

Self-assessment corner

Hearing loss and leukocytosis

Izidore S Lossos, Shimon Berger, Moshe Gomori, Gil Leibovich, Yaacov Matzner

A 79-year-old woman noticed progressive hearing loss and a sensation of pressure in her right ear. Otolaryngologic examination was unremarkable. During the next three months she developed malaise, diaphoresis and intermittent low-grade fever. Physical examination revealed mild bilateral cervical lymphadenopathy and hepatosplenomegaly. Laboratory tests revealed leukocytosis of $120 \times 10^9/l$ without anemia or thrombocytopenia. Serum protein and immune electrophoresis were normal. A peripheral blood film revealed the presence of smudge cells and 90% small mature lymphocytes, which were CD2, CD5, CD19, and CD20 positive. An audiogram performed in 2/92 and 8/92 and magnetic resonance imaging (MRI) of the auditory apparatus, demonstrated sensorineural hearing loss, more prominent in the right ear (figures 1 and 2, respectively). Treatment was initiated and a repeat audiogram was performed in 12/92 (figure 1).

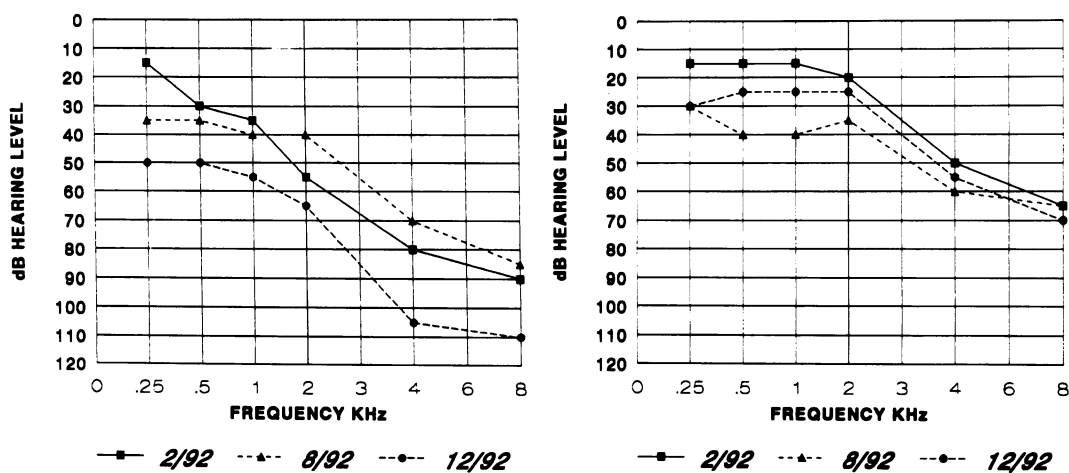


Figure 1 Audiogram results before and after treatment. Left = left ear, right = right ear

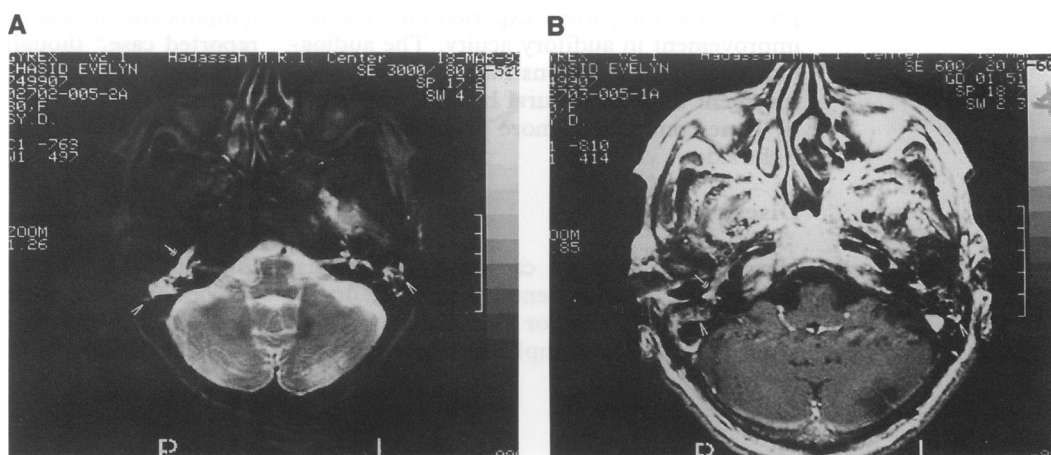


Figure 2 (A) Axial T₂-weighted image through the petrous bone. (B) Axial enhanced T₁-weighted image at the same level

