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Case Report

Radio-Cytopathological Diagnosis of Eosinophilic Granuloma of Skull in a 13-Year Old Boy

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ABSTRACT

Eosinophilic granuloma is benign end of the spectrum of the Langerhans cell histiocytosis (LCH) which is characterized by solitary or multiple lesions in bones, skin, lung, lymph node etc. Here, we present a case of a 13-year old boy with pain and swelling in the right parietal region of skull with no other complaint. A computerized tomography (CT) scan and subsequent fine needle aspiration cytology (FNAC) revealed solitary eosinophilic granuloma which was subsequently confirmed by histopathology. Minimally invasive procedures like imaging and FNAC usually suffice for diagnosing and following up of patients with this rare disease.

Keywords: **Eosinophilic Granuloma; Fine Needle Aspiration, X-Ray CT Scan**

Introduction

Langerhans cell histiocytosis (LCH) is a rare group of disorders, which falls in the category of clinical syndromes called histiocytoses that are characterized by an abnormal, neoplastic proliferation of histiocytes. Various terms like 'Histiocytosis X' and Langerhans cell granulomatosis have been historically applied to describe this group of disorders. The incidence of LCH is about five per million with most cases occurring in childhood (1). Three major overlapping syndromes are recognized. There may be unifocal disease (eosinophilic granuloma), multifocal, unisystem disease (Hand-Schüller-Christian disease) and multifocal, multisystem disease (Letterer-Siwe disease). Eosinophilic granuloma is usually seen in children, adolescents and young adults presenting with solitary, often multiple lytic skeletal lesions although other organs like skin,

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lung, and lymph node may be affected (2). Solitary lesions usually have good prognosis and are extremely radiosensitive; some lesions even showing spontaneous regression (3).

Here, we describe a case of a 13-year-old boy who presented with a swelling in the right parietal region. A computerized tomography (CT) scan followed by fine needle aspiration was done and the boy was diagnosed as suffering from eosinophilic granuloma.

Case report

A 13-year-old boy presented to the outpatient department with a painful swelling in the right lateral part of the head for the last three weeks.

There was no history of trauma. The patient had no other complaint. On examination, a swelling of about three cm diameter was seen in the right parietal bone, which was soft and tender. The underlying bone appeared to be indented on palpation. The patient had no lymphadenopathy, organomegaly, skin rash or any other abnormality. The complete blood counts and chest X-ray were within normal limits. A CT scan and fine needle aspiration was advised.

The scan revealed a solitary osteolytic lesion in the right parietal bone with erosion of both the inner and outer table (Fig. 1). The aspiration was done from the skull lesion by a 23-gauge needle fitted with a 10 cc syringe.

The wet-fixed smears were stained with Papanicolaou stain and the air-dried smears were stained with May-Grunwald-Giemsa (MGG) stain. On microscopical examination, the cellular smears showed presence of many histiocytoid cells with moderate amount of cytoplasm and folded indented, often-reniform nuclei. The most characteristic feature was the presence of nuclear grooves (Fig. 2 and 3). The nucleoli were inconspicuous and mitosis was rare. Few binucleate and multinucleated cells were also noted. Another characteristic feature was the background, which showed plenty of eosinophils with few neutrophils and lymphocytes. Based on the radiological and cytopathological findings, a diagnosis of eosinophilic granuloma was made. A subsequent histopathological examination followed by immunohistochemistry confirmed the diagnosis.

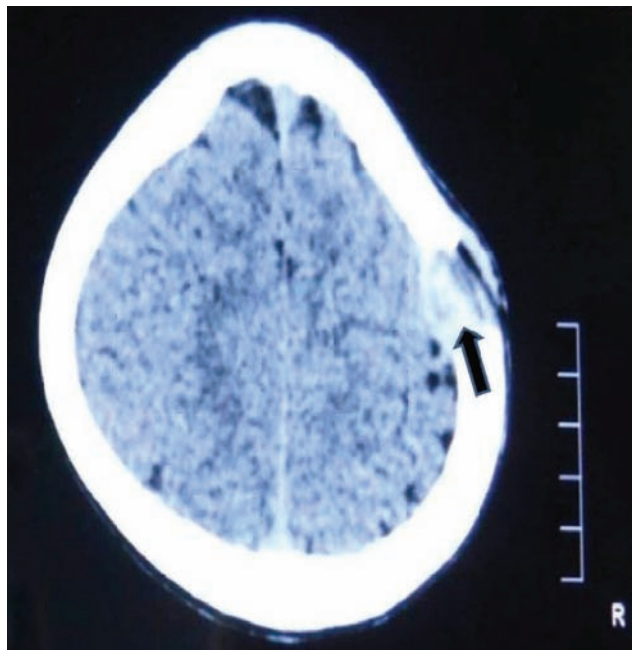

Fig. 1- Photograph of CT scan showing osteolytic lesion in skull with destruction of inner and outer table (arrow)

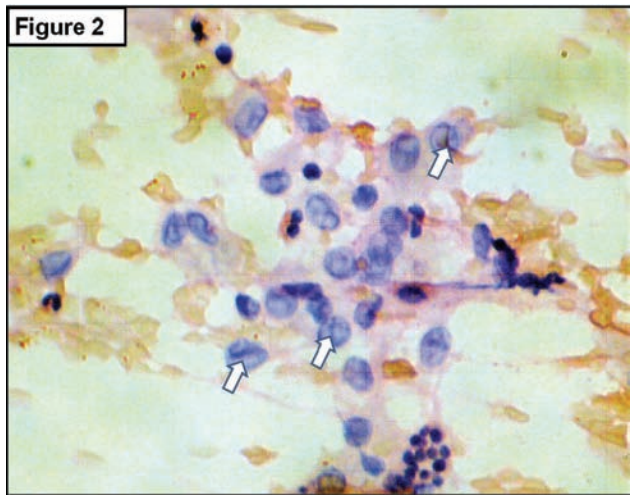

Fig. 2 - Photomicrograph showing Langerhans' histiocytes with reniform nuclei and nuclear grooves (arrows). Eosinophils and RBCs in the background (Papanicolaou stain; × 100 magnification)

