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Case Report

Hearing Loss in Systemic Onset Juvenile Idiopathic Arthritis (Sojia) -A Single Case Study

Thenmozhi K[1]*, Suriya Kala S[2], and Shanmugapriya E[3]

[1]*Audiologist & Speech Language Pathologist, Institute of Speech & Hearing, Chennai-03, Tamil Nadu, India*

[2]*Intern, BASLP, Institute of Speech & Hearing, Chennai-03, Tamil Nadu, India*

[3]*Intern, BASLP, Institute of Speech &Hearing, Chennai-03, Tamil Nadu, India*

***Corresponding author**

Thenmozhi K, Audiologist & Speech Language Pathologist, Institute of Speech & Hearing, Chennai-03, Tamil Nadu, India, Tel no: 9994609892

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Abstract

Juvenile Rheumatoid Arthritis is a common type of arthritis in children under the age of sixteen. Females are more affected than males with ratio of 2:1. Systemic onset Juvenile Idiopathic Arthritis (SoJIA) is an immune driven inflammatory condition that develops in children. Although SoJIA can occur any time during childhood it usually starts before age five. Inflammation of internal organs and MAS (macrophage activation syndrome) is closely associated with SoJIA. Children with SoJIA have high levels of two powerful inflammatory protein (cytokines) called Interleukin -1 and Interleukin-6. The management of SoJIA follows up with NSAIDs, Corticosteroids, DMARDs and inflammatory cytokine blockade. The incudomalleolar and incudostapedial joint are synovial in nature and hence could be involved in inflammatory process. Immune complex mediated vasculitis and ototoxic side effects may cause a pathological inner ear. Thus individuals with SoJIA are at higher risk of hearing impairment.

INTRODUCTION

Rheumatoid Arthritis (RA) is an inflammatory disease of multi factorial origin, including genetic predisposition and characterized by immune - driven, chronic inflammation [1,2]. Juvenile Idiopathic Arthritis (JIA) has some definite differences from RA in terms of genetic heterogeneity, phenotypically diverse presentations such as systemic type, polyarthritis, or oligoarthritis, however, both of them share the common pathogenic mechanisms [3-10].

International League of Associations for Rheumatology (ILAR) [6] has given two groups [1]. Group with Juvenile Idiopathic Arthritis (JIA): Systemic Arthritis, Oligoarticular Arthritis, Polyarticular Arthritis, Psoriatic Arthrits, Enthesitis related arthritis [2]. Other Autoimmune Disorders (AID): Juvenile systemic lupus erthemathosus, Juvenile linear scleroderma, Juvenile Sjogren syndrome.

Unlike other forms of Juvenile Idiopathic Arthritis in which arthritis is the major problem, children with Systemic onset Juvenile Idiopathic Arthritis (SoJIA) typically also have systemic symptoms. A high fever at morning which falls down at evening or vice versa that lasts at least for two weeks either precedes or accompanies the arthritis [7]. It affects one or more joints in body and inflammation of internal organs specifically lymphadenopathy, hepatomegaly, splenomegaly [9]. A rare – life threatening complication called macrophage activation syndrome (MAS) is closely associated with SoJIA. Flat, pale, pink, rash frequently appears on child`s trunk, arm, legs, sometimes associated with pericarditis or pleuritis [7]. Although SoJIA can occur any time during childhood it usually starts before age five [8].

There are two types of immune system: innate and adaptive. In SoJIA, the adaptive immune system doesn't work properly. This becomes overactive and targets the body itself leading to auto immune disease. But recently, researchers suggest SoJIA as an auto inflammatory disease rather than autoimmune disease since children with SoJIA usually don't have auto antibodies in their blood. Children with SoJIA have high levels of two powerful inflammatory proteins (cytokines) called Interleukin-1 (IL-1) & Interleukin- 6 (IL-6) [8].

In a study by A. Siamopoulou-Mavridou et al. (1990) they observed a high incidence of type A_S tympanograms and presence of acoustic reflex which strongly indicate that Juvenile Chronic Arthritis changes middle ear function. This result suggests that a pathological stiffening of middle ear occurs mainly in patients with severe polyarthritis of long duration. Conductive loss is suspected because the incudomalleolar and incudostapedial joints are synovial which might involve in inflammatory process. Copeman (1963) described three patients with Rheumatoid arthritis as otoarthritis, brought attention to the possibility of conductive impairment of Rheumatoid arthritis.

