Case Report

EOSINOPHILIC GRANULOMA OF THE TEMPORAL BONE

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ABSTRACT

Eosinophilic granuloma is the benign form of histiocytosis X, or Langerhan's cell histiocytosis. The most frequently affected bones include skull, pelvis, ribs and long bones. Involvement of the temporal bone is a rare manifestation of eosinophilic granuloma. When the disease occurs in the temporal bone, the patient may commonly present acute or chronic otitis media. We report a case of eosinophilic granuloma of the temporal bone presenting with purulent discharge.

Key Words: Eosinophilic granuloma, Temporal bone

INTRODUCTION

The term histiocytosis X (Langerhan's cell histiocytosis), proposed by Lichtenstein in 1953, refers to a diverse group of disorders of unknown etiology, characterized histologically by a proliferation of differentiated histiocytes in various body tissues (1). The stimulus for proliferation is also unknown, with theories including metabolic, genetic, infectious and malignant causes. This entity is composed of three diseases complexes that share similar pathological findings but different severity, prognosis and treatment: Letterer-Siwe disease, Hand-Schüller-Christian disease and Eosinophilic granuloma (2).

Letterer-Siwe disease is the most severe form. It usually affects infants and young children. It is manifested by fever, rash, lymphadenopathy, hepatosplenomegaly, bone marrow involvement and various visceral involvements.

Hand-Schüller-Christian Syndrome is also a systemic disease affecting children aged 1-5 years and clinical features include bony invasion, skin, brain and viscera involvement. Classic triad of the disease; diabetes insipidus, osteolytic skull lesions and exopthalmus secondary to orbital bone involvement, are seen in up to 25% of patients.

Eosinophilic granuloma is the mildest form of Histiocytosis X. It usually affects older children and adolescents but it may appear in adults too. Most commonly only one bony site is involved, although multifocal osseous involvement does occur. Skull, femur, pelvis, tibia, ribs, vertebras, and humerus are the most commonly affected sites (3). It is reported that in 15-61 % of cases with Langherhan's, cell histiocytosis (LCH), temporal bone involvement is seen (3-5).

CASE REPORT

An 11-year-old boy presented with a 2-week history of left aural discharge. Physical examination revealed purulent discharge and a granulomatous polyp blocking the external ear canal. General physical examination was normal. Results of various laboratory tests including blood count, urinalysis and blood chemistry were within normal limits. Pure tone audiometry showed a conductive hearing loss of 38 dB (pure tone average) on the left side, and a sensorineural hearing loss of 60 dB at only 6000 Hz and otherwise normal hearing on the right side. Speech discrimination score was 92 percent. Chest roentgenogram and bone survey, with the exception of left temporal bone, were normal. Computed tomography of the temporal bones showed a left sided destructive soft tissue density mass filling the

