



CASE REPORT

Sensorineural hearing loss associated to Kawasaki Disease

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Abstract

Objective: Kawasaki disease is a systemic idiopathic self-limited vasculitis of small and medium-sized vessels. Thirteen cases of sensorineural hearing loss during the evolution of this disease have been described in the literature. We describe a case of an infant with Kawasaki disease who developed sensorineural hearing loss during the acute phase. This case report shows a complication of Kawasaki disease, with few citations in literature, enhancing the importance of careful evaluation of these patients.

Description: nineteen-month-old-boy, formerly healthy, with persistent daily fever seven days before admission, associated with irritability, bilateral nonexudative conjunctivitis and maculopapular erythema on his trunk. There was later development of arthritis on wrists, elbows, knees, and ankles, and swelling associated with desquamation of hands and feet, which prevented him from walking. The diagnosis of Kawasaki disease was established according to the American Heart Association's criteria, and the child was conventionally treated. There was regression of clinical manifestations three days after the beginning of treatment. However, one month after the onset of symptoms, the family noticed an unsatisfactory response to sound stimuli. The evaluation of auditory acuity through BERA (Brainstem Evoked Responses Audiometry) revealed severe bilateral sensorineural hearing loss.

Comments: routine examination of auditory acuity in children with Kawasaki disease may help identify sensorineural hearing loss at an early stage.

J Pediatr (Rio J) 2002; 78 (1): 71-74: vasculitis, mucocutaneous lymph node syndrome, sensorineural hearing loss.

Introduction

Kawasaki disease (KD) is a self-limited, systemic vasculitis of unknown etiology that affects the small and medium-sized blood vessels of the body.¹ Coronary vasculitis is highly frequent, affecting approximately 25% of untreated patients.² Although the severe involvement of

the central nervous system is not so frequent, thirteen cases of sensorineural hearing loss were reported in the literature.^{3,4} (Table 1). In this report, we describe the case of an infant with Kawasaki disease who developed sensorineural hearing loss during the acute phase of the condition. Our objective is to show a severe complication of KD that is rarely found in medical literature, enhancing the necessity of a careful global assessment of these patients.

Case report

Healthy male aged one year and seven months, from the town of Uberlândia, state of Minas Gerais, presented with persistent fever for seven days, associated with irritability,

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nonexudative bilateral conjunctivitis, maculopapillary erythema on the trunk. The patient developed arthritis on his fists, elbows, knees, and ankles, edema and desquamation of the hands and feet, which prevented him from walking. Laboratory exams revealed Hb=11.0g/dl; leukocyte count=7,600/mm³; platelet count=161,000/mm³; erythrocyte sedimentation rate =55 mm/h, C-reactive protein=24mg/l; ASO=50IU; albumin=4.2g/l; negative serological tests for toxoplasmosis, mononucleosis, rubella and cytomegalovirus. Echocardiographic and electrocardiographic abnormalities were not observed.

The diagnosis of KD was established according to the following American Heart Association (AHA)^{5,6} criteria: persistent fever for at least five days; changes of the peripheral extremities, such as erythema and edema of the hands and feet in the acute phase, and membranous desquamation of the finger and toe tips in the convalescent phase; polymorphous exanthema; bilateral nonexudative painless conjunctivitis; oropharyngeal changes including erythema, fissuring of the lips, strawberry tongue and diffuse mucosal injection of the oropharynx. The patient met the first four criteria, which characterized him as a case of incomplete Kawasaki disease.

The patient was conventionally treated with intravenous gammaglobulin (2g/kg/day) and aspirin (100mg/kg/day up to 14 days after disease onset). The clinical symptoms

resolved three days after treatment was implemented. However, the child showed difficulty walking even though arthritis had been controlled and, one month after the onset of symptoms, the patient's family noted unsatisfactory response to sound stimuli. CT scanning exams of the brain and temporal bones were normal.

The assessment of hearing function was obtained by BERA (Brainstem Evoked Responses Audiometry - this exam is recommended for the evaluation of hearing in newborns and infants who cannot be submitted to conventional tone stimulation. This exam consists in recording electrical responses triggered off by sound stimuli traveling along the auditory pathway to the brainstem. The test interpretation is based on the action potentials that originate from the cochlear nerve (CN VIII) and from auditory pathways, and go up to the brainstem. There are usually six waveforms: I, II, III, V, VI and VII, which respectively originate in the distal portion of the cranial nerve (CN VIII), in the proximal CN VIII portion, in the cochlear nucleus, in the superior olivary complex, in the inferior colliculus, and in the medial geniculate body. No waves were generated in the presence of 90-dB stimuli in both ears, which suggests severe to profound sensorineural hearing loss on both sides. Auditory function was not improved despite prednisone treatment (initial dose 2mg/kg/day) for two months.