Here, abnormal tympanograms with worse air conduction thresholds and larger air bone gap at lower frequency suggest subclinical middle ear involvement. However hearing loss at high frequencies suggests inner ear involvement. The result suggests a dual effect of disease on both middle and inner ears of patients with Juvenile Idiopathic Arthritis. Possible explanations for sensory neural hearing loss are immune complex-mediated vasculitis of inner ear and ototoxic side effects. Goodwill et al. (1972) showed that the significantly greater hearing loss in patients with Rheumatic arthritis was of sensory neural, and not of conductive types.

Multi-disciplinary care, lifestyle modifications (non-inflammatory diets, anti-oxidants), NSAIDs (Non-Steroidal Anti-Inflammatory Drugs), DMARDs (Disease-Modifying Anti-Rheumatic Drugs), Corticosteroids and Inflammatory cytokine blockade are major lines of treatment [8]. The management protocol also includes monitoring for side effects due to drug intake until the disease achieves remission. Since the incudomalleolar and incudostapedial joint are synovial in nature, it could be involved in inflammation [8]. The SoJIA itself causes hearing impairment and the drug-line treatment too causes ototoxicity which may lead to hearing impairment. The above mentioned drugs have an ototoxic effect which makes hearing evaluation, a crucial one. However until any obvious signs of reduced hearing sensitivity, hearing evaluation doesn't take a significant place in SoJIA management during past years.

In children, the discrimination of symptoms due to eye or ear involvement is limited. Therefore, monitoring of those subjects with minimal or no symptoms are important. Since arthritis may not be the key sign, diagnosis of SoJIA is quite challenging.

The general protocol in diagnosis of SoJIA includes Health history, Physical exam, Imaging tests, Laboratory findings.

CASE PRESENTATION

A nine year and six months old female child was brought with a complaint of reduced hearing sensitivity in both ears since six months. She was diagnosed with SoJIA after all recommended test protocol results at the age of four. She was under low dose steroid medication until her six years of age. Successively at six years of age, the child also developed insignificant generalized lymphadenopathy. She was then under the medications of methotrexate and corticosteroids. At the age of seven, multiple lymphadenopathies were observered in the child and diagnosed with latent TB (Tuberculosis) from the result of MANTOUX test. Hence gradually anti-tubercular drugs were also introduced to the child.

At the age of seven years and six months, she was hospitalized for about a week due to recurrent episodes of vomiting, severe headache (not relieved by antibiotics) and pain in hip joint, abnormal eye movement and frog like posture of presentation. MRI report shows a normal study of brain and spine. At eight years of age, acute adenopathy on right side and bilateral insignificant inguinal nodes inflammation were diagnosed by MRI of neck. At nine years of age, the child had cervical lymph node TB which was diagnosed by FNAC (Fine Needle Aspiration Cytology). The child was under the second line medications of anti-tubercular drugs and Corticosteroids then. The child then had a complaint of reduced hearing sensitivity in both ears at nine years of age.

METHODS

As the child complained of reduced hearing sensitivity, the following audiological evaluation was carried out at the Institute of Speech and Hearing, Madras Medical College. The evaluation started with the collection of complete case history, health history with supportive evidences of medical reports. The audiological evaluation included Pure Tone Audiometry, Immittance audiometry and Speech Audiometry.

RESULTS

The child had frequent ear discharge in both ears which usually lasted for one to two days and relieved without any medical management. The Otoscopic examination revealed bilateral intact tympanic membrane and a dry ear canal. Pure tone audiometry examination revealed bilateral moderate mixed hearing loss with high frequency slope. The hearing loss was more prominent in high frequency regions which gradually progressed from 1 kHz. Speech audiometry results showed speech recognition threshold of 50 dBHL in right ear and 55 dBHL in left ear with speech identification score of 85 % in right ear and 80 % in left ear.

Immitance audiometry results showed bilateral 'A' type tympanogram with absent ipsilateral and contralateral acoustic reflexes in both ears. Since the child did not show any cue to Retro cochlear pathologies, the above mentioned audiometric tests were sufficient for preliminary hearing evaluation.

MANAGEMENT

Autoimmune rheumatic diseases are characterized mainly by structural and/or functional involvement of multiple organs and systems, which occurs due to inflammation and can cause permanent damage. Since they present variable evolution and prognosis in multisystem involvement, it is necessary to monitor manifestations that are not very symptomatic. Hearing impairment is a major complication of JIA which is not very symptomatic at early stages and masked by other major complications.

