A rare case of Langerhans cell histiocytosis of the skull in an adult: a systematic review

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Abstract

We report a 41-year old male who presented to the Emergency Department after falling while water-skiing. He had a previous medical history included chronic headaches, which had persisted for the last 2-3 months prior to presentation. Computed tomography of the head showed a small hypersensitivity with a small extra axial collection with a maximum thickness of 1mm. Differential diagnoses included an arachnoid cyst, haemangioma, meningoima or a secondary lesion. A diagnosis of Langerhans Histiocytosis was made based on the histopathology examination and the immunoperoxidase staining.

Introduction

Langerhans cell histiocytosis (LCH) is a rare condition that usually affects the pediatric population. The condition can affect almost any organ in the body. It is a rare condition with an estimated annual incidence between 1-7 cases per million of the population. More than 50% of patients are less than two-years of age have disseminated LCH with organ dysfunction and die of the disease. The prevalence of LCH seems to be higher among whites than other races. The incidence of LCH is greater in males than in females, with a male-to-female ratio of 2:1. Recent early data from the U.K. suggest that just as many cases of LCH present in adult life as in childhood. However, it should be noted that at present it remains unclear if these adult cases are de novo or if they are occult cases from childhood. We report a case of adult LCH with his presentation, medical history, clinical characteristics, and radiological findings as well as the success of the surgical intervention.

Case Report

A 41-year old male who presented to the Emergency Department after falling while water-skiing at 40-50 km/hr. He landed face first onto the water but did not lose consciousness and remembered that he was in trouble.

The patient recalls his face feeling puffy for 10-15 min but then settled and both his eyes were bleeding for approximately 30 min. He complained of neck and right shoulder pain and a frontal headache. His previous medical history included surgery for lumbar disc prolapse and chronic headaches, which had persisted for

Figure 1. X-ray showing lesion on skull.

Figure 2. Histopathology of lesion removed from skull.
Discussion and Conclusions

Langerhans cell histiocytosis is a clonal proliferative disorder of the antigen presenting Langerhans' cell and refers collectively to a group of diseases previously known as histiocytosis X, eosinophilic granuloma, Letterer-Siwe disease, Hand-Schuller-Christian syndrome, Hashimoto-Prizker syndrome, self-healing histiocytosis, pure cutaneous histiocytosis, Langerhans cell granulomatosis, Type II histiocytosis and non-lipid reticuloendotheliosis. Langerhans cell histiocytosis usually affects patients under 10 years of age, however, studies have reported that the average age for presentation is 25 years. Patients mainly present with localized bone pain, dyspnea and malaise, and 75% have non-disseminated disease. The skull, femur, pelvis and ribs are most commonly involved. With skull lesions, the orbit and the cranial base are frequently involved and produce the classic triad of bony defects, exophthalmos and diabetes insipidus.

Our adult case exemplifies an uncommon presentation of LCH which more commonly occurs in pediatric populations. However, this case illustrates a classic presentation of LCH which more commonly occurs in pediatric populations. However, this case illustrates a classic presentation of LCH.
in adults. Osteolytic bone lesions are a common manifestation of single system LCH in adults. These lesions tend to be unifocal rather than multifocal, often involving the skull or axial skeleton, such as in our patient. Our review of skull vault lesions in adults (Table 1) has confirmed this observation with 4 out of 6 cases of LCH involving the skull having unifocal lesions. Calvarial lesions are normally found incidentally as was the case with our patient. However they may also present with bone pain, soft-tissue swelling, hearing loss, vertigo and visual disturbances. Histologically, proliferation of Langerhans cells expressing CD1a and S100 admixed with acute and chronic inflammatory cells are consistently reported. Immunohistochemical findings in our patient were consistent with these features. This report is interesting and adds valuable information to limited literature available on unifocal skull vault lesions in adults with LCH. Local treatment with excision, systemic chemotherapy and corticosteroid injection is highly successful in treating this disease and patients have excellent prognosis. Over 90% of patients survive 3 to 5 years post diagnosis. Age at diagnosis and initial response to therapy affect the prognosis and rate of recurrence of disease.

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