Annals of Short Reports and Clinical Images

Clinical Image

An Atypical Orbital Langerhans Cell Histiocytosis

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Clinical Image

A 3-year-old female with a one-month history of rapidly progressive, non-tender, well-defined cystic mass in the superior-temporal left orbit, extending to supra-orbital, infra-orbital and temporal region presented to the Advanced eye center, triage area in August 2020 (Figure 1A). She had no history of ocular trauma, fever, lymphadenopathy, or previous hospitalization. The patient had severe mechanical ptosis with an overlying bluish hue, dilated vessels, and 7 mm inferior globe dystopia with 30-degree esotropia, and limitation on attempted abduction, elevation, and Levo-depression was observed. Anterior segment and fundus examination were within normal limits. Contrast-enhanced CT scan head and orbit revealed a 28 mm X 30 mm heterogenous enhancing irregular superior-temporal cystic orbital mass involving the superior and lateral rectus muscles with overlying frontal vault erosion (Figure 1B-D). Blood counts, liver and renal functions, chest X-Ray, and abdominal ultrasound were within normal limits. Orbital mass with evidence of bony erosion, rapidly progressive course, absence of inflammation in the first decade of life suggests malignant etiology. Ultrasound-guided fine-needle aspiration cytology revealed pleomorphic, atypical cells with nuclear grooving against osteoclast-like giant cells and eosinophilic background. Immunocytochemistry was strongly positive for CD1a and CD207/Langerin confirmed Langerhans Cell Histiocytosis (LCH) Figure 2. Urine osmolarity and Bone Skelton survey revealed normal study, therefore, our patient was diagnosed as Atypical cystic uni-system unifocal orbital Langerhans cell histiocytosis. Extensive degenerative changes in the entire mass simulate a cystic lesion creates a diagnostic dilemma. The patient responded well to the induction phase of systemic chemotherapy-vinblastine, prednisolone and cotrimoxazole and currently in the continuation phase of chemotherapy.

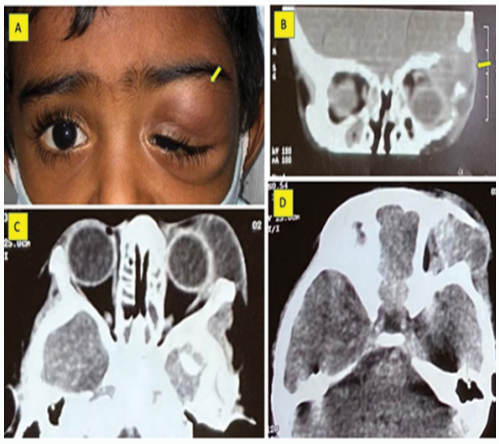

Figure 1: Clinical photograph of 3-year-old child at presentation shown by arrowhead (A). Contrast enhanced CT images- coronal cut section indicating osteolytic lesion of roof and lateral orbital wall (B). Axial CECT cut section demonstratingheterogenous enhancing, superior-temporal cystic orbital mass (C) with overlying frontal vault erosion (D).

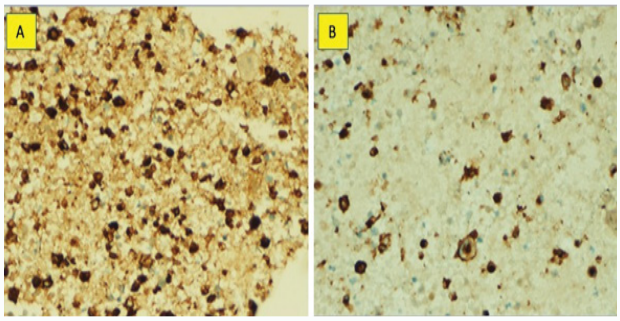

Figure 2: Photomicrograph of lesion -Positive immunostain with CD 1a (A) and CD207/Langerin (B).

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