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T cell lymphoma of the ear presenting as mastoiditis

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Abstract
Mastoiditis is a complication of otitis media characterized by suppuration and destruction of air cell septa in the mastoid and petrous pyramid. Diagnosis is made by clinical findings and computerized tomography (CT) of the temporal bone. We present a patient initially diagnosed by CT as having chronic mastoiditis who was subsequently shown to have an unusual large-cell malignant lymphoma of T cell type.

Key words: Lymphoma, large cell; Lymphoma T cell; Mastoiditis

Case report

A 16-year-old male presented with a two-week history of pain in the skull near the left ear and left peripheral seventh cranial nerve paralysis of one day's duration. Three weeks prior to his admission he complained of infection of the left ear, which was treated by cephalosporins without improvement. In the past, head trauma without loss of consciousness had been recorded. Physical examination revealed tenderness of the left mastoid, the external ear canal narrowed by oedema, a normal tympanic membrane, and left seventh cranial nerve palsy. Blood counts and chemistry were not impaired. Computerized tomography (CT) of the temporal bone showed opacification of the left mastoid characteristic of a chronic process. Paracentesis showed no purulent exudate; therefore, the patient was treated by steroids as in an idiopathic facial nerve palsy.

Minor improvement of the nerve palsy was recorded after two weeks of steroid treatment, but the patient's general condition deteriorated and he was readmitted to the neurological department with diplopia, bilateral papilloedema, sixth cranial nerve palsy, ipsilateral peripheral seventh cranial nerve paralysis, and a scalp lesion. A repeated CT scan revealed a picture which was interpreted as mastoiditis with posterior cortex destruction, lateral erosions (Figure 1) and inflammatory infiltration of tissues lateral to the left mastoid (Figure 2). A mastoid fracture was also suspected, and a space-occupying lesion of the falx was observed. The intracranial hypertension was treated by acetazolamide. On mastoidectomy, extremely thick cutaneous and subcutaneous tissues were found. The cortex and mastoid were filled with white granulation tissue and bone sequestrum.

Histological examination of the multiple biopsies taken from the mastoid showed a few bone fragments embedded in or near areas of fibrotic tissue (Figure 3). All the specimens had severe crush artefacts, but the impression was that in certain areas there were foci of large cells (Figure 4). The lymphoid cells (although crushed) stained positive by immunoperoxidase stainings for CD45 (leucocyte common antigen) and for T cell markers (CD43, CD45DO, CD3) and were negative for B cell markers (CD20, 4KB5); in addition, there were negative results for epithelial, neural, and muscle cell markers. CD30 (Ki-1) was also negative.

All the above findings were very suggestive of a T cell malignant lymphoma, and an additional biopsy was recommended.

The skin biopsy taken later from the scalp lesion revealed a dense, diffuse dermal infiltration made of large lymphoid cells. The cells had large round nuclei with central nucleoli. There were also a few necrotic foci. The cells were positive for lymphoid and T cell markers, and negative for B cell and histiocytic cell markers. CD30 was also negative.

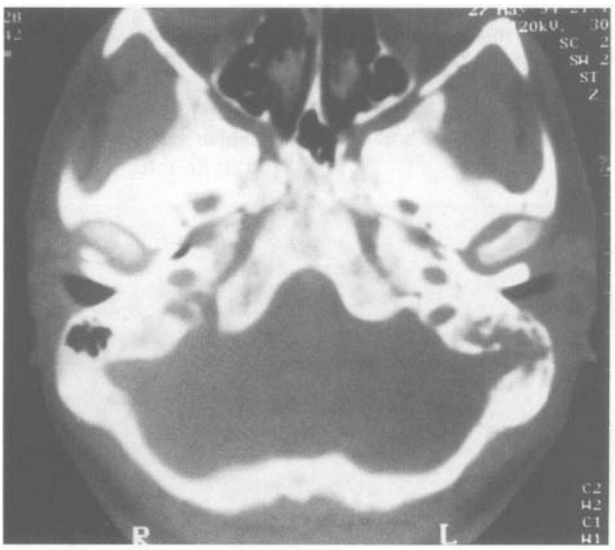

FIG. 1
Opacification of mastoid air cells and destruction of septa of mastoid medial wall near sigmoid sinus.

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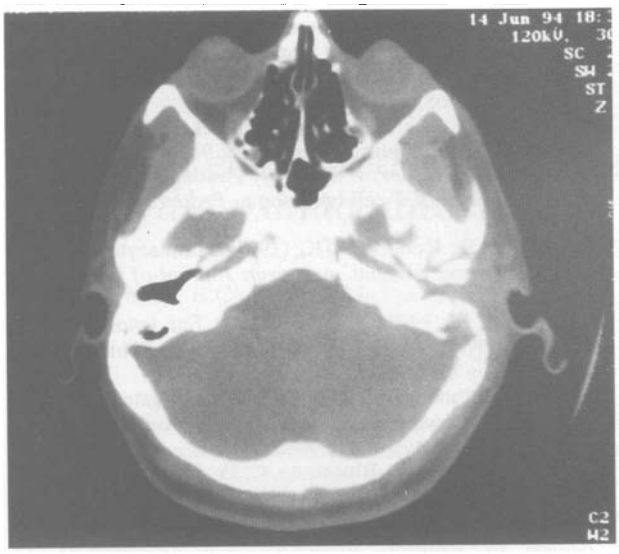

FIG. 2
Soft tissue infiltration on the mastoid and base of auricle.

The diagnosis of the skin lesion as well as of the mastoid lesion was malignant lymphoma, large cell, immunoblastic T type (CD30-negative).

