Int. Adv. Otol. 2012; 8:(2) 317-320

CASE REPORT

Sensorineural Hearing Loss and Tinnitus as Presenting Symptoms of Polycythemia Vera

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Abstract:

Polycythemia vera (PV) is a blood disorder in which the bone marrow makes too many red blood cells. Commonly encountered neurologic symptoms include transient ischemic attack, dementia, dizziness, tinnitus, and visual disturbances which are thought to result from thrombosis and hemorrhage. Although hearing impairment is one of the neurologic symptoms of PV, tinnitus or hearing impairment as a presenting symptom of PV is very rare. We report a patient with polycythemia vera whose first sign and symptom were bilateral tinnitus and hearing loss. Unfortunately, the hearing was not recovered after treatment including aspirin and phlebotomy while blood level returned to near normal range.

Submitted : 16 June 2011 *Accepted : 7 November 2011*

Introduction

Polycythemia vera (PV) is characterized by an erythrocytosis variably associated with leukocytosis and thrombocytosis. PV is a rare disease with an incidence rate of approximately 1.9-2.3 new cases per 100,000/year in Europe and the United States. The incidence of PV is slightly higher in men than women (2.8 vs. 1.3 cases/100,000/year), and is highest for men aged 70-79 years (24 cases/100,000/year) [1].

The most common presenting complaints result from thromboembolic problems. Clinical manifestations range from nonspecific complaints such as headache, weakness, pruritus, dyspepsia, and dizziness to major thrombotic events such as visual disturbances, stroke, and acute myocardial infarction. Although hearing impairment is one of the neurologic symptoms of PV, tinnitus and hearing impairment as a presenting symptom of PV are very rare. We report a 50-year-old man with PV whose first sign and symptoms were bilateral hearing loss and tinnitus.

Case Report

A 50-year-old man presented with a history of bilateral tinnitus and hearing loss in his right ear. One year ago, hearing loss and tinnitus had developed in his right ear. At that time, he visited another hospital for the hearing loss and was treated with some medication on the impression of idiopathic sudden hearing loss. However, as his hearing was not recovered and his tinnitus was aggravated, he quit taking the medication. After 6 months, tinnitus gradually developed in his left ear and he visited our clinic for management of the newly developed tinnitus. It was not associated with otalgia, otorrhea, or dizziness. There was no history of previous noise exposure, otologic trauma, or drug intake. The patient denied any medical history of systemic disease or severe illness. On physical examination, both external auditory canals and tympanic membranes were normal. There was no nystagmus or facial asymmetry.

The pure tone audiometry demonstrated a deaf ear on the right side and 45 dB of descending high tone sensorineural hearing loss in the left ear (Figure 1). Auditory brainstem response showed no response in the right ear and wave V recordable at 50 dB on the left side (Figure 2). Tinnitogram showed subjectively perceptive pitch of 2~3 kHz with loudness level of 60 dB. Laboratory results showed a platelet count of 833,000/μL (normal range: 140,000~400,000),

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leukocyte count of 11,900/ μL (3,900~9,900), red blood cell count of 5.99×106/μL (4×106~5.6×106), hemoglobin of 18.7 g/dL (12~15), and 55.6% (35~45) of hematocrit.

The patient was referred to the department of hematology and underwent further examination with suspicion of myeloproliferative disease. Erythropoietin was decreased to 6.0 mU/mL (normal range 10.2~25.2). Peripheral blood smear revealed erythrocytosis with normal morphology and leukocytosis with neutrophilia. Bone marrow aspirates showed hypercellularity with generalized hyperplasia. Chromosomal studies revealed the presence of the JAK2 V617F mutation, while major bcr/abl rearrangement was absent. EKG showed complete block of the right bundle branch, while echocardiogram revealed normal appearance and contractility. Abdomen and pelvis CT showed mild splenomegaly and a tiny low density nodule in the left hepatic lobe.

