Temporal bone metastasis as a sign of relapsing chronic lymphocytic leukemia

Hadeel M. Aljafar, MD, Sari S. Alsuhibani, MD, Mohammad S. Alahmari, MD, Musaed A. Alzahrani, MD.

From the Division of Otorhinolaryngology, Department of Surgery (Aljafar, Alahmari, Alzahrani), King Fahd Specialist Hospital, and the Division of Otorhinolaryngology, Department of Surgery (Aljafar), Department of Radiology (Alsuhibani), King Fahd Hospital of the University, University of Dammam, Dammam, Kingdom of Saudi Arabia.

Received 10th May 2015. Accepted 19th August 2015.

Address correspondence and reprint request to: Dr. Musaed A. Alzahrani, Division of Otorhinolaryngology, Department of Surgery, King Fahd Specialist Hospital, Amer Bin Thabet Street, PO Box 15215, Dammam 31444, Kingdom of Saudi Arabia. E-mail: musaed.zahrani@kfsh.med.sa

The incidence of otologic complains in leukemia patients ranges from 16.8%-48%. Hearing loss, conductive, or sensorineural, is the most common symptom. In addition, patients may complain of tinnitus, vertigo, otalgia, otorrhea, and otorhagia. Although, otologic manifestations are very common in those patients, leukemic infiltration of the temporal bone is very rare, and only few cases are reported in the literature. We report a case of relapsing chronic lymphocytic leukemia (CLL) presenting with otitis media and lower motor neuron facial paralysis. Our objective in presenting this particular case is to highlight the rare temporal bone complications and management in CLL.

Case Report. A 65-year-old diabetic patient was admitted to the hospital with suspicion of skull base osteomyelitis. He presented with bilateral persistent otitis media for one month not responding to antibiotic therapy with severe otalgia. Past medical history revealed CLL 15 years ago. He was in complete remission for 8 years before having a recurrence presented as...
Bone metastasis in relapsing CLL ... Aljafar et al

A lymphoproliferative malignancy (Figure 2). Bone marrow aspirate confirmed the diagnosis of a relapsing CLL. Cytogenetic chromosomal analysis revealed 71% trisomy 12 (which is the most common cytogenetic change in CLL, associated with an intermediate prognosis), and 13% monosomy p53. He was started on Ibrutinib, a long-term daily oral chemotherapy that resulted in complete remission. Facial nerve paralysis was completely recovered with a residual TM perforation on the left ear. Granulation tissue of the middle ear disappeared completely at one month follow up after treatment.

Discussion. An elderly diabetic patient presenting with otalgia and facial nerve paralysis has most probably skull base osteomyelitis. However, this present study shows that those findings can be signs of relapsing CLL. Chronic lymphocytic leukemia is characterized by proliferation of (mono)clonal mature B-lymphocytes in peripheral blood, bone marrow, and lymphoid tissues with a characteristic immunophenotype. In a study of the frequency of genetic abnormalities in leukemic B-cells, 111 out of 180 CLL cases (62%) displayed one or more cytogenetic abnormalities. In the whole series, del(13q-) was the most frequently observed abnormality (35%) followed by trisomy 12 (23%), 11q- (9.4%), and 17p- (8%). Peripheral facial palsy (PFP) is most commonly caused by a benign self-limiting inflammatory neuropathy condition, known as Bell’s palsy. However, systemic diseases can present with PFP including Guillain-Barré syndrome, sarcoidosis, syphilis, leukemia, Lyme disease, meningitis, and acute HIV infection.

Figure 1 - Coronal (A) and Axial (B) CT scan cuts of the temporal bones showing homogenous opacity of the mastoid and middle ear cavity. Dehiscent fallopian canal of the facial nerve is visible on the left side (arrow).

Figure 2 - Fragment of middle ear granulation tissue infiltrated by monotonous small lymphoid cells showing round nuclei, condensed chromatin, inconspicuous nucleoli, and scanty cytoplasm (arrow).
Temporal bone metastasis from distant primary carcinoma can mimic any local inflammatory or infectious process. Metastasis can reach the temporal bone through any of these patterns: a) hematogenous spread; b) tumor cells gaining access to the cerebrospinal fluid and disseminating through the subarachnoid space into the internal auditory canal; c) direct extension; d) leptomeningeal extension from an intracranial primary tumor; and e) lymphomatous infiltration.7 Druss8 in 1945, was the first otologist to call attention to the fact that patients with leukemia can present with secondary otological complications.

Buyukavci et al9 reported that bilateral PFP is a warning sign of leukocyte infiltration. The CLL can present with otologic symptoms in up to half of the affected persons, such as acute otitis media and acute mastoiditis.10 However, it is important to keep in mind the possibility of temporal bone metastasis of CLL with unusual presentations, including facial paralysis and sensorineural hearing loss.

In conclusion, unusual otologic manifestations in patients with past history of CLL should alert us to the possibility of temporal bone metastasis. Full history and physical examination is mandatory in addition to proper imaging and histological exams.

Acknowledgment. We thank our colleagues Dr. Heba N. Raslan, MD, Consultant Hematopathology, and Dr Ahmed Alsaghier, MD, Consultant Hemato-Oncology, King Fahd Specialist Hospital, Dammam, Kingdom of Saudi Arabia for their insight and expertise that greatly assisted this case report.

References


Related Articles