Following the histopathological diagnosis, the patient was investigated and treated in the paediatric oncology unit. Left cervical lymph nodes were moderately enlarged. Brain CT scan revealed a spreading tumour with petrous bone destruction. Lung and abdomen CT scans were normal. A gallium scan showed diffuse pathological bone uptake. Bone biopsy showed only fibrosis. Bone marrow aspiration was normal, but cerebrospinal fluid showed blast cells. The patient was clinically staged as Stage IV diffuse T cell large-cell lymphoma with bone and CNS involvement.

Six courses of inductive chemotherapy protocol (POG 8617) comprising cytarabine, methotrexate, adriamycin, cyclophosphamide and vincristine together with intrathecal injections of methotrexate, hydrocortisone and cytosar were begun immediately. Radiotherapy to the scalp and neck was also applied during the chemotherapy (3500 cGy). After the fourth course, a minor clinical improvement in the main tumour and lymph nodes was observed, while the patient's general health status was deteriorating. Since five months of chemotherapy failed to slow the progress of the disease, second-line therapy

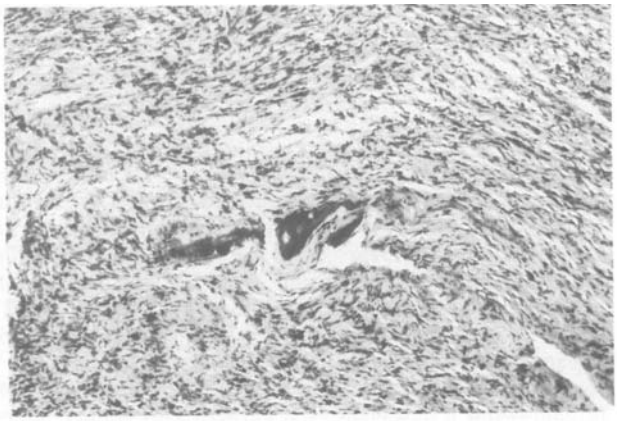

FIG. 3
Low-power photograph of mastoid lesion showing a bone fragment in the centre surrounded by fibrous tissue infiltrated by crushed lymphoid cells (H & E; × 125).

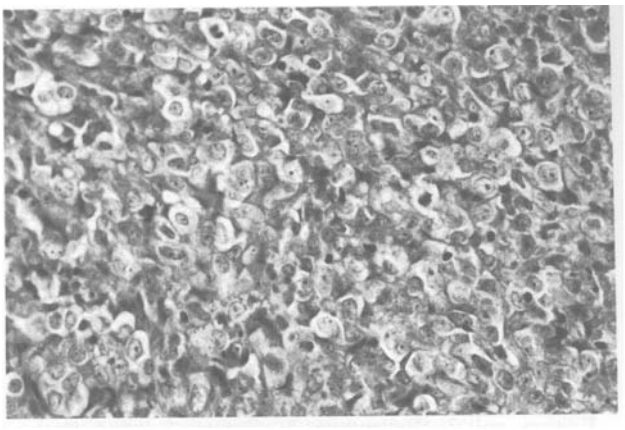

FIG. 4
High-power photograph of skin infiltration, which is made of large lymphoid cells (H & E; × 250).

protocol (dexamethasone, etoposide, ifosfamide, platinol) was started after restaging. Like the first line of chemotherapy, the first course of the second-line chemotherapy did not improve the patient's clinical status, while blast cells appeared in peripheral blood (leukaemia).

The child died of lymphoma. No remission was obtained during the chemotherapeutic treatment.

Discussion

Childhood T cell lymphomas are referred to as both lymphoblastic and aggressive high-grade lymphomas. They classically present with an anterior mediastinal mass with or without peripheral lymphadenopathy, although all combinations of nodal and extranodal sites are possible. These tumours occur predominantly in older children and young adolescents with a high male-to-female ratio. Primary central nervous systems (CNS) lymphoma without systemic involvement is quite rare, comprising 0.3 to 2.3 per cent of all intracranial neoplasms (Jellinger *et al.*, 1975; Spaun *et al.*, 1985) and approximately 2 per cent of all lymphomas (Woodman *et al.*, 1985). It occurs mostly in organ transplant patients or in patients with primary immunodeficiencies, and most of these malignancies are of B cell origin (Spaun *et al.*, 1985; Bogdahn *et al.*, 1986).

Most malignant neoplasms of the middle ear and mastoid are epithelial (Boland and Paterson, 1955; Fairman, 1971). Plasmacytomas of the mastoid bone have recently been reported (George *et al.*, 1994; Kandiloros *et al.*, 1994; Panosian and Roberts, 1994), while malignant lymphomas of the middle ear are more common (Paparella and el-Fiky, 1972; Gapany-Gapanavicius *et al.*, 1980; Takahara *et al.*, 1986). T cell lymphomas of the ear, nose and throat are rare (Norwood and Haller, 1989). The T lymphoblastic lymphoma, derived from the thymic lymphocytes, makes a distinct histopathological and clinical entity (Nathwani *et al.*, 1976). To our knowledge, T cell lymphoma of the mastoid is very unusual.

The most effective treatment regimens for non-Hodgkin's lymphoma, B-cell type, stage IV, are then derived from regimens initially designed for the treatment of leukaemia (Magrath, 1988). All of the modern protocols share two features: very intensive therapy given early in the treatment and use of multiple agents.

In conclusion, T cell lymphoma, large cell, presenting as mastoiditis with seventh cranial nerve paralysis is an unusual mastoid tumour. Lesions involving the mastoid may mimic a chronic infection. When suspected mastoiditis does not respond to standard therapy, unusual T cell

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lymphoma should also be suspected. The use of lymphocytic markers is essential in the histological diagnosis of these cases.

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