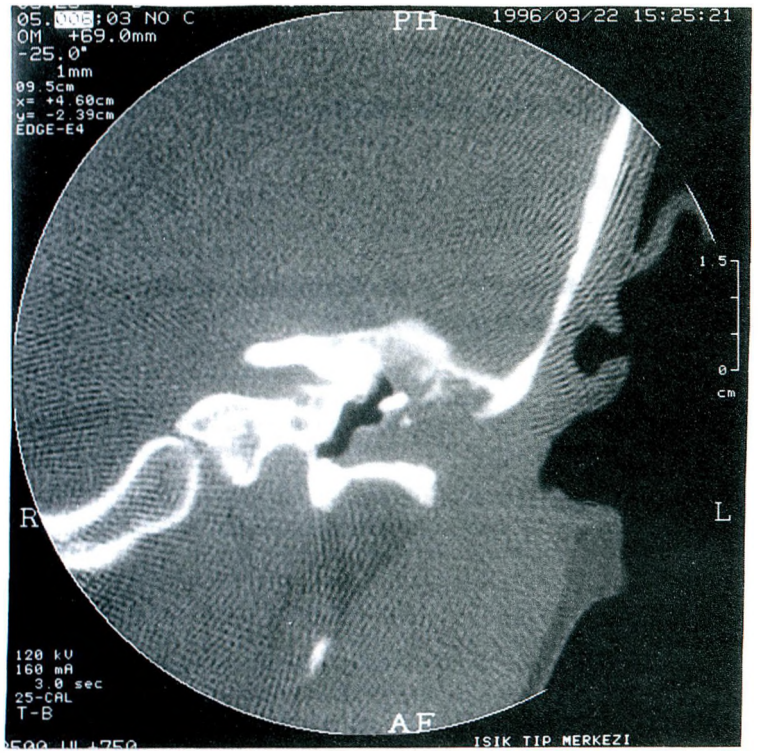

Fig. 1 A - Coronal

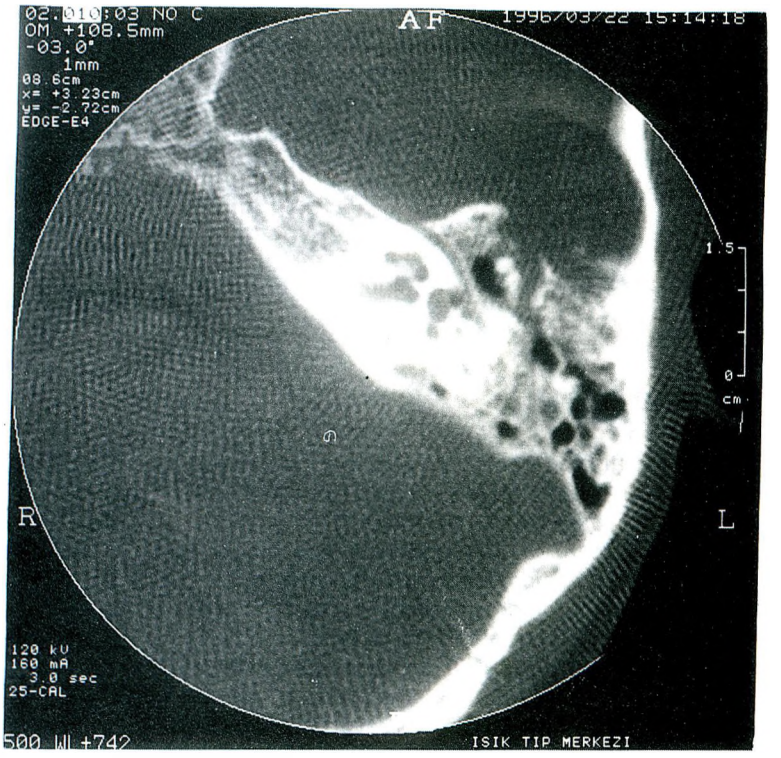

Fig. 1 B - Axial

Fig. 1: Preoperative computerized tomogram shows erosion of the left temporal bone.

attic and mastoid cavity (Figs 1a, 1b). Biopsy of the mass through the left external ear canal was reported as inflammatory polyp with eosinophilic infiltration. Surgery of the left ear was planned. Surgical exploration revealed a soft tissue mass filling the mastoid cavity and epitympanium, eroding the

posterior wall or the external ear canal and middle fossa dural plate. The mass was in contact with the middle fossa dura. Ossicular chain was suprisingly intact. The mass was completely removed from the mastoid cavity. A modified radical mastoidectomy was completed. Histopathological examination revealed congested tissue fragments with a dense cellular infiltrate. Variable proportions of histiocyte-like cells, eosinophils, leuococytes, lymphocytes, histiocytes were seen in different fields. Focal eosinophil clusters and degranulation were prominent. Histiocyte-like cells had indentated, grooved or lobulated nuclei and abundant pale staining granular cytoplasm. Mitoses were rare. There was no necrosis. Immunostaining with S-100 protein monoclonal antibody revealed cytoplasmic staining in histiocyte-like cells (Fig. 2).

Postoperatively the patient was treated with low dose radiation therapy to the left ear and temporal bone region. Computed tomography of the left temporal bone performed 3 months postoperatively, showed a normally pneumatized mastoidectomy cavity with no mass (Figs. 3a, 3b). Post-operative hearing revealed a small conductive hearing loss of 15 dB. Speech discrimination score was the same. The patient is presently free of disease 13 months after the operation.

DISCUSSION

Langerhan's cell histiocytosis is an insidious disease of the reticuloendothelial system mostly appearing in children. Although ear and temporal bone involvement is more likely to occur in children with multisystem disease, it may be the only manifestation of the disease. In a review of 62 cases of histiocytosis, 18 had temporal involvement of which 12 had multisystem disease and only one child had unifocal osseous LCH (2). Temporal bone involvement is usually unifocal and bilaterality is rare occuring in about 30% of cases (3,6-8). Involvement of petrous portion is also rarely seen (9).

Most common presenting otologic symptom is otorrhea followed by postauricular swelling, hearing loss and aural polyp. Hearing loss is usually conductive type and due to canal and middle ear obstruction by soft tissue but may also be due to ossicular erosion. Rarely sensorineural loss, vertigo and nystagmus can be seen suggesting otic capsule involvement. Although the facial nerve is exposed in the middle ear in 50% of cases, facial nerve paralysis is rarely seen (8). Otalgia is infrequently reported (3-5). Cranial nerve paralysis may be seen due to skull base invasion. A rare case with eosinophilic granuloma of the temporal bone which had intracranial hypertension as a presenting sign due to lateral sinus involvement, was also reported (10).

