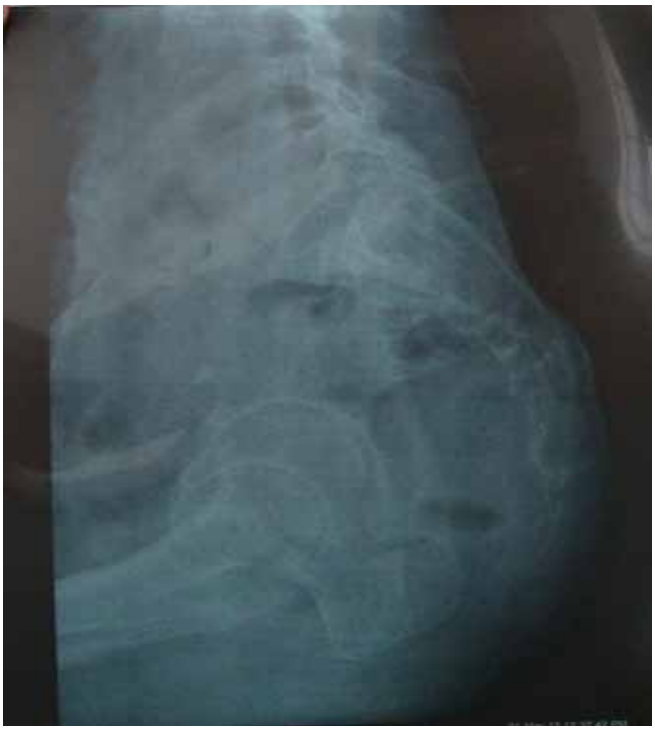

Fig.1: Plain X- ray pelvis show an ill-defined lytic lesion in the sacrococcygeal region with bone destruction and irregular amorphous calcifications anterior to the sacrum.

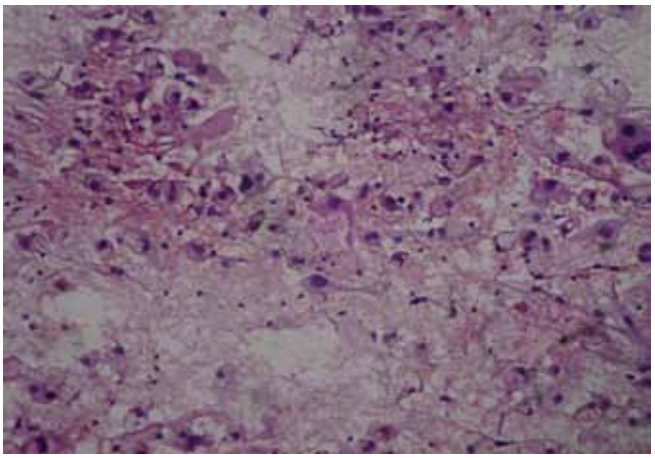

Fig.3: FNAC show physaliphorous cells and some medium sized cells with vesicular cytoplasm and prominent nucleoli in the fibromyxoid background (H&Ex100).

Fig.2: CT scan show osteolysis of sacrum with soft tissue mass in the presacral region.

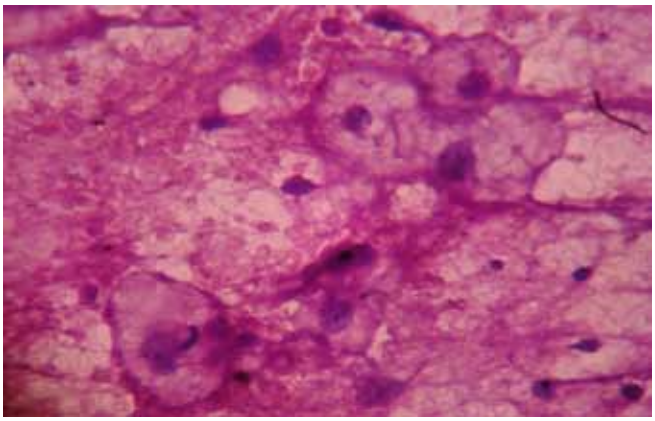

Fig.4: FNAC shows large cells with babbly cytoplasm (small arrow) and prominent nucleoli-physaliphorous cells (H&E x400).

abundant fibromyxoid material. Histopathology confirmed the diagnosis of chordoma.

Discussion

Chordoma is a rare malignant tumor, arises from notochord remnants. Duration of symptoms usually

ranging from six months to eight years [2], in the present case it took six years. Low back pain is the commonest symptom though it is very notoriously nonspecific [2]. In some of the studies, extremity weakness and sacrogluteal mass are common complaints of sacrospinal chordoma [1]. In the present case, the main complaints were low back pain and swelling in the sacrococcygeal region. Presacral tumors are often palpated by rectal examination. On plain X-ray they appear as solitary midline swelling with osteolytic lesion in the sacrococcygeal bone [7] as in the present case. CT scan shows soft tissue swelling and destruction of sacrococcygeal bone [6]. In 40% of the cases, CT scan shows focal calcifications as seen in present case [1].

Fine needle aspiration cytology shows large cells with vacuolated or bubbly cytoplasm and prominent nucleoli-physaliphorous cells also called soap bubble cells. Some were medium sized cells with vesicular cytoplasm and prominent nucleoli in the fibromyxoid background [7]. The characteristic physaliphorous cells in the fibromyxoid background were also seen in the present case. Some of the cytological studies show pleomorphic physaliphorous cells, nuclear inclusions, binucleation and multinucleation [8]. Biological behavior of chordoma is highly variable, commonly low grade neoplasm with bland nuclear features. Sacral chordoma extends into presacral, subperiosteal and also into sacral canal. Sacrum and coccyx are destroyed due to tumor invasion with rectum, bladder and uterine displacement. Wide surgical excision is the treatment of choice. Large tumors require combined abdomino-sacral approach to remove entire tumor. Histopathologically clusters of pleomorphic cells, including physaliphorous cells arranged in cords, sheets and nests amidst myxoid matrix confirms the diagnosis of chordoma [7,9]. Immunohistochemical markers are positive for cytokeratin, EMA and S100 [9].

Local recurrences are observed in 50% of cases of sacrococcygeal chordomas [3,5]. Distant metastasis occurs in 10% of cases to lungs, liver, lymph node, skin and muscles [5]. The prognosis depends on completeness of resection, age, gender and postoperative irradiation [1]. Post-operative irradiation is required in case of high sacral tumors. Chemotherapy has doubtful role [5]. Radiotherapy with complete excision achieves the best results with a disease-free survival of more than 5 years [1]. Ten years survival rate is 63% [2]. Chordoma should be differentiated from chondrosarcoma, mucinous metastatic adenocarcinoma and myxopapillary ependymoma [4].

Conclusion

Characteristic physaliphorous cells in the FNAC are the best tool to diagnose chordoma preoperatively. Early diagnosis may lead to preservation of bladder, bowel, motor function, sexual function and also prevents distant metastasis.

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