Questions

- 1 What abnormality is demonstrated on the audiograms from 2/92 and 8/92?
- 2 What abnormalities are shown on the MRI?
- 3 What is the patient's underlying disease?
- 4 How does the underlying disease cause hearing loss?
- 5 Describe the changes observed on the audiograms following specific therapy.

Hadassah University Hospital, Jerusalem, Israel
 Department of Internal Medicine
 IS Lossos
 G Leibovich
 Department of Otolaryngology/Head and Neck Surgery
 S Berger
 Department of Radiology
 M Gomori
 Haematology Unit
 Y Matzner

Correspondence to
 Yaacov Matzner, MD,
 Haematology Unit, Hadassah University Hospital Mount Scopus, Jerusalem, Israel

Accepted 1 June 1995

Answers

QUESTION 1

The audiograms performed in February and August 1992 demonstrate sensorineural hearing loss in the left ear, and mixed (conductive and sensorineural) hearing loss in the right ear.

QUESTION 2

Figure 2A shows the axial T₂-weighted image through the petrous bone, demonstrating a high-intensity eustachian tissue in the right tube (double arrowhead) and the hypotympanium (single arrowhead) with similar tissue in the right middle ear and mastoid region.

Figure 2B is an axial enhanced T₁-weighted image at the same level and demonstrates enhancement of all the areas that were high intensity on the T₂-weighted image. These findings are consistent with a solid tumour.

QUESTION 3

The patient has a classic B-cell chronic lymphocytic leukaemia with marked lymphocytosis, lymphadenopathy, hepatosplenomegaly and immunophenotyping. Bone marrow confirmed the diagnosis. Since the patient does not have anaemia and thrombocytopenia, she is in stage 2 according to Rai.¹

QUESTION 4

In the present case, the hearing loss was caused by leukaemic infiltration of the auditory apparatus, as demonstrated in figures 2A and 2B. However, chronic lymphocytic leukaemia may cause hearing loss by a different mechanism, in leukostasis, due to high leukocyte counts (hyperleukocytosis; see Discussion).

QUESTION 5

Following institution of chlorambucil and prednisone, the patient experienced a marked improvement in auditory acuity. The audiogram from 12/92 demonstrates a partial improvement in sensorineural hearing loss tested by bone conduction, more prominent on the right side.

Discussion

Hearing loss may be caused by infections, genetic and developmental disorders, drugs, vasculitis and primary or metastatic neoplastic lesions.² Otological complications of leukaemia

are infrequently described and leukaemia is a rare cause of deafness. Among the various types of leukaemia, acute lymphoblastic leukaemia is the most common form affecting hearing. Paparella *et al*³ described otological complications in 46% of 25 patients with leukaemia. The complications included otitis media, mastoiditis, haemotympanum, sensorineural hearing loss, and vertigo. They were attributed to leukaemic infiltration, haemorrhage or infection. None of their patients suffered from chronic lymphocytic leukaemia. To our knowledge, only five other cases of hearing loss attributed to chronic lymphocytic leukaemia have been published in the English literature.⁴⁻⁶ The relevant clinical information from the previously reported cases and the present case are summarised in the table.

