A RARE CASE OF LANGERHANS CELLS HISTIOCYTOSIS OF THE SKULL

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BACKGROUND

Langerhans cells histiocytosis (LCH) is a rare disorder caused by the clonal proliferation of Langerhans cells (antigen-presenting cells). LCH is a type of dendritic cell that normally helps the body fight infection. Sometimes mutations in genes that control how dendritic cell function may cause too many LCH cells to grow and build up in certain parts of the body, where they can damage tissue or form lesions in one (single-system disease) or more places (multisystem disease). A family history of cancer or having a parent exposed to certain chemicals may increase the risk of LCH.[1]

CASE PRESENTATION

A 23 years old lady with underlying bilateral sensorineural hearing loss and previous history of smear-negative PTB in 2015, completed anti-tuberculosis treatment and presented with right frontal swelling for the past 8 months. The swelling progressively increased in size over the past 6 months and was soft and boggy. It was painless and there was no associated fever. CT brain showed right frontal subgaleal collection with multiple osteolytic changes. MRI brain revealed a right frontal subgaleal collection with intracranial extension to the right frontal extra- and intra-dural spaces which were suggestive of empyema. She was admitted and had a right craniotomy and tumor excision. The excised lesion consisting of skull bone, dura, and the frontal extradural tumor was sent for a histopathology examination.

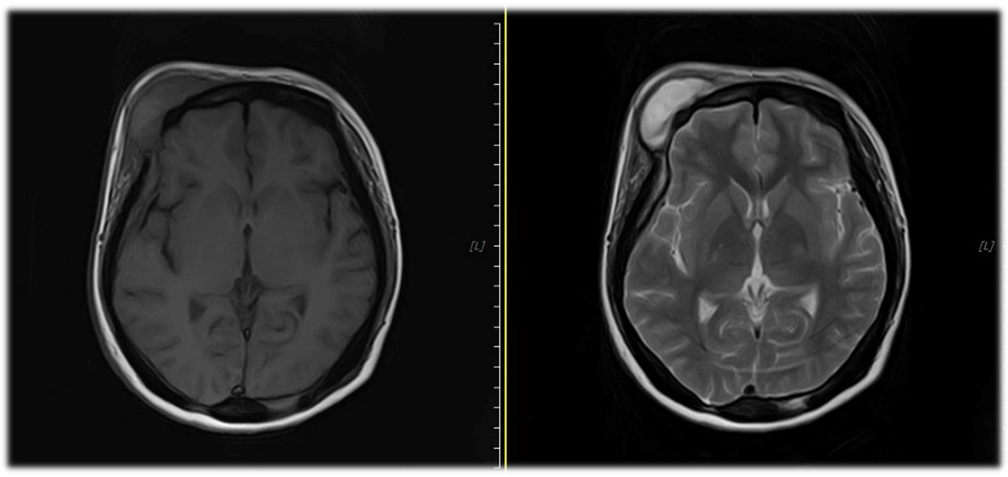

Figure 1: Lesion shows isointense on T1WI, and hyperintense on T2WI/FLAIR. The underlying right frontal bone underneath the collection is eroded with periosteal changes. Intracranially, there is a peripherally enhancing area seen at the right frontal extra-axial region with adjacent dural thickening.

