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Case Report

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An Uncommon Presentation of Chronic Lymphocytic Leukemia: Bilateral Hearing Loss

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ABSTRACT

Sudden sensorineural hearing loss that presents as the initial sign of haematological disease is very rare. We report a case with typical Rai Stage 2 (Binet Stage A) CLL, who presented with sudden onset deafness as the initial manifestation. A 48-year-old male presented with sudden bilateral sensorineural hearing loss. The patient was found to have chronic lymphocytic leukemia (CLL) during a work-up for his hearing loss. Early onset of sudden deafness in a CLL patient may be due to the hyperviscosity syndrome. Audiograms were recorded showing positive findings on presentation and recovery after therapy. The importance of early diagnosis and therapy is stressed in the light of this rapid clinical and audiological recovery observed here. *©2007, Firat University, Medical Faculty*

Key words: *Chronic lymphocytic leukemia, hearing loss, hyperviscosity*

ÖZET

Kronik Lenfositik Lösemide Nadir Bir Başvuru Şekli: Bilateral İşitme Kaybı

Ani işitme kaybı hematolojik hastalıklarda nadiren bir başvuru belirtisi olabilir. Burada başvuru nedeni ani işitme kaybı olan Rai evre 2 (Binet evre A) olan kronik lenfositik lösemi (KLL) vakasını sunmaktayız. Kırk sekiz yaşındaki erkek hasta bilateral işitme kaybı ile başvurdu. İşitme kaybının nedeni araştırılırken KLL tespit edildi. Tedavi öncesine göre odiyogramlarında belirgin düzelme elde edildi. Ani işitme kaybı hiperlökositozun yol açtığı lökostaz ile ilişkilendirildi. İşitsel düzelimin ışığında erken tanı ve tedavinin önemi vurgulandı. *©2007, Fırat Üniversitesi, Tıp Fakültesi*

Anahtar kelimeler: *Kronik lenfositik lösemi, işitme kaybı, hiperviskozite*

Chronic lymphocytic leukemia (CLL) is a neoplasm of monocytic small, round (mature appearing) B-lymphocytes in the peripheral blood which usually espress CD5, CD19 and CD23 antigens. At the time of diagnosis all of patients have bone marrow and peripheral blood involvement with a lymhocyte count >10.000/mm3. Most of patients are asymptomatic. Clinical manifestations of hyperleukocytosis are common in acute leukemia (1-3) and less common in chronic leukemia which mainly involve lungs, and the brain, including sensorineural dysfunction of cranial nerves (1,4,5). Deafness has been known to occur during the clinical course of leukemia since the initial descriptions of Vidal (1856) and Politzer (1884). In chronic leukemias three cases with CLL and hearing loss were reported (6,8).

CASE REPORT

A 48-year-old male was admitted sudden hearing loss and tinnitus in both ears. After audiological examination a sensorineural hearing loss was diagnosed which was 67% in the right ear and 69% in the left ear (Figure 1). During the work-up complete blood count revealed significant leucocytosis white blood cell count: 160.000/mm3 and referred to a specialist in Internal Medicine. Physical examination revealed lympadenopathies in cervical region and splenomegaly. Peripheral blood smear revealed mature appearing lymphocytosis (88%) and smudge cells. Bone marrow aspiration was hypercellular with 70% mature appearing lymphocytes. Flow cytometrical examination of peripheral blood cell revealed CD5, CD19, CD23 positive lymphocytosis. Other laboratory parameters are illustrated in table 1.He was diagnosed as having chronic lmphocytic leukemia. Fludarabin was started (25mg/m^2 / day, 5 days) for remission induction. In 3 weeks, when total white blood cell count decreased to 23.000/mm^3, hearing loss resolved clinically and audiogram showed normal pattern (Figure 2). The patient did not experience any new hearing loss attack during six month of follow-up.

