Langerhans cell histiocytosis of pituitary stalk

Harsh Kandpal, Subramanian Subramanian, Smriti Hari

Department of Radiodiagnosis, All India Institute of Medical Sciences, New Delhi - 110 029, India

A four-year-old male child presented with increased urine output and excessive thirst for eight months. On examination he had multiple scalp swellings with mild hepatosplenomegaly. Urine specific gravity was decreased suggestive of diabetes insipidus. Skull radiograph revealed multiple well-defined geographic lytic lesions in the calvarium with bevelled margins [Figure 1]. On MR examination of sella, the posterior pituitary bright spot was not seen on T1W image [Figure 2]. The pituitary stalk was thickened and showed uniform enhancement [Figure 3]. The characteristic radiographic feature of skull lesions, MRI findings and hepatosplenomegaly strongly suggested the possibility of Langerhans cell histiocytosis (LCH) which was confirmed on bone marrow biopsy.

LCH is a wide-spectrum disease characterized by proliferation of abnormal histiocytes. It encompasses three classical clinical syndromes which in increasing order of severity are: eosinophilic granuloma (solitary bone lesion), Hand-Schuller-Christian disease (skull lesion, exophthalmos and diabetes insipidus) and Letterer-Siwe disease (multisystemic rapidly progressive form). The skull bones are most commonly involved. Typically the lesions are round or oval-shaped with well-defined margins and bevelled edge. Individual lesions may coalesce giving a geographic appearance to the skull

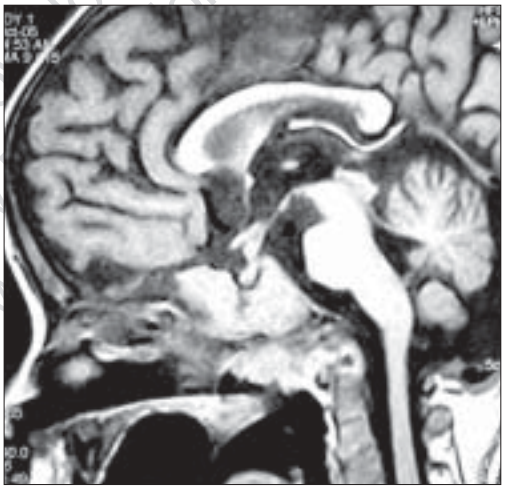

Figure 2: T1W sagittal image. The bright spot of the posterior pituitary is not seen and the pituitary stalk is thickened. Isointense soft tissue mass is seen in the clivus and sphenoid bone

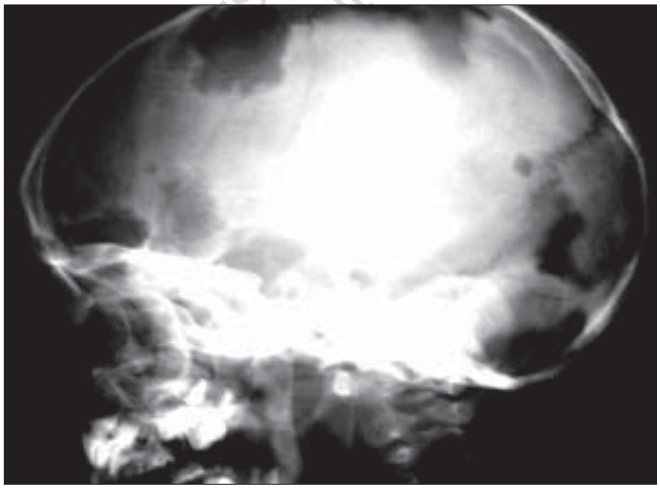

Figure 1: Skull radiograph lateral view shows multiple varying sized geographic lytic lesions with beveled margins

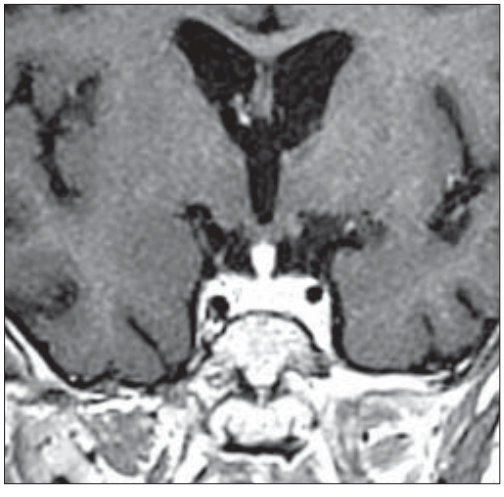

Figure 3: T1W postgadolinium coronal image shows the thickened and enhancing pituitary stalk

Subramanian Subramanian
Department of Radiodiagnosis, All India Institute of Medical Sciences, New Delhi - 110 029, India. E-mail: drsubbusmc@yahoo.co.in

vault. A button sequestrum may be seen within them. Diabetes insipidus although rare at initial presentation may occur in 25% patients during the course of the disease.[1] Cranial MRI findings in LCH include thickening and abnormal enhancement of the pituitary stalk with absence of posterior pituitary hyperintensity.[1,2] Infundibular involvement may be diffuse or focal[1] and may precede other typical manifestations of LCH.[3,4] Although germinoma, neurosarcoid, tuberculosis and occasionally idiopathic diabetes insipidus can cause pituitary stalk thickening, clinical and radiological features usually suggest the diagnosis of LCH, which can be confirmed on biopsy from lesions in the bone, bone marrow, lymph node or skin lesions.

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