On physical examination, there may be granulation tissue on the posterosuperior ear canal wall. When combined with physical signs, manifestations of the disease can easily be attributed to acute or chronic infectious disease of the middle ear as in the case represented here.
Classically on plain x-ray, radiolucent areas with

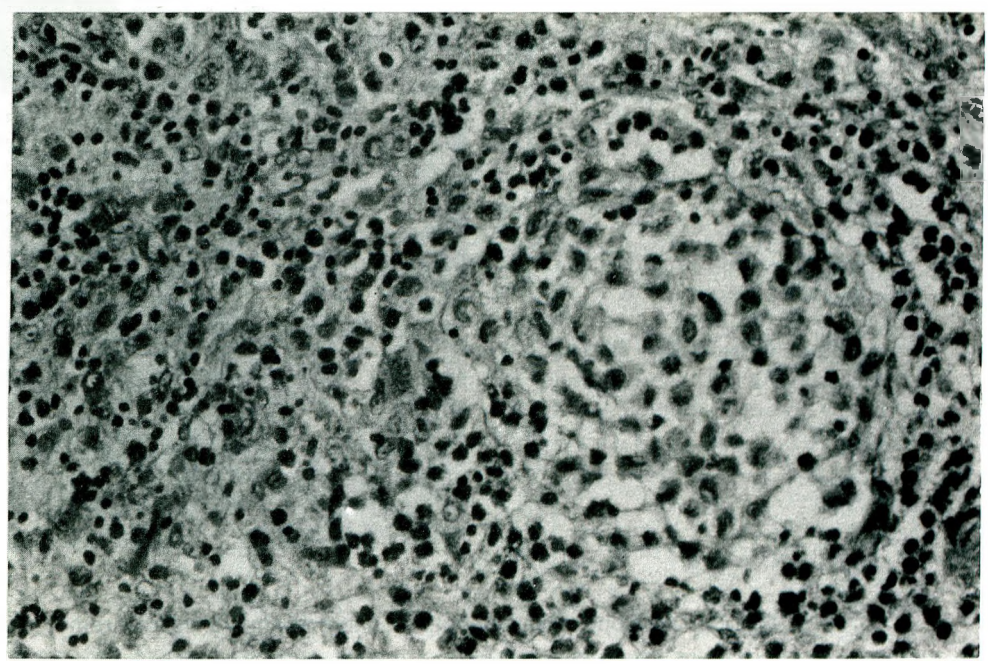

Fig. 2:
Section shows infiltrate composed of Langerhan's cells, histiocytes and lymphocytes. Nuclear indentations and clefts can be better appreciated in the aggregate of Langerhan's cells

