Sensorineural hearing loss in Kawasaki disease

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ABSTRACT

Kawasaki disease is a common nonspecific vasculitis seen in childhood. The most significant long-term sequela is coronary artery aneurysm. However, the spectrum of complications involves not only the heart, but also other organs such as the eyes, skin, kidneys, gallbladder, liver, and central nervous system. Sensorineural hearing loss (SNHL) is a relatively unrecognized complication of the disease. Although most of the complications (except coronary artery aneurysm) are self-limiting, SNHL can be persistent. It is, especially important in infants and young children who might not be able to report the hearing deficits and are most likely to have cognitive and speech delays if this hearing loss is not addressed in a timely manner. We report a child with Kawasaki disease who had SNHL during the 2nd week of the illness. The aim of this article is to briefly review the pathophysiology behind this hearing loss and strongly emphasize the importance of universal hearing evaluation in all children diagnosed with Kawasaki disease. This screening in children with Kawasaki disease may provide some timely intervention if needed. Since most Kawasaki disease patients will be seen by cardiologists, we hope to create more awareness about this complication to the cardiology community as well.

Keywords: Fever, hearing loss, Kawasaki disease, steroids

INTRODUCTION

SNHL has been an under-recognized complication of Kawasaki disease. Even after the first reported case of SNHL in association with Kawasaki disease in 1988,[1] not much research has been done with regards to this complication. With the reported incidence of SNHL in approximately 36% patients in patients with Kawasaki disease,[2] we aim to create awareness about this under-recognized complication.

CASE REPORT

A 6-year-old girl presented to the hospital emergency with 9 days history of fever up to 104 F. She had bilateral nonpurulent conjunctivitis, cracked lips, intermittent abdominal pain and decreased activity. She was seen on the 4th day of her illness by her pediatrician, diagnosed with a viral syndrome and sent home with supportive care. On follow-up at 48 h, she was also noted to have cervical lymphadenopathy. Concerns for Kawasaki disease were raised, so she was referred to the hospital for further evaluation.

Her past medical history was negative except for one episode of urinary tract infection as an infant. Her family history was noncontributory. Her immunizations were up to date for her age, and her development had always been on target.

Physical examination on admission revealed a nontoxic, afebrile child with bilateral nonpurulent conjunctivitis and cracked dry lips. Other findings included bilateral, posterior cervical, nontender lymphadenopathy measuring approximately two by two cm. She was well oriented and had clear speech comprehension. Her neurological examination was unremarkable.

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Laboratory evaluation showed normal hemoglobin of 11.7 g/dL with a mildly elevated white cell count of 11,700/cumm and normal platelet count of 452,000/cumm. Viral studies by polymerase chain reaction, including adenovirus and influenza virus were negative. Her C-reactive protein and erythrocyte sedimentation rate were elevated, 5.8 mg/dL and 63 mm/h respectively. Liver enzymes, bilirubin, kidney function tests, and urine analysis showed no abnormality. Her electrocardiogram showed normal sinus rhythm and echocardiogram done on the day of admission was normal. Her clinical presentation was met the clinical criteria for Kawasaki disease (fever for more than 4 days and cervical lymphadenopathy, nonpurulent bulbar conjunctivitis, cracked lips, and desquamation of the skin at nail beds in follow-up). She was treated with one dose of intravenous immunoglobulin (2 g/kg/dose) and high-dose aspirin (80 mg/kg/day). During hospitalization and prior to starting these medications parents noticed decreased hearing as evidence by the patient not responding to verbal commands unless talking loudly to her. Apart from this complaint, the patient improved clinically. She was discharged home on 81 mg/day of aspirin.

The patient's hearing loss progressed after discharge from the hospital, and she had trouble with the comprehension of spoken words. There was interval improvement in conjunctivitis, but the redness and swelling of her lips persisted and she was also noted to have superficial skin peeling. She otherwise remained in good health and afebrile. On follow-up with her pediatrician, a hearing test was performed, which revealed moderate to severe bilateral SNHL across all frequencies. She was readmitted for further evaluation. A brainstem evoked response audiometry performed showed absence of wave V latency below 60 dB bilaterally. This was suggestive of precochlear or cochlear receptor dysfunction bilaterally. Her tympanometry was normal bilaterally.

Blood salicylate levels were <4 mg/dL. Cerebrospinal fluid analysis showed seven white cells and three red cells with normal biochemistry. Culture of cerebrospinal fluid was negative for bacteria and cytomegalovirus. Culture from the eye was negative for herpes simplex virus, adenovirus, and enterovirus. A throat culture was negative for herpes simplex virus, cytomegalovirus, adenovirus, influenza A and B, parainfluenza 1, 2 and 3; and enterovirus. Magnetic resonance imaging of the brain was negative for any intracranial lesion with normal internal auditory canal and inner ear structures. She was started on high-dose methylprednisolone 30 mg/kg intravenous for 3 days. She was discharged home on aspirin 81 mg/day and prednisolone 30 mg oral daily for 5 more days.

An audiogram done 2 weeks after completing treatment with steroids showed significant improvement of the hearing with normalization in the high frequencies (1000-6000 Hz) and moderate improvement in the lower frequencies (250-1000 Hz) with persistence of mild SNHL.

