ORIGINAL RESEARCH PAPER

Volume - 10 | Issue - 04 | April - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

LANGERHANS CELL HISTIOCYTOSIS OF PTERIONAL REGION IN A CHILD

Neurosurgery

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ABSTRACT

We report a case of 3-year- old boy who presented with swelling in the left pterional region and on and off fever. CT and MRI suggested an osteolytic lesion with soft tissue mass. Lesion was excised and confirmed to be Langerhans cell histiocytosis.

KEYWORDS

Eosinophilic Granuloma, Langerhans Cell, Pterion

INTRODUCTION

LCH is characterized by proliferation of Langerhans cells.[1] Gene mutation or infection leads to uncontrolled and irregular proliferation of histiocytes and lymphocytes.[2] It commonly affects children below the age of 4 years, however it can occur at any age. Bone is most commonly involved organ. Histochemically , Langerhans cells are positive for the presence of CD 1a and S-100 surface markers with the presence of Birbeck granules on electron microscope[2] Recently we came across a case of pterional involvement in a child. This is a very unusual location and hence being reported.

Case report

A 3-year- old male child was brought with complaints of gradually increasing swelling on the left side of head since 8 months .He also had history of fever on and off for the same duration. There was no history of trauma. On examination: Child was fully conscious and without any neurological deficits. A 4 x 4 cm sized nontender, non-pulsatile swelling was present over the left frontotemporal region (fig 1a). Cough impulse was absent and transillumination was negative. It was soft in consistency. Hemoglobin was 10.4 g%, WBC 13.7 K , differential counts NE 52%, lymph 37.4%, MO 3-8 and eosinophils 5.8 %. CT revealed soft tissue shadow with erosion of the bone in pterional area (fig 1b). MRI revealed slightly mixed intensity well delineated lesion on T1W1 and T2W1 which enhanced inhomogeneously on contrast.

Fig 1

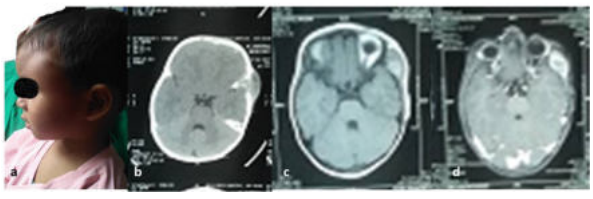

Figure 1 a) Clinical photograph showing frontotemporal swelling, b) Computed tomography showing erosion of bone with bevelled edge and soft tissue mass, c) MRI T1W1 showing mixed intensity lesion in the pterional region, d) Contrast MRI showing uniform enhancement except small central areas

At surgery, the swelling contained yellowish coloured toothpaste like substance destroying the bone and reaching up to dura (fig 2a). Bone was nibbled all-around and lesion was separated from the dura which was intact. Histopathology showed trabeculae with sheets of histiocytic cells . The cells were oval, grooved, folded, indented nuclei with fine chromatin. Mitosis was low and cytoplasm was eosinophilic (fig 2b). Numerous giant cells and eosinophils with formation of eosinophilic abscess was seen at places. Immunohistochemistry revealed histiocytic cells positive for CD 1a, S100, CD 68 (fig 2 c and d)consistent with Langerhans cell histiocytosis (Eosinophilic granuloma-old terminology).

Postoperative scan revealed no residual mass. Post-operative PET scan did not reveal any area of metabolic activity. Patient has been asymptomatic and there has not been any clinical evidence of recurrence.

Fig 2.

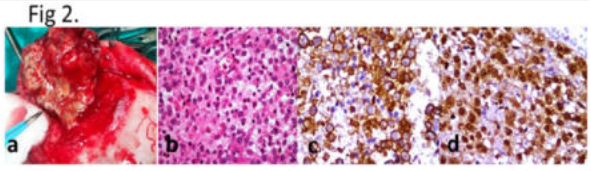

Figure 2 a) Operative photograph showing the nodular mass , b) histology showing oval Langerhans cell ,c) immunohistochemistry positive for CD 1A staining , d) nuclear and cytoplasmic positivity for S-100.

DISCUSSION

LCH previously known as Histiocytosis X is characterized by clonal proliferation of dendritic cells in various organs and tissues. There may be a single lesion or multicentric and multiorgan system involvement. It commonly affects bone especially skull, mandible, ribs and long bones and may spread to adjoining soft tissues.[3] In the skull , the lesion commonly affects temporal, parietal or frontal bone and involvement of pterion has not been reported earlier to the best of our knowledge. Differential diagnosis include meningioma, eosinophilic granuloma, plasma cell granuloma, tuberculosis, osteomyelitis. Because of its being rare and variable presentation, the diagnosis of LCH may get overlooked.

Eosinophilic granuloma is the benign of the three clinical variants of LCH. Other subtypes being Hand-Schuller-Christian disease (multifocal) and Letterer-Siwe disease (a diffuse systemic). The lesion may be asymptomatic or discovered incidentally or it may present as painful mass. Systemic symptoms may include general fatigue, rarely fever with leucocytosis as seen in our patient. Rarely there may be eosinophilia[4]. At times they may present as extra dural hematoma.[5] Plain x-ray reveals punched out lytic area without sclerosis of the margins. CT scan demonstrates cortical erosion and soft tissue mass. MRI shows low signal on T1W1 and hyperintense or isointense on T2W1 and enhances on contrast.[6] Surgical excision suffices if there are no other lesions in the body. Radiotherapy may be used as adjuvant therapy .In our case as there was no evidence of lesion anywhere else in the body , the adjuvant therapy was not advised. However, chemotherapy is indicated in multifocal and multi-organ involvement. Spontaneous remission of the eosinophilic granuloma has been reported in the literature in skeletally immature patients.[7,8] Recurrences are treated by irradiation and multiorgan disease requires systemic chemotherapy to bone marrow transplantation.[9]

Exact incidence of Eosinophilic Granuloma is not known. Etiology may be autoimmune, inflammatory or uncontrolled replication of Langerhans cells.[10]

CONCLUSIONS

The occurrence of eosinophilic granuloma involving the pterional region is very rare. While evaluating and managing the lesion of this site, it is worthwhile to include eosinophilic granuloma in the differentials.

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