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Atypical Cogan Syndrome Mimicking Acute Rheumatic Fever

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Cogan syndrome is a syndrome of non-syphilitic interstitial keratitis associated with vestibuloauditory deficits. We report a 10 year-old male child who presented with fever, acute polyarthritis, and unilateral red eye and was diagnosed as acute rheumatic fever. Subsequently unilateral hearing loss was detected and the child was diagnosed to have atypical Cogan syndrome.

Key words: *Acute rheumatic fever, Arthritis, Atypical Cogan's syndrome, Polyarthritis.*

Cogan syndrome is a syndrome of non-syphilitic interstitial keratitis associated with vestibuloauditory deficits, that progresses to complete deafness within 2 years [1]. The term 'Atypical Cogan's syndrome' was suggested later for cases in which sensorineural hearing loss is associated with ocular inflammation distinct from interstitial keratitis, such as uveitis, scleritis, conjunctivitis or retinal vasculitis [2,3]. Patients in which the interval between the onset of ocular and audiovestibular manifestations is more than two years are also labeled as atypical Cogan's syndrome [2,3]. Majority of patients with this syndrome develop significant systemic symptoms and often present a diagnostic challenge [3-6]. We describe a 10 year old male child who presented to us with acute polyarthritis and was later diagnosed as atypical Cogan syndrome.

CASE REPORT

A 10 years old male child was referred to our center as a case of acute rheumatic fever. There was history of low grade fever for 7 days, severe pain and swelling in multiple large joints for 4 days and redness of the right eye for 4 days. There was no history of any antecedent upper respiratory tract

infection. On examination, there was redness, swelling, tenderness and restriction of movement in bilateral knee, ankle and wrist joints. On ophthalmologic evaluation, visual acuity in the right eye was 2/60 with presence of anterior uveitis and inflammatory hypopyon. Left eye was normal. Bilaterally pupillary reaction was normal. Except for mild hepatomegaly, rest of the systemic examination was unremarkable. Initial laboratory evaluation revealed mild polymorphonuclear leucocytosis, raised ESR and elevated C-reactive protein. His chest radiograph, electrocardiogram and echocardiogram were unremarkable. A differential diagnosis of acute rheumatic arthritis, and autoimmune disorder was considered and he was started on high dose aspirin (75mg/kg/day). In addition topical steroids and mydriatics were given for the right eye. The joint symptoms responded dramatically to aspirin, which further tilted the diagnosis in favour of acute rheumatic arthritis.

On day 3 of admission, patient complained of right-sided hearing loss and tinnitus for last 7-8 days. There was no history of vomiting and vertigo. Local ear examination was normal. A BERA screen showed no response in right ear. Auditory Steady State Response (ASSR) assessment showed profound deafness of right ear with mild hearing loss of left ear. MRI of the petrous bone was suggestive of right sided labyrinthitis. Other laboratory investigations including anti-streptolysin O, VDRL, RA factor and ANA were normal.

A diagnosis of atypical Cogan's syndrome was made and the child was started on oral prednisolone at the dose of 2mg/kg/day, and aspirin was stopped. The uveitis and the residual joint symptoms subsided completely within 3 days of starting steroids but hearing loss persisted. ASSR repeated after 7 days showed normal hearing on the left side and some improvement on the right side. The child was discharged on oral steroids. A follow up ASSR after 2 months showed further deterioration of hearing on the right ear. However, there was no recurrence of visual and joint symptoms. In view of worsening hearing loss, weekly oral methotrexate (10mg/m^2) was started as a second line immunosuppressive agent and oral steroids were concurrently tapered off within next 4 weeks. After one year of methotrexate, there have been no flare ups of the disease but the right sided hearing loss is persisting.

DISCUSSION

Cogan syndrome is a systemic vasculitis that mainly affects young adults but can appear from ages 3.5 month to 81 years [6-7]. In approximately 30% of the cases, the disease is preceded by an upper respiratory tract infection [4-7]. The disease generally begins with ocular or audiovestibular symptoms separated by months, or occasionally these can occur concurrently [4,5]. Audiovestibular symptoms of typical Cogan syndrome are similar to that of Meniere's disease (sudden onset of nausea, vomiting, tinnitus and vertigo) that usually progress to complete deafness within several hours to days [7]. Various systemic features like fever, headache, myalgia, arthralgia, arthritis, rash, abdominal pain, gastrointestinal bleeding, pleuritis, lymphadenopathy, splenomegaly, encephalitis, cerebral infarction and other vasculitis may also be present [3-8]. Approximately 10% cases are complicated with aortic insufficiency associated with aortitis and inflammatory or myxoid degenerative valve changes [3-8].

Cogan syndrome is considered to be an autoimmune disease. Autoantibodies to inner ear and epithelial proteins have been documented in some patients with Cogan syndrome [5-9]. The diagnosis is based on the constellation of clinical features. Corticosteroids are most widely used drugs in the management. Steroid resistant cases are managed with methotrexate, azathioprine, cyclophosphamide and cyclosporine. However, audiovestibular symptoms are generally resistant to drug therapy [3-8].

The disease course is variable. Mostly it presents as an initial flare followed by a chronic and slowly progressive course. Some may have recurrent flare-ups secondary to intercurrent infections with remissions in between. Deafness is usually permanent. Death can occur due to cardiac failure. The patient should be kept under regular follow-up in view of possibility of life-threatening complications and possible side effects of immunosuppressive agents [5].

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Scarlet Fever Caused by Community-associated Methicillin-resistant *Staphylococcus aureus*

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We describe a previously healthy 2½-year-old boy with staphylococcal scarlet fever associated with acute suppurative otitis media due to community-associated methicillin-resistant *Staphylococcus aureus*. The patient was successfully treated by spontaneous drainage in combination with trimethoprim-sulfamethoxazole therapy.

Key words: *Acute suppurative otitis media, Community-associated methicillin-resistant Staphylococcus aureus, Staphylococcal scarlet fever.*

Acute otitis media (AOM) is pervasive in children with high incidence rates reported both in developed and emerging nations [1]. Recently, strains of community-associated methicillin-resistant *S. aureus* (CA-MRSA) are increasingly found in skin and soft-tissue infections of children, usually linked to intravenous drug abuse, cystic fibrosis, chronic diseases and repeated antimicrobial therapy [2]. However, CA-MRSA otorrhea after tympanostomy tube insertion in well children has been reported previously [3]. Here we describe a child with bilateral acute suppurative OM and staphylococcal scarlet fever (SSF) associated with CA-MRSA. To the best of our knowledge, this is the first pediatric case of such an association.

CASE REPORT

A previously healthy 2-year-and-6-month-old boy was seen in the emergency department because of the acute onset of turbid discharge with pus and bloody component over the left ear, and generalized