Hearing loss in chronic lymphocytic leukaemia can be caused by a leukaemic infiltration of the auditory apparatus or can be a manifestation of leukostasis. Since hyperleukocytosis is uncommon in chronic lymphocytic leukaemia, hearing loss as a manifestation of leukostasis is rare. There are several reasons for this: *a*) the small size of the lymphocyte requires very high lymphocyte counts to produce leukostasis; *b*) the rheologic properties of lymphocytes, in comparison with myeloid cells, are minimal; *c*) there is an inverse correlation between haematocrit and leukocrit, keeping total cytocris low. Consequently, all the four reported cases of deafness caused by leukostasis due to chronic lymphocytic leukaemia had leukocyte counts above $500 \times 10^9/l$ and manifested additional neurological signs and symptoms. Deafness was not the presenting manifestation of the disease in these patients.

It was speculated that leukaemic infiltration of the auditory apparatus was involved in the pathogenesis of hearing loss due to chronic lymphocytic leukaemia in a single recently reported case,⁶ though neither radiologic nor pathologic confirmation of this was provided. In the present case, infiltration of the middle ear and mastoid cell was clearly demonstrated by computed tomography (CT) and MRI. In both cases, sensorineural hearing loss was a presenting manifestation, although in neither was the leukocyte count sufficiently high to implicate leukostasis in the pathogenesis. Both responded well to chemotherapy with recovery of auditory acuity. In contrast, chemotherapy and radiotherapy failed in a single patient with

Table Hearing loss due to chronic lymphocytic leukaemia – summary of reported cases

Ref	Year	Age/sex	Hearing loss at presentation	WBC* ($10^9/l$)	Audiogram	Pathogenesis	Treatment	Recovery of auditory acuity
4	1978	70/F	no	647	NR	leukostasis	leukapheresis	yes
		83/F	no	1000	NR	leukostasis	leukapheresis	no
5	1985	42/M	no	968	NR	leukostasis	leukapheresis	yes
		40/F	no	>500	NR	leukostasis	radiation and chemotherapy	no
6 Present case	1993	35/M	yes	30	sensorineural	not confirmed	chemotherapy	yes
		79/F	yes	120	sensorineural	leukaemic infiltration	chemotherapy	yes

*White blood cell count at the time of appearance of hearing loss. NR – not reported

leukostasis-induced deafness,⁵ while leukapheresis improved auditory acuity in two of three such patients.^{4,5}

Physicians should be aware of hearing loss as a rare presenting manifestation or complication of leukaemia, including chronic lymphocytic leukaemia. Since hearing loss can progress rapidly to permanent deafness, early diagnosis and prompt treatment are essential. Definite determination of the mechanism of hearing loss (leukostasis versus leukaemic infiltration) is important in providing appropriate therapy (leukapheresis or chemoradiotherapy, respec-

tively). Imaging techniques, including CT and MRI, may help in demonstration of leukaemic infiltration of the auditory apparatus, thus providing the pathogenesis of the hearing loss.

Final diagnosis

Chronic lymphocytic leukaemia resulting in hearing loss

Keywords: chronic lymphocytic leukaemia, MRI, hearing loss

- 1 Rai KR, Sawitsky A, Cronkite EP, *et al.* Clinical staging of chronic lymphocytic leukemia. *Blood* 1975; **46**: 219–34.
- 2 Nadol JB. Hearing loss. *N Engl J Med* 1993; **329**: 1092–102.
- 3 Paparella MM, Berlinger NT, Oda M, El Fiky F. Otological manifestations of leukemia. *Laryngoscope* 1973; **83**: 1510–26.
- 4 Preston FE, Sokol RJ, Lilleyman JS, Winfield DA, Blackburn EK. Cellular hyperviscosity as a cause of neurological symptoms in leukemia. *BMJ* 1980; **1**: 476–8.

- 5 Baer MR, Stein RS, Dessypris EM. Chronic lymphocytic leukemia with hyperleukocytosis. The hyperviscosity syndrome. *Cancer* 1985; **56**: 2865–9.
- 6 Nageris B, Or R, Hardan I, Polliack A. Sudden onset deafness as a presenting manifestation of chronic lymphocytic leukemia. *Leukemia Lymphoma* 1993; **9**: 269–71.
- 7 Lichtman MA. Rheology of leukocytes, leukocyte suspensions and blood in leukemia. *J Clin Invest* 1973; **52**: 350–9.