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Primary T-cell lymphoblastic lymphoma in the middle ear

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ABSTRACT

T-cell lymphoblastic lymphoma (T-LBL) is a highly aggressive lymphoma characterized by precursor T-cell malignancy and lymphadenopathy or mediastinal involvement. We present the case of an 11-year-old boy with a diagnosis of middle ear T-LBL, which manifested as a headache, hearing loss and peripheral facial paralysis. The child was given intensive chemotherapy and had a complete response. To our knowledge, this is the first case reported in the literature of T-LBL originating in the middle ear. This case aims to help clinicians to be vigilant about the possibility of primary lesions at atypical sites in some special diseases.

1. Introduction

T-cell lymphoblastic lymphoma (T-LBL), a neoplasm of precursor lymphoid cells in the T-cell lineage, is the second most frequent subtype of childhood non-Hodgkin lymphoma (NHL). Most frequently, T-LBL is categorized as a precursor T cell malignancy and is accompanied by lymphadenopathy or mediastinal involvement; by contrast, a middle ear presentation is extremely rare. Here, we describe a rare manifestation of T-LBL presenting with the following clinically significant involvement of the middle ear: headache, hearing loss and mild facial weakness, at initial presentation. To the best of our knowledge, this is the first case of pediatric T-LBL that primarily manifested as a mass in the middle ear.

2. Case report

A previously healthy 11-year-old boy sought medical attention due to a three-week history of headache along with right-sided hearing impairment and right-sided facial palsy (House-Brackmann grade II). He denied fever, vertigo and emesis. Meanwhile, computed tomography (CT) and magnetic resonance imaging (MRI) of the temporal bone revealed extensive soft tissue density in the right middle ear cavity without an intracranial extension (Fig. 1A.1). Based on the clinical presentation and radiological findings, congenital cholesteatoma of the middle ear was originally considered. Therefore, he received a modified radical mastoidectomy in a local hospital after finishing the routine preparation for surgery. Two days after the operation, he complained of a persistent fever (38–39.7 °C) and progressive headache and swelling of the right-sided face. Antibiotics were immediately given for one week, but the symptoms were not successfully managed.

Finally, he was referred to our hospital (without the pathological result). According to the history of surgery and the symptoms, a preliminary diagnosis of a postoperative intracranial infection was considered. Thus, the packing was removed from the external ear canal, where a few granulation tissues were found. In addition, he was treated with intravenous cefatriaxone for seven days for a presumed "infection" to no response. Meanwhile, a lumbar puncture was performed but showed no evidence of an intracranial infection. During the anti-infective therapy, we found a soft mass instead of the granulation tissue that we had previously found, and the mass progressively filled his external auditory canal without evidence of lymphadenopathy or abdominal organomegaly. Enhanced computer tomography located a 3.7 × 3.6 cm, significantly enhanced mass occupying his external auditory canal and tympanic cavity (Fig. 1A.2). The chest radiograph was normal, and tests for HIV were negative; other laboratory examinations were also normal, including complete blood counts, electrolytes, liver function, and renal function. Considering the rapid growth of the soft tissue, we doubted whether it was a malignant mass and its suitability for surgical resection; therefore, a mass biopsy was performed and revealed diffuse small to medium sized lymphocytes that were inclined to a lymphoma (Fig. 2B.1). Immunohistochemistry stains showed that the T cells were positive for CD3ε (Fig. 2B.2), CD4, CD43, CD99, LCA and GB, with a partial co-expression of CD56. The Ki-67 proliferation rate was 80%. Meanwhile, the neoplastic cells were negative for CD20, CD8,