Loss can be reversible (mostly in initial stages of the disorder) or irreversible and also can be fluctuating (on disease course – remission). Hence a constant follow up is crucial to enhance adequate hearing management. Hearing aid is a better choice for sensory neural hearing loss which can be more beneficial when programmable aids are used. In case of conductive hearing loss, a medical or surgical management is recommended. They include antibiotics (ear infection), Stapedectomy, Stapedotomy (stapes fixation), or Ossicular replacement surgery. However surgical management is not a first line of treatment in case of SoJIA induced hearing loss.

DISCUSSION

Auditory impairment may be related to several factors such as: inflammatory and autoimmune mechanisms, involvement of the small middle ear joints, and sensitivity to some anti-inflammatory drugs and immunosuppressants including non-hormonal anti-inflammatories, prednisolone, methotrexate, hydroxychloroquine, azathioprine and biological agents [6].

The child was diagnosed with SoJIA at the age of four. The child has been living with SoJIA for about six years and with its complications such as lymphadenopathy, hepatomegaly, and lymph node tuberculosis. The child is under medication of corticosteroids, NSAIDs, DMARDs and anti-tubercular drugs.

The hearing impairment due to SoJIA can occur by two ways.

(1) (1) Disorder mediated and (2) Drug mediated. Disorder mediated:

- The inflammation of synovial ossicular joint is followed by stiffness of tympanic membrane and ossicular chain [1].
- Osteogenesis in rheumatoid arthritis leads to fixation along with bacteria mediated acute otitis media (Salomonsen et al, 2010).
- Pro inflammatory cytokines (Interleukin-6) may harm the inner hair cells by an oxidative process [1].
- Destruction of the cochlear hair cells or the inner ear due to immune complex deposition [1].
- Rheumatoid nodule, swollen and tender joints. Rheumatoid nodules are the most prevalent extra-articular manifestation that may present in different locations, however commonly found subcutaneously at points of pressure. Various ear, nose, throat sites may be infected [1].

(2) Drug mediated:

- Drugs like Isoliazid and Methyl prednisolone causes insufficient heme formation and electrolytic changes [9]. This in turn causes insufficient local flow cochlea (stria vascularis and spiral ligament).
- DMARDs (methotrexate, sulfasalazine) cause immune suppression which may result in higher risk of infection that includes frequent middle ear infection. Specifically Hydroxychloroqine and chloroquine are associated with variable injuries to the cochlear sensory hair cells and also a decrease in neuronal population and supporting structures as well as atrophy of striavascularis [1].
- Corticosteroids taken for a long time may cause increased blood pressure, risk to infections since it suppresses the immune system, osteoporosis and fractures (thinning of bones), fluid retention, impaired electrolyte exchange mechanism which plays a main role in sound transmission, aseptic necrosis (damage to bones) [9].
- NSAIDs intake for long duration causes tinnitus, increased blood pressure, anemia [9].

Conductive hearing loss in RA may be related either to stiffness or to discontinuity of the ossicles in the middle ear [5]. Since the child was under immunosuppressants, she is vulnerable for infections leading to recurrent ear infection. These pathologies lead to the conductive component of hearing loss.

Ototoxic drugs which were taken by the child may have caused inner ear damage that lead to sensory neural component of hearing loss. The presence of a mixed type of hearing loss suggested a multifocal involvement of the audiological system in RA [5].

Evaluation of audiometric test should be integrated in RA assessment to detect and manage possible hearing impairment caused by the disease itself and/or by therapeutic intervention. Furthermore, patients must be informed by their physician of the risk of having auditory damage as a complication of the disease [5].

CONCLUSION

Risk of hearing loss is higher in Juvenile Idiopathic Arthritis patients and seems to be associated to disease duration. Hearing loss in Juvenile Idiopathic Arthritis is directly proportional to the Disease Activity Score - 28 [5]. Hearing impairment may be caused by the dual effect of middle and inner ear pathologies in SoJIA patients. Symptoms related to hearing loss and audiological alterations were more frequent in children and adolescents with Juvenile Idiopathic Arthritis. Researchers say that the hearing loss may be reversible or irreversible. The hearing alterations also occurred in patients with no symptoms, indicating the need for systematic hearing assessment for those patients in their clinical routine [6]. Since study of hearing impairment in SoJIA patients are very limited, an elaborative study is necessary to understand the involvement of hearing acuity. Hence a constant follow up is crucial [11].

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