

Intradural Langerhans' Cell Histiocytosis Invading Frontal Bone of Skull: A Case Report with A Brief Review

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1. Abstract

In Langerhans Cell Histiocytosis (LCH), there is a slow and abnormal proliferation of histiocytes. It is also called as Eosinophilic granuloma. In the case of bone involvement, there is an expansion of bone along with a lytic reaction. We present a 22 years male who presented with headache and solitary swelling over the right of the skull's frontal bone. In this case, the CH was originating from dura with secondary involvement of the bone. The X-Ray showed an osteolytic lesion of the frontal bone of the skull. CT scan finding showed a lytic area involving the inner and outer plate of the frontal bone. Intraoperative, the lesion was arising from Dura matter. It is quite rare to have a dural attachment. We could not find another case of frontal bone involvement with dural extension in the literature.

2. Introduction

The exact cause of LCH is still unknown and has varied manifestations. Earlier, Langerhans' cell histiocytosis is a reactive disorder rather than a neoplastic process, but recently, studies have supported its neoplastic origin [1]. In 1953, Lichtenstein proposed that the disease entities can be called an eosinophilic granuloma, Hand-Schuller-Christian disease, and Letterer-Siwe disease share a common pathologic appearance; these are analogous to LCH.

Other researchers disagree with this view [2]. It may present as single or multisystem CH. Its incidence is about 0.5-5.4 per million people. Its incidence is higher in children but can affect all age groups. It is more predominant in males (1.6-1.7 times more than females). In 70% of cases, it is solitary and least aggressive. Of Patients with the multisystem disease (30%), around 50% have equal or more than the one risk organ involved. It can affect bone (80%), lungs (15%), or lymph nodes (5-10%) [3]. Langerhans' cell histiocytosis affects one or many-body systems or tissues. It can affect bone, lung, brain, skin, lymph node, liver, and various other soft tissues. The extent of involvement of different body tissues decides the clinical picture. The patients with single organ disease have localized pain. The patients with disseminated disease may present with lymphadenopathy, skin lesions, or diabetes insipidus [2]. Treatment range from excision, chemotherapy, and radiation [3]. We report an unusual and first case of LCH involving frontal bone of the skull, arising from dura matter in a 22 yrs.' male who presented with headache and swelling.

3. Case Report

A 22-year male patient reported to the OPD with a history of headache and a swelling skull at the right frontal region of a 6-month duration (Figure 1). He gave a history of slow-growing swelling. There was no other relevant complaint. Examination revealed a 4

cm diameter swelling without tenderness and firm in-consistency. The overlying skin was normal. The systemic study revealed no abnormality. Aspirate revealed approximately 7-8 ml thin blood-stained fluid. Direct smears and centrifuged deposit were prepared and sent for cytology.



Figure 1: showing frontal Bone swelling

X-ray skull (anteroposterior, lateral, and tangential view) showed a well-defined soft tissue swelling over the right frontal bone. There was a destruction of the outer table frontal bone underneath the soft tissue swelling. A single well-defined lytic lesion was seen in the right frontal bone. Intracranial tension was within normal limits without any sign of intracranial calcification. The bony lesion confirmed the diagnosis of Histiocytosis-X on radiology and computerized scan (Figure 2, Figure 3).

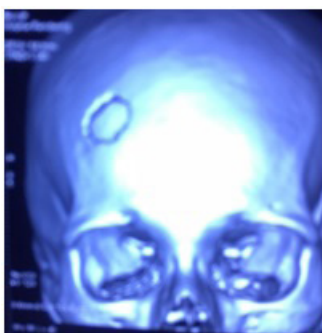


Figure 2: X-Ray showing the bony defect

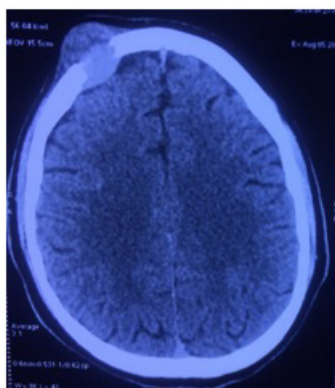


Figure 3: CT scan showing swelling of frontal Bone

The patient underwent frontal craniotomy. The lesion was originating from dura matter. The lesion was excised along with the involved dura and rim of the frontal bone (Figure 4). A Dural patch was created along with the periosteum, and the acrylic resin was used to repair the bone defect. Postoperatively, the patient was free from headaches.



Figure 4: Photo of an excised tumor with dura and bony rim

Histopathology of the tissue exhibited aggregates and sheets of histiocytes with some lying separately. The histiocytes showed nuclear folding, coffee bean grooving, and multinucleation. Scattered giant cells were seen. Many cells had voluminous bubbly cytoplasm. The RBC, lymphocytes, polymorphs, and several eosinophils were seen in the background. No cytophagy, epithelioid cells, filarial worm, or necrosis was seen. No mitotic figures were seen. The final opinion was histiocytic proliferative lesion suggestive of Langerhans' cell histiocytosis (Histiocytosis X) (Figure 5). Immunocytochemistry was also performed to confirm the diagnosis (Figure 6). The post-operative period was uneventful, and the patient was discharged after five days. The followed up was done for six months, and there was no recurrence after surgery.