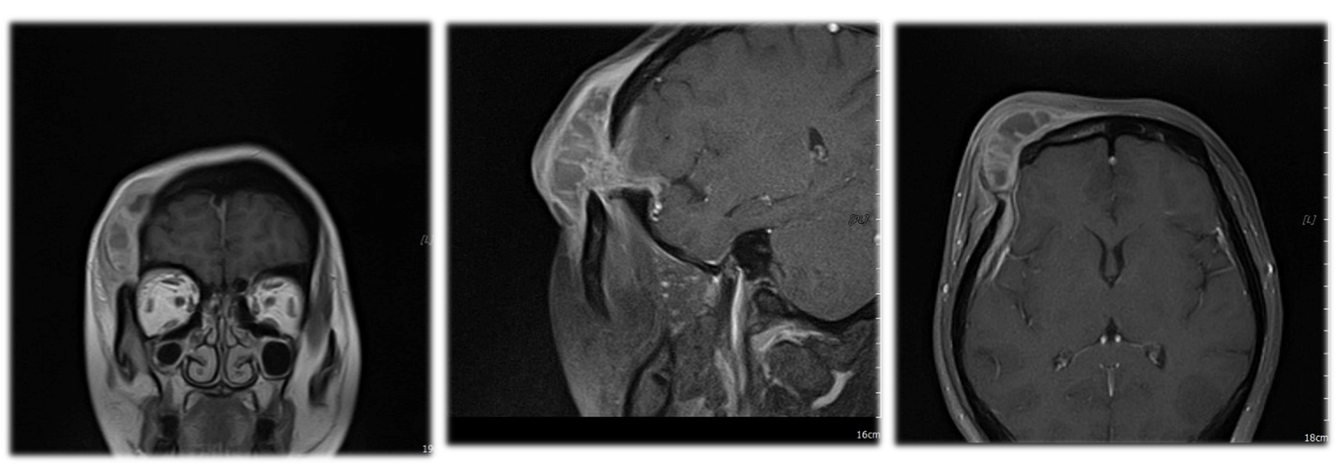

Figure 2: Shows peripherally enhancing extradural collection with numerous enhancing septa seen at the right frontal scalp region with mild extension to the right periorbital region inferiorly.

DISCUSSION

The presence of cortical destruction, endosteal scalloping, and a periosteal reaction on CT or MRI; the margin of soft tissue masses, the presence of bone marrow edema, and a “budding” appearance on MRI; and the presence of sclerotic margins or septations on CT may be helpful signs for differentiating LCF from malignant tumors in adult.[2] Early lesions appear aggressive with poorly defined margins and lamellated periosteal reaction. Late lesions appear well-defined and may show sclerotic margins and an expanded remodeled appearance.[3]

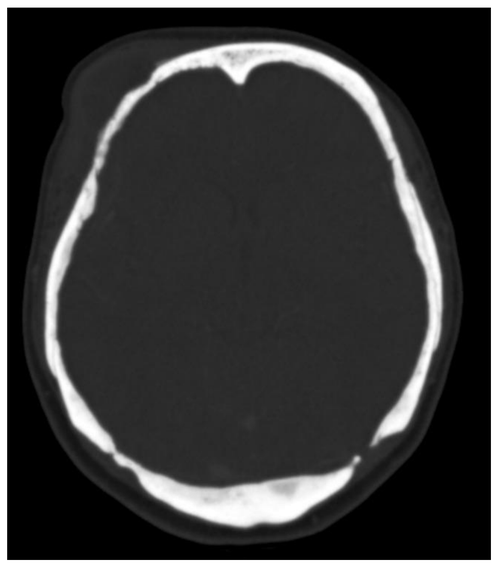

Figure 3: shows right frontal irregular cortical bone destruction.

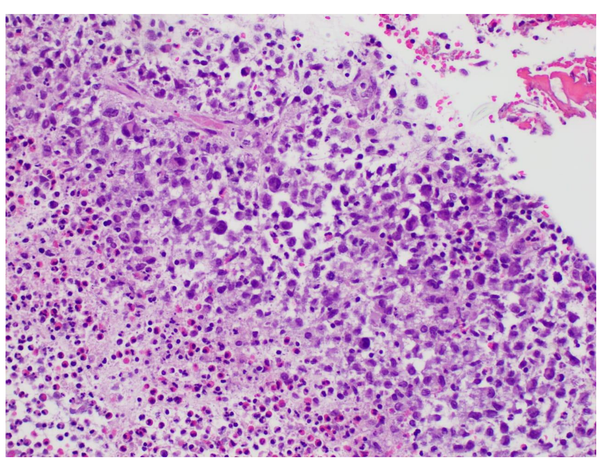

Figure 4: showed fragments of fibrous tissue exhibiting proliferation of neoplastic cells in sheets and clusters, displaying irregular cleaved nuclei with prominent nuclear grooves and folds, fine chromatin, indistinct nucleoli, and moderately abundant pale eosinophilic cytoplasm

CONCLUSION

It is of utmost importance to interpret the scan and the case to make an astute decision for operative planning. There was a high index of suspicion that it was not merely a case of empyema, hence dura and bone was sent for Histopathology examination (HPE) as well. It is of vital importance to send HPE for suspicious cases to prevent missed or late diagnosis, as Langerhans Cells Histiocytosis and empyema are two distinct conditions with two different treatment regimes.

REFERENCE

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