Langerhans Cell Histiocytosis In Postauricular Region

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ABSTRACT

We present a rare case report of eosinophilic granuloma in 10 year old female which was managed surgically. We also review the case literature, the clinical presentations briefly with surgical management.

Key words: Langerhans Cell Histiocytosis, Postauricular Region
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INTRODUCTION

Langerhans cell histiocytosis is the term currently recommended to refer to the spectrum of diseasescharacterized by proliferation of Langerhans cells which were previously referred as Histiocytosis X causing local or systemic effect(1–3).

Langerhans cell histiocytosis (LCH) is a rare disease of unknown cause. LCH, formerly known as histiocytosis X, is a disease entity composed of three rare proliferative disorders of bone marrow–derived antigen-presenting cells of the dendritic cell line, also known as Langerhans cells(1–5). LCH is composed of three distinct clinical syndromes that show indistinguishable histology. Characteristically, these lesions stain positively with histochemical stains, S-100 and CD1a. Eosinophilic granuloma is limited to bone in patients usually 5–15 years old. HandSchüller-Christian disease is characterized by multifocal bone lesions and extraskeletal involvement of the reticuloendothelial system (RES) and pituitary gland, usually seen in children 1–5 years old. In Letterer-Siwe disease, there is disseminated involvement of the RES with a fulminant clinical course in children less than 2 years old(1–3,5–7)

This is rare case report we report of eosinophilic granuloma in 10 year old female which was managed surgically.

CASE REPORT

A 10 year old female child presenting with history of recurrent swelling in the postauricular region for a duration of 5 months. She had a history of, the swelling burst opening on its own and thick material brownish in clour pouring out from the swelling which was managed conservatively. There was no history of surgery or incision and drainage. At the time of

presentation patient had persistent sinus with discharge in the right postauricular region (Fig: 1). There was a cortical bone defect on palpation. External auditory canal and tympanic membrane were normal on examination. Tuning fork tests were normal. Pure tone audiometry was also within normal limits. Contrast enhanced computerized tomography of the temporal bone was done which showed heterogenous soft tissue density involving the retrosigmoid region which was purely extradural. MRI brain also showed the lesion in the retrosigmoid region which was extradural with no intradural extension (Fig:2). Middle ear was normal in imaging. Patient was taken up for surgery under general anaesthesia. Postauricular incision with elliptical incision to include the sinus skin was given. Lesion was dissected and could be separated from the surrounding skin (Fig:3). Drilling of the mastoid bone around the lesion was done and the lesion was exposed completely. Its extradural location was confirmed, lesion was removed in toto with intact dura left behind without any CSF leak (Fig:4). Temporal bone defect was closed with temporalis fascia and soft tissue from the postauricular region that was harvested. Wound was closed in two layers.

Fig : 1 Persistent sinus with discharge in the right postauricular region

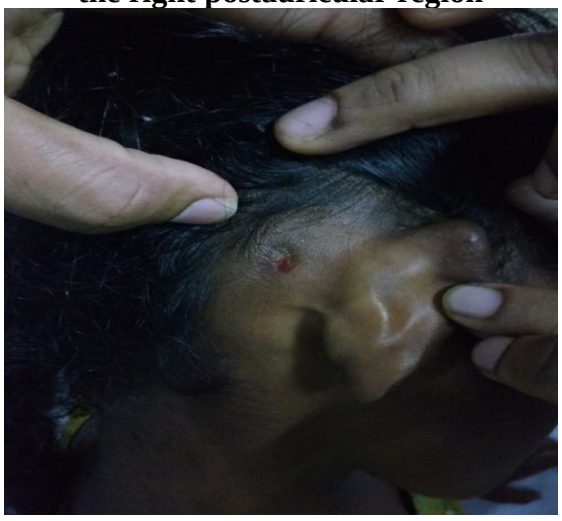


Fig:2 T2 weighted MRI showing a hyper dense lesion in the retroauricular region which is extradural