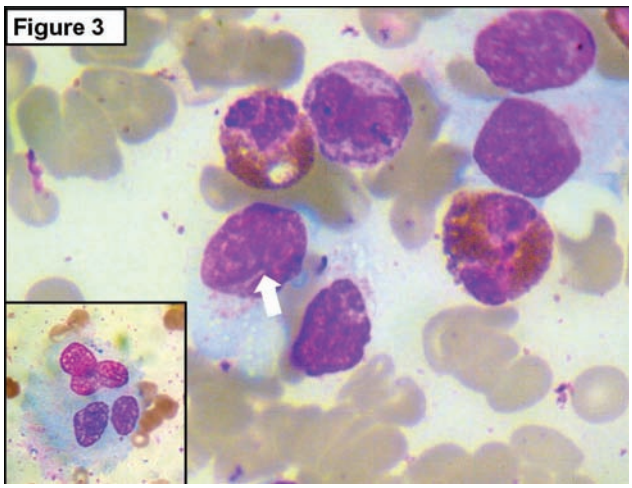

Fig. 3 - Photomicrograph showing Langerhans' histiocytes with nuclear grooves (Arrow) with eosinophils and a neutrophil. Inset shows a multinucleated histiocyte (MGG stain; × 400 magnification)

Discussion

Eosinophilic granuloma is the benign form of the spectrum of Langerhans Cell Histiocytosis (LCH). The acute, severe form in infants known as Letterer-Siwe disease present as a fulminant, disseminated disease while the intermediate Hand-Schuller-Christian disease in older children and young adults present with diabetes insipidus, proptosis and lytic bone lesions. Cutaneous lesions are more common in the Letterer-Siwe disease. Eosinophilic granuloma, predominantly affecting the children (with a median age of diagnosis being 3.8 years) is characterized by single or multiple osteolytic skeletal lesions. There is a predilection for males (male: female ratio 3.7: 1) and whites of Northern European descent (2). Any bone can be affected but the more common sites include the skull, mandible, spine, ribs, and the long bones (4-6). Extra-skeletal sites like lung, lymph nodes and skin can also be involved (2, 7, 8) .

Our patient was a 13-year-old boy with complaints of a tender, soft swelling in the right parietal region of the head. Although a fraction of patients may be asymptomatic, the majority of patients with eosinophilic granuloma in bone present with pain, swelling and sometimes with pathological fractures. Radiologically, osteolytic lesions are most common as was seen in our case. While neither clinical nor radiological evidence is sufficient for diagnosis, fine needle aspiration cytology appears sufficient to clinch the diagnosis. The characteristic appearance of Langerhans' histiocytes with grooved, indented, reniform nuclei and plentiful cytoplasm lying in a background of variable number of eosinophils, neutrophils, and lymphocytes helps in the cytological diagnosis. The important differentials include osteomyelitis, Ewing's sarcoma, Hodgkin's lymphoma, and sometimes melanoma (7, 9). Aspirates from osteomyelitis lack the eosinophil-rich background and characteristic nuclear features of histiocytes as seen in eosinophilic granuloma although both clinically and radiologically it is often very difficult to differentiate these two entities. Ewing's sarcoma can easily be differentiated cytologically by the presence of small, round cells with increased nuclear to cytoplasmic ratio and nuclear moulding along with few pseudorosettes. The background, consisting of eosinophils, neutrophils and histiocytes, also make Hodgkin's lymphoma a very close differential. However, the demonstration of Reed Sternberg cells usually clinches the diagnosis in favor of Hodgkin's lymphoma (9). Melanoma cells usually lie dispersed or in loose clusters. The cells exhibit eccentric nuclei, binucleation and occasional intracytoplasmic or nuclear inclusions while the amount of melanin pigment is variable.

Ultrastructural demonstration of Birbek granules, immunocytochemistry using CD 1a (10) and enzyme cytochemistry with alkaline phosphatase (2) are helpful methods to reliably confirm the diagnosis. Apart from the traditional immunohistochemical markers like CD 1a and S-100, Langerin (CD 207) is a recently identified lectin for which antibodies can be used as immunohistochemical markers of Langerhans

cells (10, 11).

The prognosis of these patients depends on the number of organs affected at presentation, with an overall survival of 95% for patients with unifocal disease. However, 10% of patients with unifocal disease eventually progress to multisystem disease (2). As per Kaul *et al.*, radiologists need to be aware that additional eosinophilic granuloma of the bone occurring as long as four years after initial diagnosis, should be interpreted as a localised form of Langerhans cell histiocytosis (8). Thus, the patient of even solitary eosinophilic granuloma needs a prolonged follow-up for as long as five years or more. This is important for the treatment. While solitary lesions respond well to local excision or limited radiation, chemotherapy is needed for multisystem diseases.

To conclude, a characteristic cytopathological picture coupled with radiological evidence is helpful in diagnosing as well as following up the patients of this rare disease.

Acknowledgments

The authors declare that there is no conflict of interests.

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