CASE REPORTS

EOSINOPHILIC GRANULOMA OF MASTOID WITH SKIN LESIONS AND DIABETIS INSIPIDUS

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A case of eosinophilic granuloma of pars mastoida of temporal bone is reported in a 6-year-old boy. He had diabetes insipidus and skin lesions suggestive of histiocytosis X, clinically and histologically. Recent views about pathogenesis of histiocytosis X are discussed.

Key words : Eosinophilic granuloma, Histiocytosis X, Diabetes insipidus.

Histiocytosis is a group of closely related diseases characterised by the proliferation of histiocytes due to an unknown cause. Three clinical forms are recognized, viz. Letterer-Siwe disease, Hand-Schuller-Christian disease and eosinophilic granuloma. Eosinophilic granuloma is the most benign form and presents usually as a single bone lesion in older children and adults. Letterer-Siwe disease on the other hand is the most severe and manifests in the first year of life with often fatal visceral involvement. Hand-Schuller-Christian disease occurs during early childhood and presents with multiple bone lesions but with less visceral involvement. The three conditions are not demarcated sharply from each other and thus transitional forms are common. Skin lesions are seen most commonly in Letterer-Siwe disease but occasionally in the other two types also. The common types of skin lesions observed are the scaly papular and crusted lesions simulating seborrhoeic dermatitis, infiltrated plaques with or without ulceration and xanthomatous lesions. Histologically, one observes a proliferative, a granulomatous or a xanthomatous infiltration in the skin and viscera, depending on the type of skin lesions and the stage of the disease. The predominent cells in the infiltrate are large histiocytes having abundant, slightly eosinophilic cytoplasm and large pale irregular shaped nuclei.

On electron microscopic examination, Langerhans granules are seen in the cytoplasm of about one half of the histiocytes present in the infiltrate in all three forms of histiocytosis X[1]. These granules are morphologically similar to the granules seen in the epidermal Langerhans cells and hence Pinkus prefers the term 'L cell granulomatosis' instead of histiocytosis, for this group of diseases. We are reporting a case of eosinophilic granuloma of the pars mastoida of the temporal bone with skin lesions and diabetes insipidus.

Case Report

A six-year old boy was seen with asymptomatic scaly papular and crusted lesions on the scalp, neck and upper part of the chest since two years. He had recurrent attacks of purulent discharge from the left ear since three years and polyuria since one year. He had received various systemic antibiotics and topical antiseborrhoeic preparations, but without much benefit. The developmental milestones were normal. There was no parental consanguinity and none in his family suffered from a similar disease. Dermatologic examination revealed multiple, bilateral, papular, scaly and crusted lesions over the scalp, front and sides of the neck and upper part of the

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chest. Auspitz sign was negative. Hairs and nails appeared normal. There was no significant lymphadenopathy. The left ear had purulent discharge and granulation tissue in the external auditory canal. Tympanic membrane was not visible. There was no swelling of the mastoid region but it was slightly tender. Nose, throat and the right ear were normal. Facial nerve was intact. Ophthalmologic examination including a fundoscopy did not reveal any abnormality. All other systems were clinically normal.

Routine examination of blood and urine were normal. Average daily total output of urine was 3.8 litres, the specific gravity of urine being 1010. The serum proteins, lipids, calcium and phosphate values were within normal limits. Blood sugar and urea also were normal. Liver function tests showed normal levels of serum bilirubin, SGOT, SGPT and alkaline phosphatare. X-ray of the skull showed a punched out lesion of the mastoid region on the left side. All other bones were normal. Biopsy of the granulation tissue taken from the left external auditory canal showed a granulomatous reaction consisting of aggregates of histiocytes, lymphocytes, multinucleated giant cells and a few eosinophils and neutrophils. The skin biopsy taken from the neck showed a proliferative reaction consisting of an extensive infiltration of the upper dermis. The infiltrate invaded the epidermis in some areas. The histiocytes in the infiltrate appeared as large cells with irregularly shaped vesicular nuclei and abundant, slightly eosinophilic cytoplasm. A few lymphocytes and eosinophils were also seen.

Comments

The first case of eosinophilic granuloma was reported by Finzi in 1929. He described it as a myeloma with eosinophils. The term histiocytosis X for the three diseases– Letterer-Siwe disease, Hand-Schuller-Christian disease and eosinophilic granuloma, was introduced by Lichtenstein in 1953[2]. Eosinophilic granuloma represents a monosymptomatic form of histiocytosis X and involves one organ system only, usually the skeleton with one or at the most a few lesions. But irrespective of the presence or absence of bone lesions, cases with skin and oral lesions, pulmonary lesions and even those with diabetes insipidus have been classified as having eosinophilic granuloma [3-5]. Sood et al [6] in 1973 reported a case of eosinophilic granuloma of temporal bone with cutaneous lesions. Similar cases were reported by Mecreary in 1942[7] and Curtis and Cawley in 1947[8].

The present case had eosinophilic granuloma of pars mastoida of the temporal bone on the left side which presented as granulation tissue in the external ear canal with purulent discharge. The skin lesions in our case closely simulated the seborrhoeic dermatitis, which on histology showed a proliferative reaction of histiocytosis.

Histology of the granulation tissue from the ear canal also proved to be eosinophilic granuloma. Low specific gravity of the urine and symptoms of polyuria and polydypsia in the present case suggested a diagnosis of diabetes insipidus. Considering the high risk to the patient, water deprivation test was not performed. Diabetes insipidus in histiocytosis is usually the result of a granulomatous infiltration of either the posterior pituitory gland, the tuber cinereum or the hypothalamus. Rarely, cases originally diagnosed as eosinophilic granuloma have progressed into typical Hand Schuller Christian disease[9]. Such a possibility cannot be completely excluded in the present case where it started as ear symptoms and progressed with the development of diabetes insipidus and skin lesions.

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