Table 1 - Summary of cases of Kawasaki disease, with sensorineural hearing loss, described in the literature

Case	Age	Disease period	Hearing loss	Treatment
1 ⁽³⁾	4 years	1 year	Unilateral (R)	NC
2 ⁽³⁾	4 years	28 days	Severe bilateral	AAS+corticoid
3 ⁽³⁾	5 years	5 years	Unilateral (R)	NC
4 ⁽³⁾	5 years	5 years	Unilateral (R)	NC
5 ⁽³⁾	1 year	10 days	Trans. unilateral (L)	AAS+IVIg
6 ⁽³⁾	3 years	10 days	Severe bilateral	AAS+corticoid
7 ⁽³⁾	5 years	6 months	Severe bilateral	AAS
8 ⁽³⁾	1 year	13 days	Trans. moderate bilateral	AAS+IVIg
9 ⁽³⁾	4 years	10 days	Severe bilateral	AAS+IVIg+cortic.
10 ⁽³⁾	12 years	14 days	Unilateral (R)	AAS+IVIg
11 ⁽³⁾	7 months	?	Severe bilateral	AAS
12 ⁽³⁾	6 years	18 days	Severe unilateral (L)	AAS+IVIg+cortic.
13 ⁽³⁾	13 years	14 days	Moderate bilateral	AAS+IVIg+cortic.
14*	1 year	28 days	Severe bilateral	AAS+IVIg+cortic.

*described case; NC = not carried out; AAS = acetylsalicylic acid; IVIg = intravenous immunoglobulin; trans. = transient; R = right; L = left

Discussion

The major KD complications are related to cardiac involvement, especially coronary vasculitis.^{2,7,8} We observed another kind of severe and irreversible complication in this case: sensorineural hearing loss; which is not frequently reported in the literature.^{3,4} The presence of hearing loss during the subacute phase in an infant with KD underscores the idea that this is an inflammatory systemic disease. Therefore, it is important to carefully assess these patients, since vascular lesions may be found in any human tissue.

Neurological symptoms are very rare, although extreme irritability, possibly due to aseptic meningitis, is common in the acute phase of KD.⁸ Seizures, involvement of cranial pairs (7th pair) and hemiparesis caused by infarction or thrombosis are reported in the literature.^{1,3,4}

Until today, only thirteen cases of sensorineural hearing loss associated with Kawasaki disease have been reported.^{3,4} However, this seems to be an underestimation, since hearing function can be discreet and transient in this disease, being detected only by audiometry or BERA (younger children). As with our patient, hearing loss is normally severe, bilateral, and irreversible, but may be unilateral and transient. Although most patients with hearing loss have been treated with aspirin (anti-inflammatory doses), it is unlikely that the condition is caused by the use of this medication, since the serum levels of aspirin always remained under 20mg/dl. In addition, differently from aspirin-induced ototoxicity, hearing loss was not transient in most reported cases.

The time between the diagnosis of KD and the perception of hearing loss ranges from 10 days to 5 years in reported cases.^{3,4} This delay is correlated with the difficulty in establishing the diagnosis in younger children, especially those aged less than two years, when parents may fail to identify the problem perception may fail, and conventional audiometry cannot be performed. In our case, two factors may have contributed to the delayed perception of hearing loss: the persistence of irritability one week after disease onset and the fact that the child had just acquired a more elaborate speech.

Some patients with sensorineural hearing loss associated with KD showed other neurological symptoms, such as facial palsy and ataxia.^{3,4} Our patient had frequent falls initially attributed to joint pain and later related to vestibular involvement, due to the presence of dizziness.

Several immunological abnormalities are observed in patients with KD.⁸ Initially, there is activation of vascular endothelial cells and adhesion molecules induced by cytokines associated with the increased number of CD4 cells, with the reduced number of positive, active CD8 cells, and with the enhanced production of immunoglobulins by B cells. The physiopathological mechanism that causes

vasculitis in the middle ear in patients with KD and with sensorineural hearing loss is still unknown. Possibly, there is impairment of cochlear vessels or vasa nervorum, which results in the involvement of the auditory and vestibular nerve.

The conventional treatment of KD with aspirin and intravenous gammaglobulin and, in some cases, with oral or parenteral corticosteroids, does not interfere with the progression of hearing loss.^{8,9} The impairment of the internal ear may occur in the first days, that is, during the initial febrile phase, before the administration of specific treatment, which is usually implemented between the fifth and tenth day after disease onset when diagnosis can be made by the established criteria. Early treatment (first week) with intravenous gammaglobulin and/or corticosteroids may help prevent this complication.

Sundel et al.⁴ suggest that it is necessary to have at least one audiometric evaluation during the follow-up of all patients with Kawasaki disease, since transient or persistent sensorineural deafness may affect these patients, especially those with long-lasting fever and with lab exams that suggest severe systemic infection.

Our conclusion is that routine exams to assess hearing function in infants with Kawasaki disease may detect sensorineural hearing loss at an early stage.

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