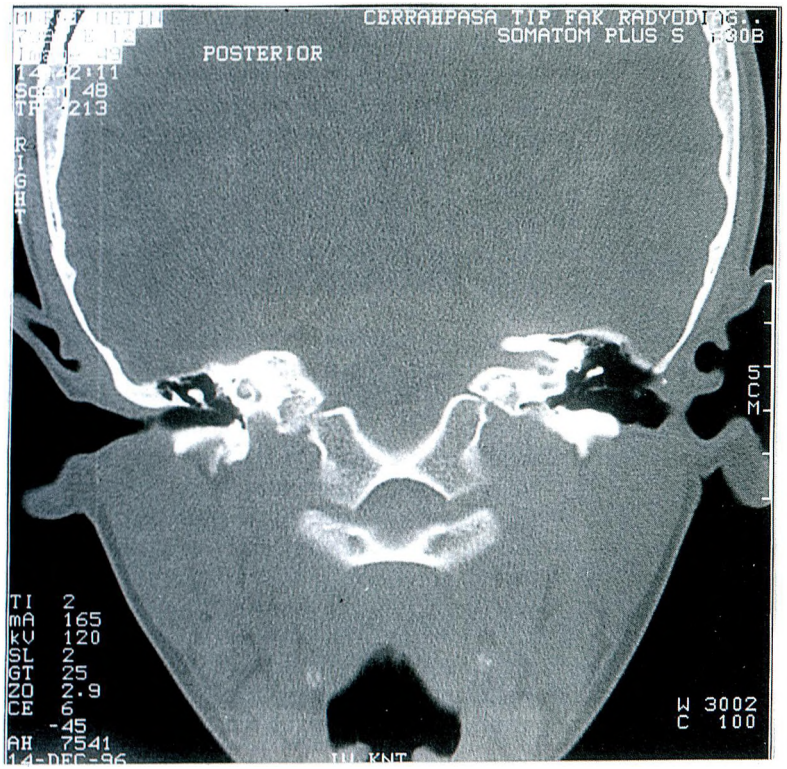

Fig. 3 A - Coronal

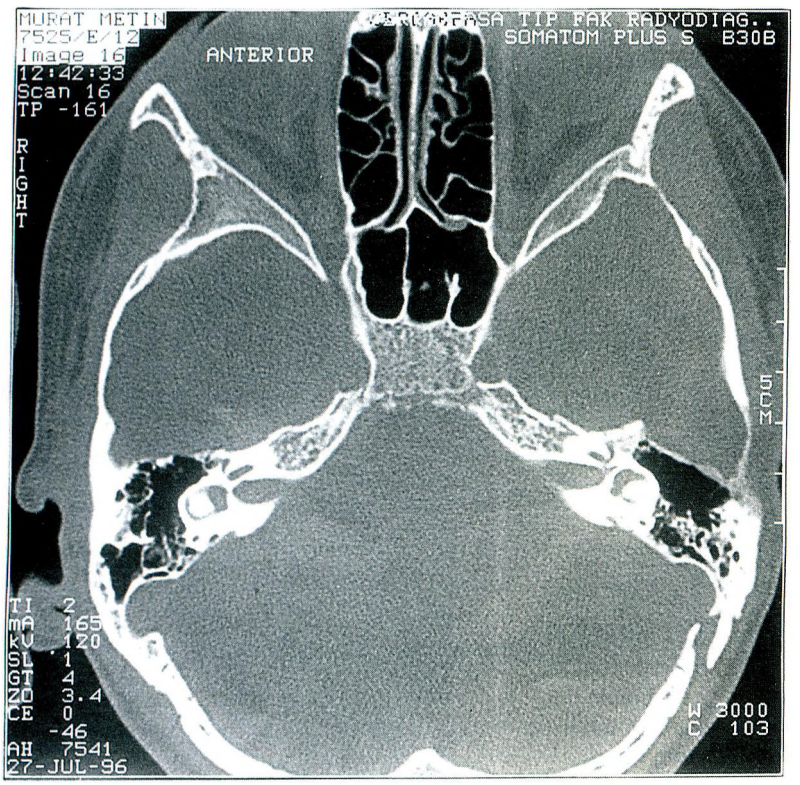

Fig. 3 B - Axial

Fig. 3: Postoperative computerized tomogram shows total removal of the mass

"punched out" appearance are seen. Computed tomography gives a better delineation of both osseous and soft tissue components of the lesion and shows margins of the lesion, erosion of the bony labyrinth and ossicles. There are many disorders causing lytic lesion in the temporal bone, such as cholesteatoma, mastoiditis, malignant tumors including sarcomas and lymphomas, metastatic tumors, hemangiomas. An expansing soft tissue mass is unlikely to be present in mastoiditis. Cholesteatoma usually causes lesions with a sharp border but rarely causes expansion. Hemangiomas show increased contrast enhancement. Metastatic tumors almost exclusively occurs in adults.

Other than radiologic findings there are no distinctive laboratory features of otologic form of the disease. In a study it was reported that elevated erythrocyte sedimentation rate in the absence of acute infection, should raise the suspicion of LCH in children under 3 years of age (11).

In most of the cases, the initial diagnosis is cholesteatoma which is similar and much more common than eosinophilic granuloma. Eventual diagnosis of temporal bone eosinophilic granuloma is done by histologic examination of a biopsy specimen which is obtained by either transcanal or transmastoid approach. Immunohistochemical and electron microscopic examination of the specimen gives the definitive diagnosis (12). A superficial biopsy, usually done transcanally may give only nondiagnostic granulation tissue suggesting chronic inflammation as in our case. Surgical exploration and biopsy is the most appropriate action for diagnostic sampling.

Treatment of LCH is mainly determined by the presence of systemic disease or not. Treatment modalities for unifocal temporal bone eosinophilic granuloma include surgical resection, radiotherapy or combination of the two (5,7). The greatest risk of surgical resection in such cases is facial nerve injury (3,13). Lesions limited to the mastoid cortex and middle ear can be treated by modified radical mastoidectomy but residual disease warrants postoperative radiotherapy (3). When disseminated disease is present or there is recurrence after radiation therapy, chemotherapy is usually indicated (7). Doses ranging from 600 to 1000 cGy are recommended (14). Cure rates for unifocal LCH are greater than 90% regardless of the therapy chosen (15).

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