Table 1. Laboratory parameters

Leukocyte count/mm^3	160.000
Hemoglobin gr/dL	11.6
Thrombocyte count/mm^3	213.000
Uric acid (mg/dL)	8.9
LDH (U/L)	974

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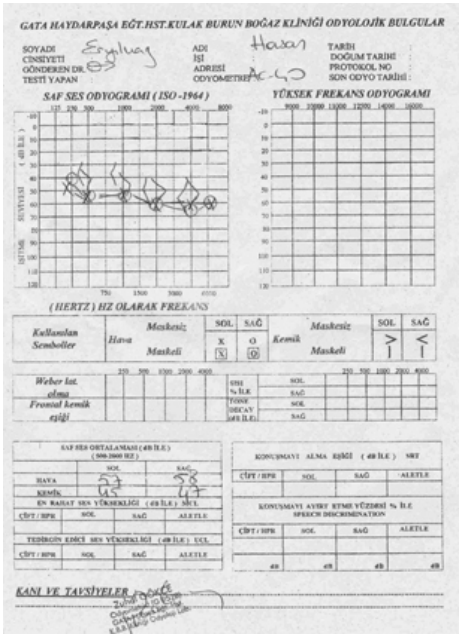

Figure 1. Audiogram before CLL treatment

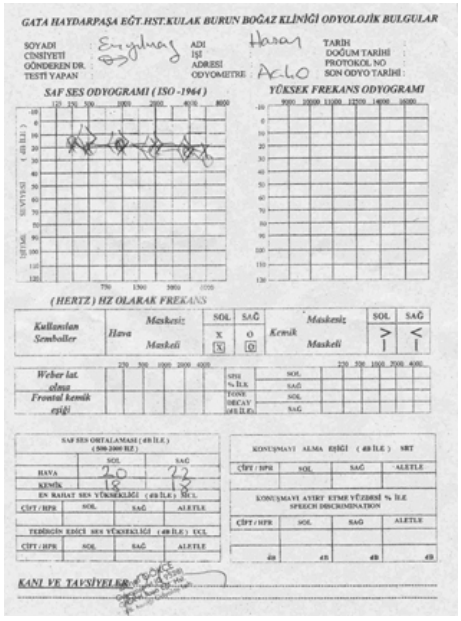

Figure 2. Audiogram after CLL treatment

DISCUSSION

Hearing loss is an uncommon symptom in hematological diseases. There are only few case reports with regarding hearing loss in leukemias. The pathogenesis is not well established. In some cases, postmortem lesions in the inner ear were reported. Nageris et al (6) reported a CLL case with sudden hearing loss in 1993. In that case, the hearing loss was also sensorineural and improved dramatically after chemotherapy. This unique observation was associated with decrease of the circulating CLL cells, which corresponded to a partial response, and lasted three years. Chae et al (9) reported a similar observation in 2002. The hearing loss in a chronic myelogenous leukemia was resolved after leukoapheresis. They presumed that cochlear vessel occlusion, as a result of elevated blood viscosity was responsible for the hearing loss. Harada et al (10) also reported a case with unilateral sensorineural hearing loss in a patient with acute lymphoblastic leukemia. The hearing loss was reversible.

There are also some cases with blindness due to retinal vein occlusion as a result of hyperviscosity syndrome. Serum viscosity can increase with large molecules such as IgM or IgA dimers, which usually occur in plasma cell discrasias. In vivo this can cause sludging of capillary blood flow and so vision disturbances, and some clotting disorders. The pathogenesis of the clinical features of leukemic hyperleukocytosis is complex. Multiple factors may involved including number, size, and deformability proliferation rate; vascular adhesion/invasion ability of leukemic cells, release of tissue harmful substances by different leukocyte types, specific characteristics of the microcirculation and competition of leukocytes and tissue cells for O_2 (1,3,11,12). Hyperviscous blood becomes an important factor if leukocyte is above 15ml/dl, which contributes injury at the microcirculatory level. The mechanisms may change depending on the leukemia type. The increased leucocytes result in small aggregates and/or leukocyte thrombi, that lead to tissue infarctions or vascular and tissue invasion with hemorrhage (1,3). However, in some postmortem series in CLL patients, thrombi and aggregated leukocytes have been seen described (13).

In our case, the bilaterally sensorineural type hearing loss resolved after chemotherapy, within three weeks.

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