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NOTOCHORDAL TUMOUR- CHORDOMA: A RARE CASE REPORT

Pathology

Dr Junu Devi Associate Professor, Department of Pathology, Gauhati Medical College and Hospital.

Dr Upasana Kalita Post Graduate Trainee, Department Of Pathology, Gauhati Medical College And Hospital.

Dr Eveeta Baruah* Post Graduate Trainee, Department of Pathology Gauhati Medical College And Hospital.
*Corresponding Author

KEYWORDS

INTRODUCTION

Chordoma is a rare primary malignant bone tumor of notochordal origin. It accounts for approximately 5% of primary bone tumors[2]. Location is central to the diagnosis of Chordoma. Most Chordomas occur at either end of the primitive notochord, approximately 40% in the clivus and 45% in the sacrum; the rest occur from notochord vestiges along the spinal column, 10% along cervical, 2% along thoracic, and 2% along lumbar portions of the spine[1].

CASE REPORT

We present a case of a 25 years old female with a vague swelling at the back of her neck since 1 year. She presented with progressively worsening headache, neck pain, diplopia and weakness of lower limbs. Radiological studies revealed a large heterogenous mass in C4-C5 region. Ultrasound guided FNAC show features suggestive of Chordoma. The patient was treated with surgical excision of tumour and histopathological examination was performed. Histopathology confirmed the diagnosis of Chordoma. Postoperatively, the patient experienced an improvement in neurological symptoms.

Cytological Findings

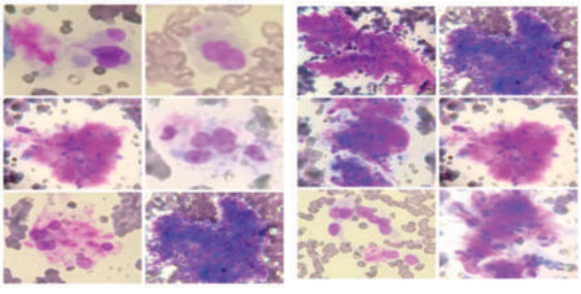

Fig 1: **(a)** **(b)**

FNAC- Cohesive clusters and singly dispersed round to cuboidal cells mixed with myxoid ground substance. The cytoplasm contains conspicuous cytoplasmic vacuoles. Cells with an abundance of vacuolated, bubbly cytoplasm and well differentiated cell borders are also noted (physaliferous cells).

Histopathological Examination

Microscopic Examination:

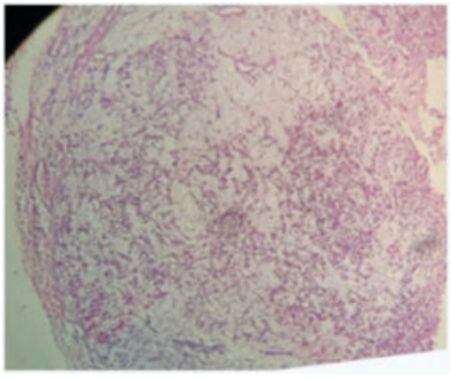

Fig 2: (a)

Scanner view: fig- Lobulated structure showing cords, nests and sheets of cells in a myxoid matrix

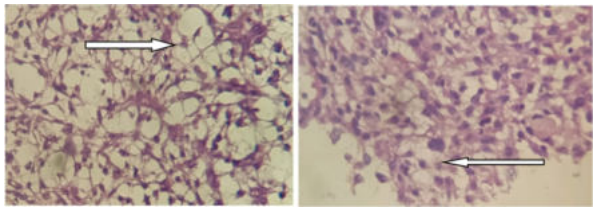

Fig 2: **(b)** **(c)**

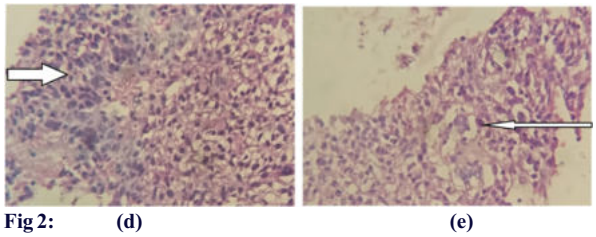

Fig 2: **(d)** **(e)**

Low power view: fig 2: b, c- Sheets of large vacuolated cells (physaliferous) with centrally placed hyperchromatic nuclei.

fig 2: d, e- Sheets of malignant cells with hyperchromatic nuclei and vacuolated cytoplasm. High grade area noted (fig d).