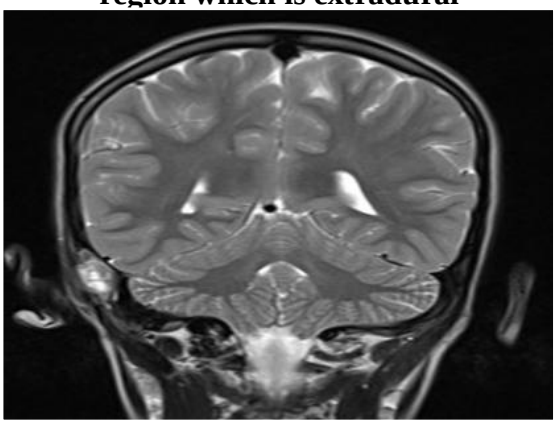


Fig 3 - Lesion visualised after elevation of subperiosteal flap

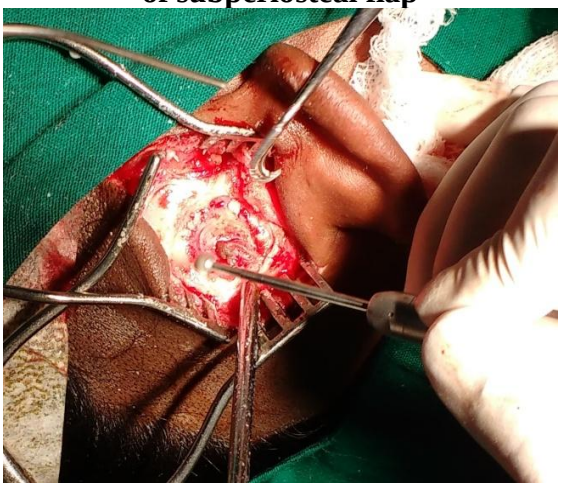
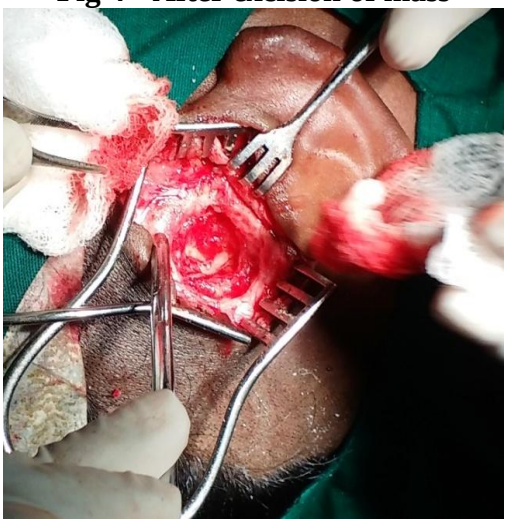


Fig 4 - After excision of mass



Histological picture

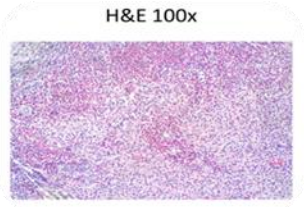


Fig:5 Section shows typical LC histiocytes with cleaved nuclei admixed with numerous eosinophils

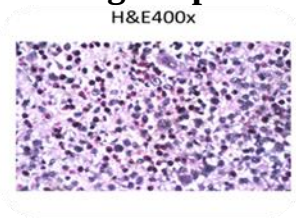


Fig: 6 Highlights the LC histiocytes in high power

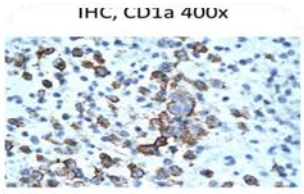


Fig: 7 Histiocytes are positive for CD1a

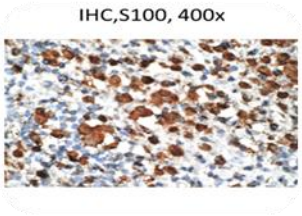


Fig:8 Histiocytes are positive for S-100

DISCUSSION

Langerhans' cell histiocytosis also known as histiocytosis X in the past is a group of disorders including(Eosinophilic granuloma, Hand-Schüller-Christian disease, Letterer-Siwe disease) that shows varing disease manifestations(2). Etiopathogenesis of the disease is not known. Most of the works done previously had shown it to have an autoimmune etiology (3).

Eosinophilic granuloma of bone, Hand-Schuller-Christian disease, and Letterer-Siwe disease are varied presentation of one disease which had been grouped by Lichtenstein into "histiocytosis X," as these condition had proliferation of histiocyte in common(5). In the clinical presentation, however, they seem to have difference to separate the three variety of disease. 1893, author Hand had described a case report of 3-year-old male child with polyuria, hepatosplenomegaly, cutaneous petechiae, exophthalmos and destructive skeletal lesions. Similarly at the same time Schuller reported a 5-year-old female child with "map-like" lytic defects in the femur, ilium and skull and Christian reported a 5-year-old female child with bony defects, exophthalmos and diabetes insipidus so all these three rare case report where grouped and named as Hand-Schuller-Christian disease. Eosinophilic granuloma of the bone first described by Lichtenstein and Jaffe after they reported two cases with solitary lytic bone lesions which when seen microscopically phagocytic cells with prominent collections of eosinophils were seen. Letterer-siwe disease is characterized by hepatosplenomegaly, a hemorrhagic diathesis, lymphadenopathy, anemia and skeletal defects.