DISCUSSION

Kawasaki disease is a systemic vasculitis of unknown cause and the leading cause of acquired heart disease in North American and Japanese children.[3] This disease was first reported in 1967 by Dr. Tomisaku Kawasaki.[4] In the United States, the incidence in 2006 was estimated to be 20.8/100,000 in children <5 years of age.[5] Coronary artery aneurysms develop in approximately 20% of untreated children and are the most common reported the cause of morbidity and mortality in this disease.[6]

Despite being reported as early as 1988 by Suzuki *et al.*[1] from Japan and in 1990 by Sundel *et al.*[7] from the United States, hearing loss associated with Kawasaki disease has been fairly unrecognized in the literature. In a recent review by Smith and Yunker,[2] 36% of patients with Kawasaki disease were noted to have some degree of hearing loss presenting usually in the first 30 days of disease onset. The severity of SNHL was mild to profound, and it was noted to be bilateral in 73% of patients. SNHL persisted in 14% of the patients at follow-up. The period of follow-up varies for each study ranging from 8 days to 6 years with the majority of them being followed for approximately 6 months.

The exact etiology for the SNHL is not clear. High-dose acetylsalicylic acid has been hypothesized to be the cause for the hearing loss.[8] This was definitely not the cause in our patient as the hearing loss was noticed even before she received any doses of aspirin, and her salicylate levels were undetectable. Since Kawasaki disease is a multisystemic vasculitis, it might be that vasculitis results in an altered osmotic balance in the inner ear hair cells or possibly axonal neuropathy of the auditory nerve. The cause of Kawasaki disease has been proposed to be an infectious etiology and it may be argued that the SNHL is due to direct cytopathic effects on the labyrinth or cochlea as is seen in other viral diseases such as rubella or mumps.[9]

Several factors have been hypothesized to predict the SNHL and these include delayed initiation of treatment (after 10 days of symptom onset), thrombocytosis, elevated erythrocyte sedimentation rate and prolonged anemia. Our patient presented late and received intravenous immunoglobulin on the 10th day of illness at which point she had already developed SNHL.

It is important to realize that coronary artery aneurysms occur in nearly 20% of patients with Kawasaki disease, and the incidence drops down to around 5% with proper treatment with aspirin and intravenous

immunoglobulin.[6,10] The reported incidence of SNHL (36%) is much higher than coronary artery aneurysms and is bilateral in 73% of these patients. Approximately, 85% of children affected with Kawasaki disease are <5 years of age with peak incidence occurring at 18-24 months.[11] Since speech and language development is very much dependent on hearing at this age, unrecognized SNHL can cause cognitive and speech delays. Because of the reported high incidence of the SNHL we strongly recommend screening all children with Kawasaki disease for hearing loss. Since the SNHL can happen at any time in the disease process, we consider it reasonable to screen at the time of initial diagnosis and if normal repeat an audiogram 6 weeks after. Further screening can be guided by clinician judgment.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Suzuki H, Yanagawa T, Kihira S. Two cases of hearing loss associated with Kawasaki disease. Clin Pediatr (Jpn) 1988;41:167-72.
2. Smith KA, Yunker WK. Kawasaki disease is associated with sensorineural hearing loss: A systematic review. Int J Pediatr Otorhinolaryngol 2014;78:1216-20.
3. Yanagawa H, Nakamura Y, Ojima T, Yashiro M, Tanihara S, Oki I. Changes in epidemic patterns of Kawasaki disease in Japan. Pediatr Infect Dis J 1999;18:64-6.
4. Kawasaki T. Pediatric acute mucocutaneous lymph node syndrome: Clinical observation of 50 cases. Arerugi (Jpn J Allergy) 1967;16:178-222.
5. Holman RC, Belay ED, Christensen KY, Folkema AM, Steiner CA, Schonberger LB. Hospitalizations for Kawasaki syndrome among children in the United States, 1997-2007. Pediatr Infect Dis J 2010;29:483-8.
6. Dajani AS, Taubert KA, Gerber MA, Shulman ST, Ferrieri P, Freed M, *et al.* Diagnosis and therapy of Kawasaki disease in children. Circulation 1993;87:1776-80.
7. Sundel RP, Newburger JW, McGill T, Cleveland SS, Miller WW, Berry B, *et al.* Sensorineural hearing loss associated with Kawasaki disease. J Pediatr 1990;117:371-7.
8. Kim SM, Jo JM, Baek MJ, Jung KH. A case of bilateral sudden hearing loss and tinnitus after salicylate intoxication. Korean J Audiol 2013;17:23-6.
9. McKenna MJ. Measles, mumps, and sensorineural hearing loss. Ann N Y Acad Sci 1997;830:291-8.
10. Newburger JW, Takahashi M, Beiser AS, Burns JC, Bastian J, Chung KJ, *et al.* A single intravenous infusion of gamma globulin as compared with four infusions in the treatment of acute Kawasaki syndrome. N Engl J Med 1991;324:1633-9.
11. Gardner-Medwin JM, Dolezalova P, Cummins C, Southwood TR. Incidence of Henoch-Schönlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. Lancet 2002;360:1197-202.