



CASE RAPORT

BILATERAL TOTALLY SUDDEN SENSORINEURAL HEARING LOSS IN A PATIENT WITH JUVENILE IDIOPATIC ARTHRITIS

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SUMMARY

A 9 year old child with a three year history of juvenile idiopathic arthritis developed bilateral sudden sensorineural hearing loss. This type of sudden totally bilateral sensorineural hearing loss has not previously been described in association with juvenile idiopathic arthritis. He did not responded to steroid treatment.

Keywords: juvenile idiopathic arthritis, sensorineural hearing loss, steroid treatment

JÜVENİL İDİOPATİK ARTRİTLİ BİR OLGUDA GELİŞEN BİLATERAL TOTAL ANİ İŞİTME KAYBI:OLGU SUNUMU

Anahtar Kelimeler: juvenil idiopatik artrit, sensörinöral işitme kaybı, steroid tedavisi

INTRODUCTION

Sudden hearing loss (SHL) is defined as a sensorineural hearing loss which develops in less than three days and determined at 30 dB and over in at least three consecutive speech frequency¹. Although the incidence of SHL is reported as approximately 5-20/100000 per year, it is considered as self-limited and these patients do not apply health institutions, so the real incidence is higher than estimated². It can be seen in all age groups although it is usually noted in young and middle ages. Although no difference between two sexes is present, it involves one side in 90% of the patients.

In this article, a case regarding a 9 year old patient who had diagnosis of Juvenile Idiopathic Arthritis two years ago and has developed total sensorineural sudden hearing loss in his left ear during the first month of treatment, and in his right ear two years later is reported.

CASE PRESENTATION

The nine year old male patient applied to our ENT (Ear Nose Throat) outpatient's clinic by his family with sudden hearing loss in his right ear. In history, it was noted that the patient came to pediatrics clinic 3 years ago because of bilateral ankle pain caused by motion with redness and swelling.

As a result of clinical examination, laboratory evaluation and follow-up, Naproxene Na 10 mg/kg twice a day was initiated with the diagnosis of persistence oligoarticular juvenile idiopathic arthritis. During the second month of the treatment, sudden hearing loss has developed in the left ear, but the patient's family stated that they did not apply to any health institution. They applied to our clinic the day after hearing loss also developed in his right ear within the third year of his diagnosis. By the history taken from the family, it has been determined that his mother had diagnosis of romatoidal arthritis, but there was no other people in his family experienced hearing loss. By the audiometric evaluation, bilateral total hearing loss was determined. As it was learned from the history of the patient that, sudden hearing loss firstly occurred in left ear three years ago and then in right ear at third year of treatment for oligoarticular juvenil romatoid arthrititis, the patient was diagnosed with sudden right ear totally sensorineural hearing loss. No other pathologies were found during otorhinolaryngological examination. Chest X-Ray and neck ultrasound evaluations were found as normal. There was no pathology in cranial MRI. The whole blood count, peripheral smear, biochemical values of the patient were found to be normal. Also, the ANA (-), RF (-), dsDNA (-), C3, C4, and BUN were normal. ASO, CRP, hepatitis markers, HIV and coagulometry tests were normal. Brucella group agglutination test, Monotest, VDRL, ASMA, AMA tests were negative. In LP and fundoscopic examination, no peculiarity was determined and also no abnormalities found in blood pressure values. The patient was supervised and the treatment is planned as one dose of prednisolone injection of 250 mg, followed by oral prednizolon of

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80 mg for twelve days, tapering it by 10 mg every day. On the 5th, 10th and 15th days following the start of treatment, no amelioration was determined in audiometric evaluation.