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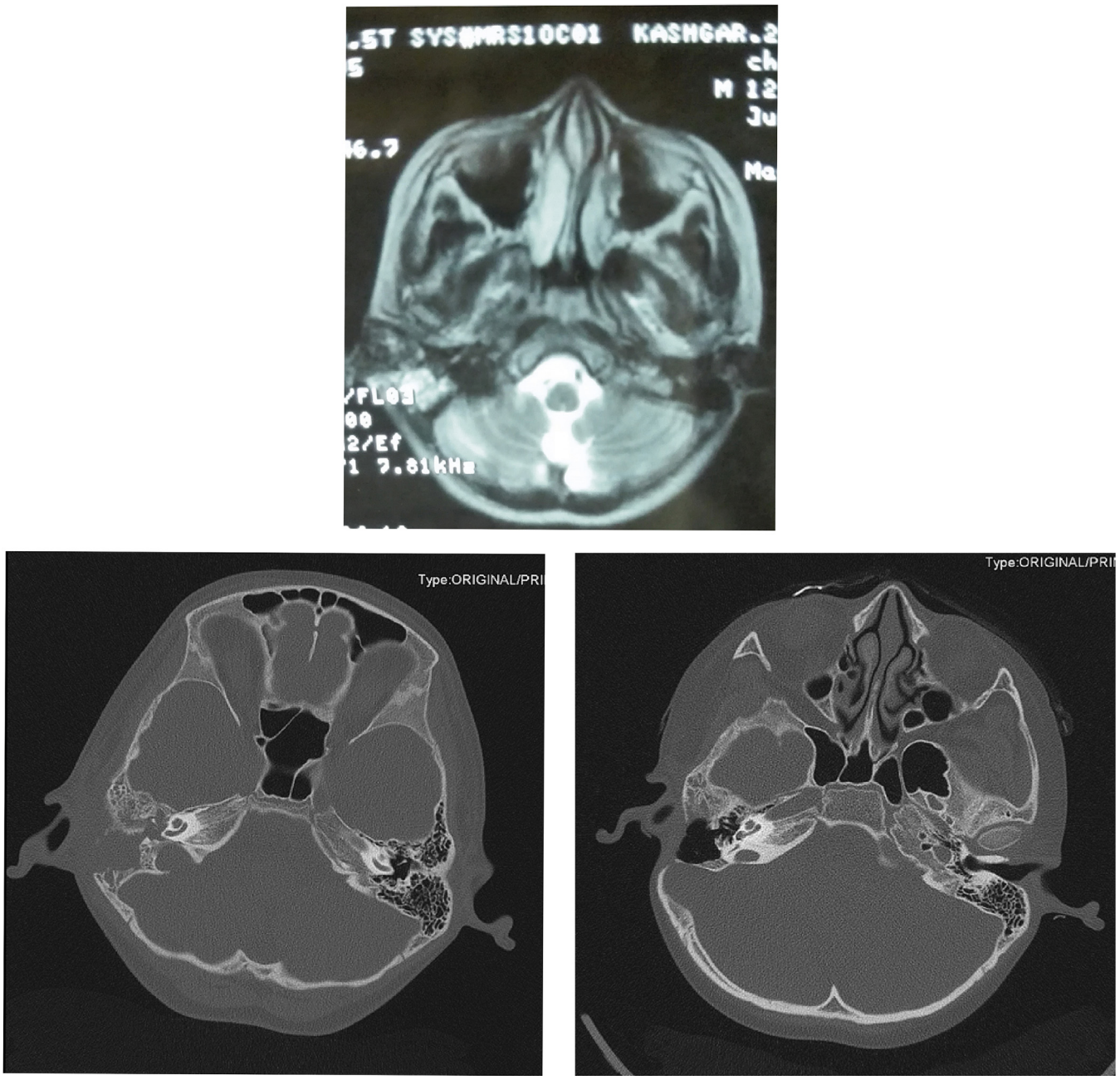

Fig. 1. (A1) MRI illustrating an extensive mass filling the right middle ear cavity. (A2) Temporal high resolution computerized tomography (HRCT) demonstrating soft tissue filling the external ear canal and middle ear. (A3) Temporal HRCT in remission.

PCK, desmin, myogenin, MPO, CK7, P63, S-100, and EBER. Despite a negative expression of Terminal deoxynucleotidyl transferase (TdT), identical clonal rearrangements of the gene for T-cell receptor γ (TCRγ) in samples were recorded. Moreover, bone marrow (BM) aspirates and biopsy showed no BM involved. These findings were strongly consistent with T-LBL.

Subsequently, the boy was scheduled to start on an intensive chemotherapy regimen that was based on protocols of the Berlin–Frankfurt–Munster (BFM) cooperative group that consists of vincristine, daunorubin, L-asparaginase, steroid and intrathecal methotrexate. On Day 5, his symptoms were quickly managed, including headache, facial palsy and swelling of his right face. Moreover, his parents noticed the neoplasm abscission in his external ear canal on Day 7. The patient became symptom free after the first cycle of chemotherapy. Tolerating the regimen well, the patient received a BM biopsy three months later that still showed no involvement. At the same time, he was given a hearing test that showed normally. The tumor also showed complete response using a follow-up positron emission tomography (PET/CT) scan. After receiving nine cycles of chemotherapy, he achieved complete remission (Fig. 1A.3) and is still in a maintenance phase.

3. Discussion

A diagnosis of chronic infectious ear disease could be first considered according to the following symptoms: hearing loss, otalgia, otorrhea, and even facial palsy. When these same presentations are attributed to a tumor, considerable time may elapse before an accurate diagnosis is made. For the patient who just received a mastoidectomy, continuous fever and headache always point to a postoperative intracranial infection. We first administered antibiotic therapy to the patient with no response.