On the basis of peripheral blood, bone marrow aspiration, and chromosomal study results, a diagnosis of polycythemia vera (PV) was made. Phlebotomy was performed 4 times and 100 mg of aspirin was prescribed. After 2 months, laboratory findings improved to a platelet count of 429,000/μL, leukocyte count of 10,080/μL, red blood cell count of 5.26×106/μL, hemoglobin of 16.1 g/dL, and 47.6% of hematocrit. Unfortunately, there was no change in his hearing clinically. A hearing aid for the left ear was recommended.

Discussion

Polycythemia vera is a clonal myeloproliferative disease characterized by erythrocytosis variably associated with leukocytosis and thrombocytosis. PV is a rare disease with an incidence rate of approximately 1.9-2.3 new cases per 100,000/year in Europe and the United States. The incidence of PV is slightly higher in men than women (2.8 vs. 1.3

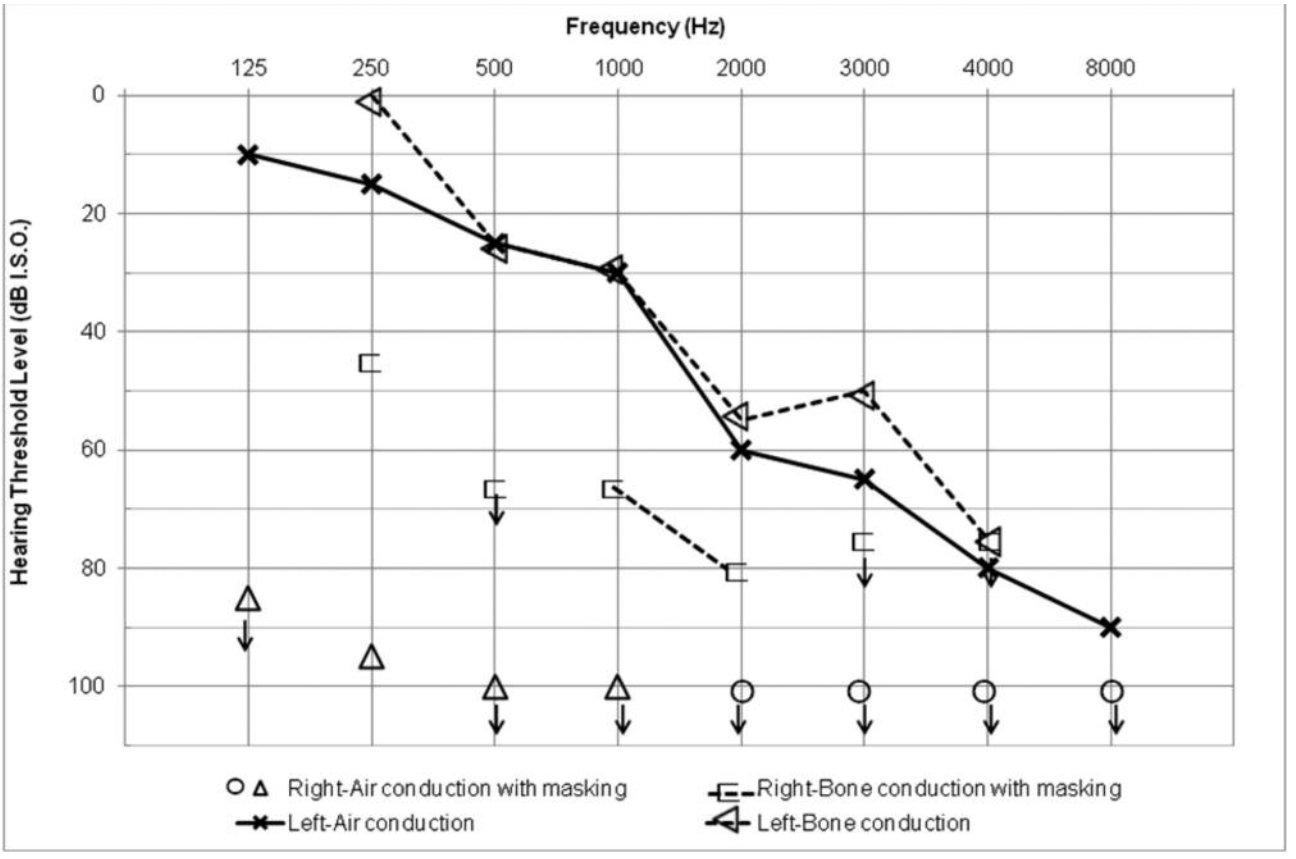

Figure 1. Pure tone audiogram showing deafness in the right ear and descending pattern of sensorineural hearing loss in the left ear.