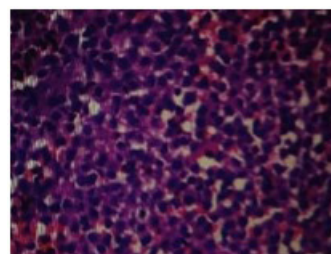


Figure 5: Slide of Histopathology



Figure 6: Immunocytochemistry slides

4. Discussion

LCH is an uncommon lesion associated with temporal, parietal bone, orbital bone, and mandible. The involvement of the frontal bone of the skull is infrequent, as in our case. LCH commonly occurs in the first four years of life but can present at any age. The incidence of LCH is 4-8 children per million [4-6] and 1 to 2 adults per million each year. More common in males and higher in whites of northern European descent [7]. LCH can present as a solitary lesion or as possibly a lethal disseminated disease. The disease process is, therefore, usually classified as three clinical variations based on organ involvement. These categories are named as eosinophilic granuloma, usually a solitary osseous lesion; Hand Schuller-Christian disease, the chronic recurrent form, which classically shows a triad of a skull lesion, exophthalmos, and diabetes insipidus; and the fulminant formation named as Letterer Siwe disease- multiple organ involvement [8-10].

Its etiology is unclear. It is a slow proliferative disorder of Langerhans' cells, but its nature, whether reactive, benign, or malignant, is still debatable. The virus or genetic may be predisposing factors, yet the exact of LCH cause is still unknown [11]. Recent research has discovered a clonal myeloid origin of the LCH cells and identified several activating mutations alongside the MAPK signal transduction pathway [12]. The progress decides the form of LCH, prognosis, course of the disease, single or multiple lesion, and onset.

The most frequently affected tissue is bones, followed by lung, lymph nodes, skin, liver, spleen, and reticuloendothelial system. The involvement of skull bones with brain or dura is uncommon, but it originated from the dura in our case. The pathophysiological phenomenon for intradural LCH is the migration of Langerhans cells with the dural membrane's inflammatory process [13]. In most patients, the presenting symptoms are swelling and headache or pain depending upon tissues' involvement.

The differential diagnosis for LCH is osteomyelitis of bone, meningioma, meningosarcoma, bony metastases, tuberculosis, sarcoidosis, bacterial and fungal abscesses, intracranial granulomas, malignant lymphoma [14, 15].

Langerhans' cell histiocytosis cells are frequently positive for S-100 protein. However, the most specific marker of Langerhans' cell histiocytosis is CD1a. CD1a expression can be perceived in frozen or paraffin-embedded tissue using the monoclonal antibody O10 [16]. In malignant lymphoma, the typical nuclear features of Langerhans' cell histiocytosis are absent.

According to the diagnostic standards of the international organization cell association for LCH in 1987, diagnosis of LCH depended upon the following staging: 1) the initial diagnosis bank on the results of the clinical laboratory, as well as bone and lung X-rays and common pathological changes (e.g., abnormal Langerhans' cells are shown to be present in pathological sections). 2) the di-

agnosis can be made if analysis by immunohistochemistry demonstrates a positive result for S-100 protein based on the preliminary diagnosis. 3) the diagnosis is made from a detailed examination of the initial diagnosis, along with an ultrastructural study that shows Birbeck particles or a positive result for CD1a expression by immunohistochemistry [11].

The radiographic features are usually quite characteristic. This disease mostly involves the skull, spines, and long bones with typical a hole-in-hole (punched out lytic) appearance because of the different destruction rates of the bone's two tables [3,17,18]. Radiology can indicate the presence of LCH, but the biopsy (histopathology) is essential for the final diagnosis.

Surgery, chemotherapy, or radiation are the treatment of choice [19]. The nature of therapy depends on the lesion's character, whether single or multiple, along with disease course. Surgery is the treatment of choice in the single and localized early lesion, as in our case. Radiotherapy or chemotherapy are required for multiple and late lesions. During surgery in our case, it was confirmed that this LCH originated from dura: i) complete dural attachment with round granuloma ii) LCH remained attached with dura matter after removing bone rim with granuloma (Figure 4).

The prognosis of the disease depends upon histologic features. The disease stage is more critical than the histologic appearance in predicting prognosis [20]. Kilpatrick et al. [21] have reported that the presence of hepatosplenomegaly, thrombocytopenia, young age at diagnosis, and polyostotic occurrence (three bones or more) are bad prognostic features.

5. Conclusion

Langerhans' cell histiocytosis is a rare disease; it is quite challenging to diagnose LCH due to the variable clinical and radiologic findings. Given this disease's rarity and unpredictable clinical course, one must have a high index of suspicion when interpreting the imaging findings. Frontal bone disease is often nonspecific and most commonly presents as a frontal bone mass with or without a headache. Headache is common if dura is involved, as in our case. Frontal bone with dura involvement is uncommon in an adult male.

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