DISCUSSION

The congestion of the inner ear has a dangerous nature and cochlear vascular supply can be easily interrupted³. Cochlea is a very sensitive tissue prone to ischemia. Although the endocochlear potential could be re-established in cochlear anoxia lasting for 10-12 minutes, it fades away irreversibly in anoxic situation surpassing thirty minutes⁴. Sudden hearing loss cases arising from full or partial vascular occlusion have been reported in Buerger disease, atherosclerosis, intracranial aneurism, sickle cell crisis, macroglobulinemia, polycythemia vera, cardiopulmonary by-pass operation by the involvement of vascular occlusion⁵. Gussen has described serious degeneration in stria vascularis, thorough destruction in corti organ and such vascular degenerative changes as fibrosis and ossification in the cochlea in the advanced stages in histopathological studies of patients with sudden hearing loss associated with hypertension, congestive heart failure and kidney failure⁶. The most popular view in SHL today is the decrease of cochlear blood flow caused by etiological factor. It is believed that the disease in SHL develops in vascular pathological basis⁷.

Jaffe and Maassah have stated that viral pathogen causes sudden hearing loss by affecting blood flow⁸. The sudden development of hearing loss, its accompaniment by systemic vascular diseases and histopathological symptoms support vascular hypothesis.

Although the etiopathogenesis of collagen diseases is not definitely known, autoimmune mechanisms are considered. The finding that many patients with SNHL appear to benefit from glucocorticoid therapy, as well as the finding of cross-reacting circulating antibodies in many patients with sudden and rapidly progressive SNHL, suggest that at least a subset of SNHL cases are caused by inner ear autoimmunity. In addition, a number of well-known autoimmune diseases have been associated with SNHL. These include Cogan's syndrome, systemic lupus erythematosus, temporal arteritis and polyarteritis nodosa.

Juvenile Idiopathic Arthritis (JIA) is a disease emerging basically with peripheral arthritis, in which endogenous or exogenous antigens play a role in its pathogenesis, and which becomes evident with increased inflammatory response in the immune

system^{9,10}. The main diagnosis criteria of the disease are that it starts prior to 16 years of age, lasts for more than 6 weeks, with arthritis in at least one joint, and with no known etiology. Arthritis is defined as swelling in joint, temperature increase, movement limitation or redness. Pain may also accompany these symptoms^{9,11}.

The prevalence of JIA differs in countries. Incidence is 9.2-25/100.000, and average prevalence is between 12-113/100.000. In a study in our country, JIA prevalence was noted as 64/100.000¹². The etiopathogenesis of the clinical picture of symptoms under the title of JIA is not fully known. But two main reasons are considered. First is the immunological tendency in patients and the second is environmental effects. Although infections are the most emphasized ones among the environmental effects, stress and trauma also play an important role. The JIA diagnosis is fully based on clinical criteria. The diagnosis of JIA and establishment of entire clinical picture may take a long time. The patient may be followed with different diagnoses in the beginning. The disease does not have a specific laboratory data. Laboratory data only assist in differential diagnosis, in the differentiation among subgroups and in the follow-ups¹³.

Aspirin or non-steroidal anti-inflammatory drugs (NSAIDs) are administered in the treatment in the beginning. The most frequently used non steroid anti-inflammatory drugs in children are ibuprofen, indomethacin, tolmetin or naproxen sodium. These medications are administered especially in children less than 12 years of age. Most oligoarthritis respond NSAID treatment dramatically¹⁴.

The side effects of NSAIDs are anorexia, gastritis and gastrointestinal bleeding; less frequent side effects are those of hepatic, renal and central nervous system. Patients administered with high doses of NSAID should be followed at intervals of 3-6 months for occult blood in the stool, hepatic enzymes and renal functions. Since NSAID alone is not mostly effective in treatment, other lastingly effective and stronger anti inflammatory drugs are required. Sensorineural hearing loss related to NSAID use has not been determined so far.

Markusse et al. have presented a case of 26 years old patient with sensorineural hearing loss in an acute stage of Still disease, and have determined that the patient's hearing loss recovered after the steroid treatment dramatically¹⁵.

In the light of these findings, a patient with the diagnosis of persistent oligoarticular Juvenile Idiopathic Arthritis has been presented in our article



with the first time with total bilateral sudden hearing loss in the literature; and no recovery has been detected after the steroid treatment following the diagnosis at the audiometric examination repeated on the 5th, 10th and 15th days of the treatment.

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