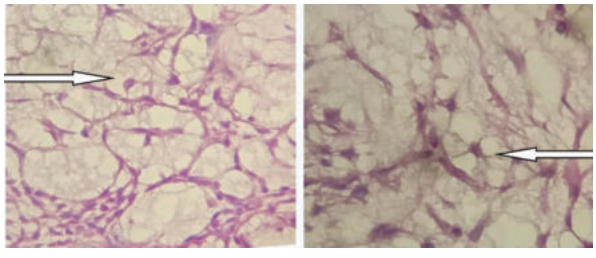

Fig 2: **(f)** **(g)**

High Power View: Physaliferous cells with abundant vacuolated bubbly cytoplasm

DISCUSSION

The study of chordoma begins in 1846 when Rudolf Virchow first observed its occurrence on a dorsum sellae; he coined the term "chordomata" 11 years later. The chordoma's origin was greatly disputed by members of the scientific community. Eventually, Muller's notochord hypothesis was accepted 36 years after its proposal. Chordomas were considered benign and slow growing until the early 1900s, when reported autopsy cases drew attention to their possible malignant nature. Between 1864 and 1919, the first-ever symptomatic descriptions of various forms of chordoma were reported, with the subsequent characterization of chordoma histology and the establishment of classification criteria shortly thereafter[7].

Chordoma is the only malignant neoplasm derived from notochordal elements, the embryonic notochord is a midline structure prescribing

the sites of Chordomas. This rare, midline lesion usually occurs at one of the two ends of the axial skeleton[3]. The annual incidence of Chordoma is 0.8 cases per 1,00,000 population, with 32- 42% arising in cranial sites, mainly in the base of the skull[5]. Most Chordomas occur in adults; only 5% of tumors develop in patients under 20 years of age. Males are affected twice as often as female.

The clinical symptoms of Chordoma depend on the anatomic location. Patients may have symptoms for months to years. Pain is almost always present[2]. Patients may present with headache, cranial nerve palsy, or brain stem compression, depending on the anatomical structures compromised[5].

Radiologically, heterogenous mass is seen often destructing the surrounding soft tissue areas and also calcifications in chordoma. MRI and CT scan have complementary roles in tumor evaluation. CT evaluation is needed to assess the degree of bone involvement and to detect patterns of calcification within the lesion. MRI provides excellent anatomical delineation of adjacent structures and is able to characterize the signal of the lesion usually allowing for a confident preoperative diagnosis[8].

On FNAC, cohesive clusters and cords of round to cuboidal cells are found in a rich myxoid matrix. The cytoplasm contains conspicuous cytoplasmic vacuoles. Some cells have an abundance of vacuolated cytoplasm and are called physaliferous (from the Greek physallis, meaning bubble). FNAC provides an early pre-operative diagnosis of chordoma because of the distinct cytological features.

On HPE, lobulated growth pattern is seen. The tumour lobules are separated by fibrous bands. An abundant myxoid matrix is always present. The tumour cells are arranged in sheets or cords or seen as single elements floating in the background matrix. Typical tumour cells are large with abundant (multi)vacuolated cytoplasm and rounded or ovoid nuclei (physaliferous cells). The physaliferous cells are admixed with smaller, epithelioid-like cells and spindly cells. Cellular atypia is usually not prominent although multinucleated tumour giant cells and cells with hyperchromasia and prominent nucleoli may be present[6].

Chordomas express cytokeratin, EMA, and brachyury[1]. Brachyury is the most helpful marker because of its high specificity for chordoma[2]. The differential diagnosis for Chordoma is Chondrosarcoma. Both Chordomas and Chondrosarcomas express S-100. Chordomas express cytokeratin, EMA, and brachyury, whereas Chondrosarcomas lack these antigens[1].

Chordomas of the skull base are treated with surgery; however, proton beam radiation also has been shown to be an effective mode of treatment at this site. The recent identification of abnormal epidermal growth factor receptor (EGFR) signaling in Chordomas has opened new avenues for the treatment of this tumor with small molecule inhibitor.

The most important prognostic factor is complete surgical resection, which can be achieved only rarely in cranial sites . The 3-, 5-, and 10-year overall survival rates are 80.9%, 73.5%, and 58.7%, respectively[5].

CONCLUSION:

Chordomas are rare tumors of a notochordal origin that often have an insidious onset. As they progress, they cause local destruction, and hence neurological deficits. Early diagnosis is essential, as aggressive surgical resection followed by adjuvant therapy improves long-term outcomes[4].

Chordoma is a rare tumour with male preponderance twice that of female. Here we presented a case of a young female who was diagnosed with Chordoma on FNAC. Characteristic physaliferous cells in myxoid rich matrix in the FNAC is diagnostic for chordoma preoperatively. Thus early diagnosis of chordoma is important as it leads to preservation of neurological function and also prevents distant metastasis.

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