Cytodiagnosis of Sacral Chordoma

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ABSTRACT

We report the cytological findings of a sacro-coccygeal chordoma in a 53 year male diagnosed preoperatively by guided fine needle aspiration cytology. The smears shows characteristic Physalliphorous cells in a metachromatic background of myxoid material entrapping cords of cuboidal cells. Differential diagnosis in cytology include conventional and myxoid chondrosarcoma, myxoid liposarcoma, myxoid malignant fibrous histiocytoma(MFH), metastatic mucinous carcinoma and myxo-papillary ependymoma. The distinguishing features between these neoplasms are discussed. Preoperative diagnosis of chordoma permits optimum planned surgery.

Keywords: *chordoma; myxoid; sacral.*

INTRODUCTION

Chordoma is an unusual encapsulated malignant tumor of fetal notochord origin that commonly affects the axial skeleton involving the sacrum, base of skull and cervico-thoracic spine.[1] The cytomorphological features of the tumor are varied but characteristic and diagnostic. An important prerequisite for optimal management of these tumors is a correct pre-operative diagnosis. We report an unusual case of a sacral mass where the fine needle aspiration cytology (FNAC) along with radiological findings clinched the diagnosis.

CASE REPORT

A 53 year old male presented with a swelling the sacro-coccygeal region which was accompanied with pain in lower spinal region, heaviness in anorectal region and constipation. Per-rectal examination detected a soft cystic mass in presacral region. Computed tomography scan revealed osteolysis of sacrum with large soft tissue mass in the pre-sacral region pushing the posterior rectal wall forward.(Fig,1) FNAC of the mass was performed. The smears were moderately cellular and revealed cords of cuboidal cells in a myxoid background. The cuboidal cells had eosinophilic vacuolated cytoplasm with peripheral nuclei. Physalliphorous cells with moderate amount of vacuolated cytoplasm and scalloped centrally placed nuclei were also observed. The myxoid material stained with Leishman's stain showed metachromasia (Fig2). The tumor was completely excised under general anaesthesia and sent for histopathology examination. Grossly, the tumor was soft and gelatinous with areas of hemorrhage. The histology revealed lobulated tumor composed of an admixture of large multivacuolated (Physalliphorous) cells and cuboidal cells, arranged in cords, sheets, and nests in a myxoid matrix. The histomorphological features confirmed the diagnosis of chordoma. (Fig 3). The tumour was immunoreactive for epithelial markers like cytokeratin, epithelial membrane antigen and S-100.

DISCUSSION

Chordomas are slowly growing malignant tumors that have a tendency to recur.[1,2] Chordomas are divided into

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three subtypes, based on microscopic morphology: 1) conventional, 2) chondroid, and 3) dedifferentiated. Conventional chordomas are slowly growing tumors and characteristically show large physaliphorouscells with vacuolated cytoplasm arranged in nests, cords, or sheets within a myxoidstroma. Chondroidchordoma is a variant showing foci of chondroid differentiation. Dedifferentiated chordoma is a biphasic tumor showing areas of high grade sarcoma and conventional or chondroidchordoma. Patients with chondroidchordomas have a longer survival than those with conventional chordoma whereas patients with dedifferentiated chordoma have a very poor prognosis. Larger tumor size, inadequate surgical margins, microscopic tumor necrosis, Ki-67 > 5%, and local recurrence are also associated with poor prognosis.[3]

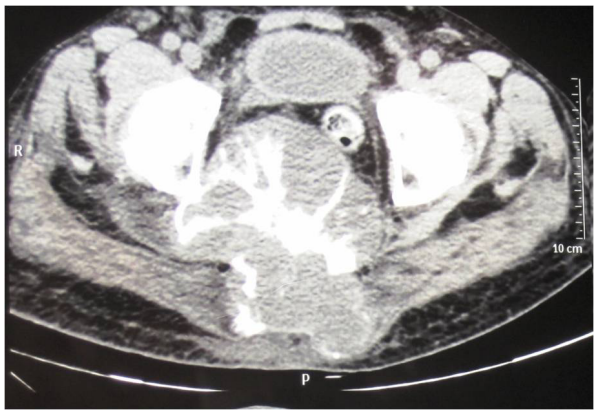

Figure 1. CT scan: Osteolysis of sacrum with large soft tissue mass in the pre-sacral region.

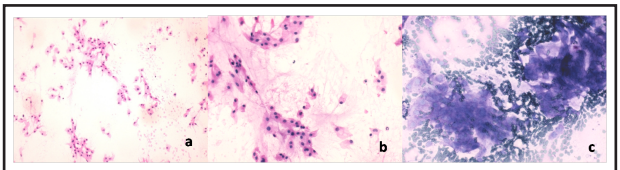

Figure 2. (a) Cuboidal cells and Physalliphorous cells over a myxoid background (H & E x 40) (b) cuboidal cells with eosinophilic vacuolated cytoplasm ,peripheral nuclei and physalliphorous cells with moderate amount of vacuolated cytoplasm and scalloped centrally placed nuclei (H & E x100) (c) metachromasia of the myxoid material (Leishman stain x 40).

The cytologicaspirates are generally cellular. Physaliphorous cells are pathogonomic which have cytoplasmic vacuolation, nuclear indentation and signet ring–like morphology. Cytoplasmic borders of physaliphorous cells in aspirates have been reported to be both distinct, or well demarcated and indistinct, or syncytial-like. Rarely these cells are absent in chordomas which results in misdiagnosis.[4] Walaas et al have reported the presence of pleomorphism with nuclear hyperchromasia, nucleolar prominence including binucleation or multinucleation of the physaliphorous cells.[5] Cristallini et al reported the presence of nuclear inclusions in chordoma aspirates, which is believed to be a feature of dedifferentiation.[6] The myxoid background is another important, though nonspecific, diagnostic feature of chordoma.