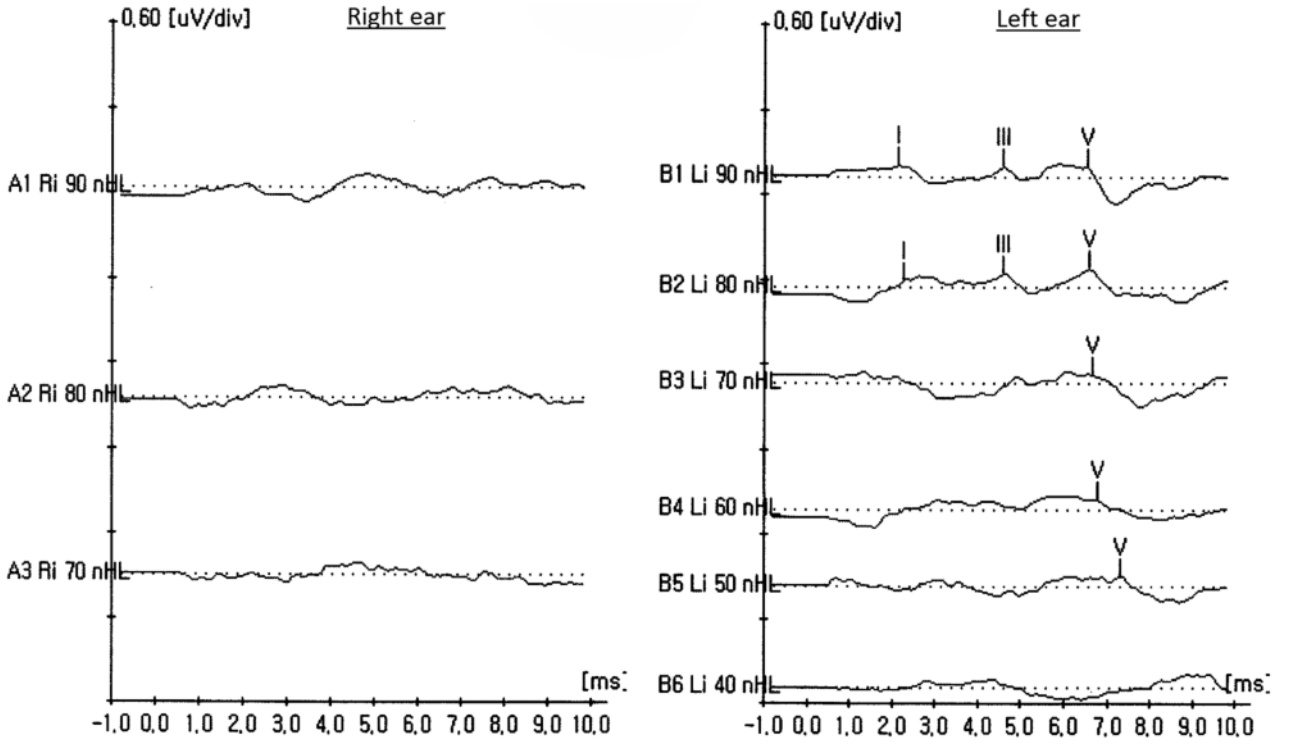

Figure 2. Auditory brainstem evoked response of right ear showed no response by click stimulus at 90 dB nHL. Wave V was recorded at 50 dB nHL in left ear.

cases/100,000/year), and is highest for men aged 70-79 years (24 cases/100,000/year) [1].