Involvement of the temporal bone was seen in 15–61% of all cases of Langerhans' cell histiocytosis. Patients with suspected Langerhan's cell histiocytosis show undergo full body elevation to exclude multisystem involvement. Most common otologic symptom will be otorrhoea that is resistant to medical treatment. Other finding can be mastoid swelling, aural

polyp, eczema, sinus. Some cases erosion of the posterior canal wall and sagging in the external auditory canal is seen. In most cases as the finding are likely to mimick mastoiditis so diagnosing this type of diseases is little time consuming. Chronic otitis media or Cholesteatoma in children between1- 3 years of age one should always consider excluding Langerhans cell histocytosis as one of the differential diagnosis especially when ESR is elevated and when the diseases is not responding to medical management. Langerhans cell histiocytosis will be differential diagnosis especially when there is major radiological changes that are found out of proportion with significantly mild clinical findings (2)

Deafness in histiocytosis X can be both conductive and sensory neural hearing loss. When the disease involves middle ear (i.e) the ossicle chain, or when the external auditory canal get obstructed conductive hearing loss happens. Sensory neural deafness and vertigo happens when the bony labyrinth gets involved(2,3,5). Cranial nerve paralysis are uncommon, but if it involves it happens mostly with the seventh and eighth nerves. Cranial nerve involvement happen only when the skull base gets involved by disease. This rarely happens in langerhorn cell histiocytosis. So if cranial nerve palsy happens then other differential diagnosis like rhabdomyosarcoma or other malignancy should be roled out. Usually, imaging with high resolution temporal bone CT scan will show temporal bone destruction mostly involving the mastoid , squamous and middle ear(less commonly involved). Ossicles and the internal ear involvement is very rare(1–3). CECT will show an uniform contrast enhancement of lesion. On MRI scan, the lesion will appear with strong intensity in T2 and variable intensity in T1, often there can be surrounding edema or inflammation which will show marked enhancement after gadolinium(1–3,5). Differential diagnosis after radiological studies will be mastoiditis, rhabdomyosarcoma, and metastasis. But in mastoiditis there would be extensive bone destruction so it can be rolled out but other condition will have similar radiological features(2,3). Biopsy of the lesion will help to differentiate between different condition(2,3) .

Treatment of Langerhans cell histiocytosis involving the temporal bone includes mastoidectomy followed by chemo or radiotherapy, nowadays there is the trend toward local steroids injection (2–5). Treatment of disease with multisystem involvement includes chemotherapy, steroid injections, and immunotherapy. Fernande et al said that high frequency of involvement of the hypothalamic–hypophyseal axis leading to diabetes insipidus(2).

Eosinophilic granuloma is rarest form of langerhorn cell histiocytosis which involves only the bone without other systems involvement represents less than 1% of them. In 90% of the reported cases where children less than 10years.And male children where more commonly affected(M:F ratio 5:1).Eosinophilic granuloma affect commonly the flat and long bones presentation usually will be monostatic lesion. The Langerhans cell with Birbeck granules are characteristic of this condition(5). Eosinophils, lymphocytes, fibroblasts and foam cells can also be found but none is pathognomonic of the condition(2,5,6). The most reliable immunological marker is the OKT6. But the other common S- 100 protein is usually positive too. Clinical feature in eosinophilic granuloma will generally be a asymptomatic swelling without any pain or signs of inflammation. In most of the cases there will not be any history of aural symptoms. Swelling will have a chronic progressive course which may occasionally rupture and become a sinus. In general in eosinophilic granuloma there will not be any

history of fever or other sign of inflammation. But the blood report may or may not show an elevated ESR.

Bone scan with technetium or gallium reveals a mass with enchancement and help in easy detection other foci or recurrence points. Ultrasound is helpful for guided biopsies. Biopsy help in histological diagnosis and treatment plan. Some patient disease gets cured without any treatment. But a few cases it may relapse especially during the first year. Treatment of choice for eosinophilic granuloma is surgical curettage with regular follow up. If it relapses then chemotherapy or further excision can be attempted (5).The present theory is inflammatory reaction/ autoimmune reaction causes an out of control langerhan cell proliferation and these langerhan cells secrete IL-1 and PG-E2 that cause bone destruction and disease progression(5).

Malignant transformation does not happen with Eosinophilic granuloma.Without bone involvement if it present elsewhere in the body it is called Hand-Schuller-Christian disease. Features include diabetes insipidus, cerebellar, hypothalamic and with other central nervous system symptoms(5).Age of diagnosis and the number of foci are important prognostic factors.

CONCLUSION

This is one of the rarest cases of langerhan cell histiocytosisof the skull. It is important to include langerhan cell histiocytosis (eosinophilicgranuloma) in the differential diagnosis of temporal bone. It is very much uncommon to see an isolated osteolytic lesion in the temporal bone. Without any involvement of the middle ear and mastoid . This is one of the first case report showing the presentation in a female child with postauricular swelling with showing only inflammatory change without any middle ear or mastoid disease.

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