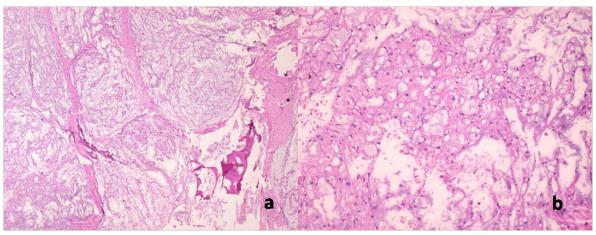

Figure 3. (a) Lobulated tumour with a myxoid background (H & E x40) (b) Admixture of large multivacuolated (Physalliphorous) cells and cuboidal cells, arranged in cords, sheets, and nests in a myxoid matrix (H & E x 100).

The other cell which is commonly seen in chordoma is a non vacuolated intermediate sized, round to polygonal epithelial cell. These cells were observed both singly and in clusters and often were intermixed with physaliphorous cells.

Other cell types described in conventional chordoma include mononucleated, nonvacuolatedspindly cells usually arranged within the myxoidstroma. Mitoses are rare.

The cytologic differential diagnosis of chordoma includes chondrosarcoma (conventional/ myxoidchondrosarcoma), metastatic carcinoma, myxoidliposarcoma, myxoid malignant fibrous histiocytoma (MFH), and myxopapillary Ependymoma.[3,7-9]

Conventional chondrosarcoma may involve the sacrum but unlikechordoma, the aspirate material shows malignant cartilage cells, sometimes within lacunae, which usually lack the prominent cytoplasmic vacuoles noted in physaliphorous cells. The background cartilaginous matrix of chondrosarcoma is mucicarmine negative whereas the matrix of chordoma is mucicarmine positive. Cytologic preparations from chondrosarcomas may be very difficult to distinguish from chordomas. The cells of chondrosarcoma are positive for S-100 protein and negative for epithelial markers including EMA and cytokeratins which often helps to distinguish the two as chordoma is positive for EMA and cytokeratins and often shows reactivity for S-100 protein.[3,8,9]

Myxoidchondrosarcoma is a tumor that usually

arises in the soft tissues of the lower extremities as opposed to the spinal axis and therefore is less of a consideration in the differential diagnosis of chordoma. However, they can arise in axial bone. FNAC findings from myxoidchondrosarcoma may show features nearly identical to chordoma. Aspirates contain uniform, round to oval cells with finely granular chromatin, conspicuous nucleoli, and a moderate amount of delicate, finely vacuolated cytoplasm in a background of abundant, brightly metachromatic myxoidstroma. Immunoreactivity of myxoidchondrosarcoma is similar to conventional chondrosarcoma, a feature useful in distinguishing it from chordoma.[3]

Metastatic carcinoma also confused with chordomas. However, metastatic adenocarcinoma lacks true physaliphorous cells and may show glands, papillary clusters, or columnar cells, features not observed in chordoma. Signet ring cells may be observed in both metastatic adenocarcinoma and chordoma. The nuclear features of chordoma are usually more benign than those of adenocarcinoma. Epithelial markers will not be helpful because both chordomas and carcinomas stain positively for EMA and cytokeratins. Mucicarmine stain is also not helpful because positive staining may be observed in both chordoma and metastatic carcinoma.[1,3]

Myxoid MFH most commonly involves the metaphyseal region of the distal femur, proximal tibia, and proximal humerus in contrast with the axial skeleton location of chordoma. Myxoid MFH aspirates are cellular and consist of pleomorphic, spindle shaped cells with pleomorphic forms in the higher grade tumors. The cells have a moderate amount of cytoplasm, which may be foamy or vacuolated, but physaliphorous cells are not observed. Immunohistochemically, myxoid MFH is distinct from chordoma, showing reactivity for vimentin, but not for cytokeratins, EMA, or S-100 protein.[3,9]

Myxoidliposarcomas usually affect the soft tissue of the lower extremities and only rarely arise in the bone. The aspirates consist of cellular smears with abundant, granular myxoid background that is metachromatic in modified Romanowsky stains and is alcian blue positive, hyaluronidase sensitive, and mucicarmine negative. In contrast, the background matrix of chordoma is alcian blue positive, hyaluronidase resistant, and mucicarmine positive. The cells of myxoidliposarcoma are bland, monomorphic, spindle or stellate, and contain mildly hyperchromatic nuclei with evenly distributed chromatin. Plexiform vessels are typically observed, a feature not present in chordoma. Immunohistochemically, the tumor cells are negative for epithelial markers and positive for S-100 protein, similar to chondrosarcoma.[3,8]

Myxopapillaryependymoma usually arises in the filumterminale and rarely in the presacral or postsacral soft tissue. Cytological features are papillary vascular cores lined by columnar cells. Physaliphorous cells are absent in myxopapillaryependymomas. Immunohistochemically, ependymal cells show reactivity for glial fibrillary acidic protein, a feature not observed in chordoma.[3]

CONCLUSIONS

The primary treatment of sacral chordoma includes complete excision followed by radiation therapy. FNAC is a safe, simple and quick method for early pre-operative diagnosis and to plan the surgical treatment.

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