It is classified as one of the chronic myeloproliferative disorders, which are chronic diseases originating from the mutation of a single myeloid stem cell. With the mutation, the cell acquires the capacity for autonomous proliferation. This results in a growth advantage over normal cells that leads to the progressive expansion of the mutated clone and to the hyperproduction of one or more mature blood cells [2].

The most common presenting complaints of PV are thrombosis and hemorrhage. Clinical manifestations range from nonspecific complaints such as headache, weakness, pruritus, dyspepsia, and dizziness to major thrombotic events such as visual disturbances, stroke, and acute myocardial infarction. In about one-third of subjects, PV manifests with a major thrombotic event such as stroke or transient ischemic attack, acute myocardial infarction, Budd-Chiari syndrome or pulmonary embolism. Bleeding events include epistaxis, oral mucosal hemorrhage, gastrointestinal hemorrhage, or nonspecific ecchymosis.

Otological symptoms and signs such as vertigo, tinnitus, hearing loss, facial weakness, and otitis media are not uncommon findings in hematological diseases including polycythemia vera. The proposed mechanism for hearing loss in patients with hematologic disease is complex. Many studies have revealed histopathologic changes of the temporal bones in patients with leukemia and they are classified into 4 main categories: 1) leukemic infiltration, 2) hemorrhage, 3) infection, and 4) hyperviscosity [3,4].

The mechanism of hearing loss in PV is slightly different from that in leukemia. Thrombotic pathogenesis is highly suggested as a pathomechanism of hearing loss. In PV, thrombosis is caused by elevated hematocrit, increased RBC mass, high blood viscosity, persistent granulocyte or platelet activation, and increased acute phase reactants [5]. The cochlea and vestibule are supplied by the labyrinthine artery, which is a terminal artery arising from the basilar artery. Occlusion of the labyrinthine artery by thrombosis will result in hearing loss [4]. Inner ear micro-hemorrhage is another possible mechanism of hearing loss in PV,

although hemorrhage is not as common as thrombotic complications in the pathogenesis of general symptoms of PV. De Ciccio [6] reported that 28 of 43 (65%) patients presented with bilateral sensorineural hearing loss in their study group, which comprised PV and essential thrombocytosis. In their cases, the patients presented with hearing loss as one of several symptoms in the course of their disease after diagnosis, not as an initial manifestation. Although hearing loss and tinnitus are among the various symptoms of PV, hearing loss or tinnitus as the first sign of PV has rarely been reported. Indeed, we could not find any case of PV with hearing loss and tinnitus as a presenting symptom in a search of the PubMed service. The patient in this case did not have any other typical symptoms related to PV except for tinnitus and hearing loss. Our patient's sudden hearing loss in the right ear of 1 year ago might also be the result of PV, although we could not confirm this because we were not able to collect his test results from last year.

Diagnosis of PV requires both major criteria and one minor criterion or the first major criterion and two minor criteria. This patient fills both major criteria, which are hemoglobin > 18.5 g/dL and the presence of the JAK2V617F mutation, and two minor criteria, which are hypercellularity in bone marrow biopsy finding and decreased serum erythropoietin level [7].

Treatment of PV is focused on minimizing vascular risks. Thrombosis prevention with low-dose aspirin is recommended for all PV patients. In newly diagnosed PV patients, cytoreduction by phlebotomy is usually performed. 350-450 mL of blood is drawn to obtain a normal hematocrit value.(1) In some cases of deafness in leukemia, hearing loss has improved with treatment [4,8]. Davis and Nilo [9] reported a patient with PV who showed hearing improvement following withdrawal of 500 mL of blood by phlebotomy. In our case, phlebotomy was performed 4 times as an initial treatment of PV. The laboratory values, including hematocrit and platelet number, improved to near normal limits, but hearing and tinnitus were not improved. Delayed treatment is thought to be the reason why his hearing was not recovered compared with the results of Davis and Nilo.

Conclusion

We report a patient with PV whose presenting symptoms were hearing loss and tinnitus. Hematologic diseases including PV should be considered as etiologies of sensorineural hearing loss and tinnitus. Early diagnosis and intervention may rescue hearing from permanent loss in these patients.

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