Multiple Cranial Neuropathy in Cogan's Syndrome

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Abstract

A 20 year old woman presented with recurrent alternative keratitis for four months. One month before admission, she developed progressive hearing loss, visual impairment, facial diparesis and bilateral trigeminal neuropathy. Cogan's syndrome was diagnosed. Prompt treatment with corticosteroid resulted in dramatic improvement of the ocular, otological and neurological dysfunctions.

Key word : Cogan's Syndrome, Cranial Neuropathy, Interstitial Keratitis

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Cogan's syndrome is an uncommon disease of young adults. It is characterized by nonsyphilitic interstitial keratitis and vestibuloauditory dysfunction. Unilateral or bilateral interstitial keratitis with photophobia and lacrimation are the common findings[1,2]. They may precede the audiovestibular symptoms for months. Systemic manifestations such as systemic vasculitis, aortitis, lymphadenopathy, gastrointestinal hemorrhage, musculoskeletal and neurological disorders have been reported[1]. Neurological symptoms and signs including head and neck pain, peripheral neuropathy, cranial neuropathy and mental changes have been associated with Cogan's syndrome[3]. However, to our knowledge, there have been only a few cases of this syndrome with cranial neuropathy. We report a patient with Cogan's syndrome who had facial and trigeminal nerves dysfunction. The multiple cranial neuropathy as well as the auditory and visual symptoms dramatically improved after treatment with high dosage of corticosteroid.

CASE REPORT

ND is a 20-year-old female who presented to the hospital in January 1997. She had been well until four months before admission when she experienced ocular pain, severe photophobia and redness of the left eye. She also noticed a low grade fever and blurring of vision on the left. The symptoms persisted for 4-5 days and gradually subsided. One month later, she again developed ocular pain and photophobia in the left eye which lasted about 2 weeks. After the symptoms in the left eye had resolved, severe photophobia occurred in the

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right eye. She also experienced progressive blurring of vision in both eyes which she described as looking through a white tinted glass. A week later, she developed intermittent pain and tightness in both ears. Hearing was gradually decreased. Two weeks thereafter, she experienced numbness of the face which was more on the right side. The visual loss progressed and she was unable to see with her right eye. One week before admission, she developed bilateral facial weakness.

On examination, she was a healthy looking young female with severe photophobia. Her blood pressure was 120/80 mmHg and the pulse rate was 72/min. She was afebrile. Ophthalmological examination revealed bilateral numular keratitis more on the right side. Mild conjunctival injection was also noted. There was no evidence of anterior or posterior uveitis. The pupils were round, 4 mm in size, with good pupillary light reaction. The eye grounds were normal. Visual acuity was limited to the recognition of hand movements on the left and only light perception on the right. The corneal reflexes were absent bilaterally. Pinprick sensation was decreased in the distribution of all divisions of the trigeminal nerve on both sides. The patient had bilateral lower motor neuron facial weakness which was slightly more prominent on the right. Decreased hearing of both ears was also noted. There was no tongue deviation. Her motor power was full. The deep tendon reflexes were 2+ in the upper extremities, 2+ in both knees and 1+ in both ankles.

Laboratory evaluation revealed a hemoglobin 12.8 g/dl, platelets $306x10^9$/l, white blood cells $9.7x10^9$/l with 69 per cent neutrophils and 19 per cent lymphocytes, erythrocyte sedimentation rate (ESR) 60 mm/h and a non-reactive VDRL. A test for human immunodeficiency virus (HIV) antibody was negative. Lumbar puncture examination disclosed an opening pressure of 10 cm CSF. The CSF was clear with no cells. CSF protein was 4.8 g/l and the CSF glucose was 5.7 mmol/l (plasma glucose 7.6 mmol/l). The antinuclear antibody was negative. Cogan's syndrome was diagnosed on the basis of the interstitial keratitis and vestibuloauditory dysfunction in the absence of syphilis.

She was treated with steroid eyedrop and oral prednisolone 60 mg/day. After 5 days of treatment, the photophobia gradually improved and the keratitis became less pronounced. The visual acuity was 20/40 on the left and 20/100 on the right. Audiogram on the 6th day after treatment revealed normal hearing of both ears. The facial weakness and trigeminal neuropathy were also improved. Only mild right facial weakness was noted after 2 weeks.

DISCUSSION

Cogan's syndrome is characterized by interstitial keratitis and aural involvement. It was first reported by Morgan and Baumgartner in 1934 as a syndrome of nonsyphilitic interstitial keratitis associated with audiovestibular dysfunction[4]. However, the syndrome was named after David Cogan who classified this syndrome as a distinct entity in 1945[5]. The disease usually occurs in young adults in their third decade with a median age at onset of 25 (range 4 to 63); 80 per cent of patients are between the ages of 14 and 47[1]. The etiology of Cogan's syndrome is still not known, although its association with vasculitis may suggest an autoimmune basis. Preceding upper respiratory infection has been reported in about half of the cases[1,2]. This may suggest viral or postviral etiologies, but no causative agent has been identified[6]. Some other infectious etiologies have been postulated including spirochetal infection and Borrelia burgdorferi but no association was found[7]. Interstitial keratitis, manifests by intense photophobia, lacrimation and eye pain, is the most common presenting symptom. Relapse and remission is common but severe visual impairment is rare. Ear involvement typically begins unilaterally with Meneire like attacks of tinnitus and vertigo accompany with nausea and vomiting and progressive hearing loss. Our patient had recurrent alternative classical ophthalmological manifestation of Cogan's syndrome and hearing impairment. Neurosyphilis was excluded by a negative serological study. Laboratory investigations in Cogan's syndrome usually reveal a raised erythrocyte sedimentation rate and leukocytosis[1]. Complement levels, antinuclear antibody, rheumatoid factor, serum protein and immunoelectrophoresis are normal[2]. Circulating immune complexes have not been found[2]. Abnormal cerebrospinal fluid studies are found in one-fourth of the cases. In most of the cases, only a few mononuclear cells are present[1]. Electroencephalographic studies (EEG) are abnormal in half of the patients with Cogan's syndrome. Generalized, low-voltage, slow waves are the common findings[1].

Various neurological complications of Cogan syndrome have been reported. These include

meningoencephalitis, cerebrovascular syndrome, venous sinus thrombosis, head and neck pain and mononeuropathy[3]. However, there are very few reports about the cranial nerves involvement in this disease and most of them have other evidence of systemic vasculitis suggesting polyarteritis nodosa[3,6,8,9]. In 1953, Oliner et al reported a 23 year old woman who presented with nonsyphilitic interstitial keratitis and bilateral deafness. During the course of her illness, she developed bilateral ptosis and right inferior oblique paresis in association with fever, generalized myalgia, lymphadenopathy, splenomegaly and purpuric cutaneous nodules. Lymph node and skin biopsy revealed polyarteritis nodosa[8]. Crawford WJ reported a 38 year old man who had headache and trigeminal neuralgia. The patient also had fever, significant weight loss, arthralgia and lymphadenopathy. Muscle biopsy revealed necrotizing arteritis compatible with polyarteritis nodosa[9]. A case of questionable facial weakness associated with Cogan's syndrome was reported by Eisenstein B et al[10]. A 38 year old woman who developed a questionable right facial weakness with right sided hypalgesia in association with progressive hearing loss and low grade fever. Two years later, aortic insufficiency developed, leading to intractable heart failure and death. In our case, multiple cranial neuropathy manifested by bilateral facial nerve and trigeminal nerve paresis were the early prominent symptoms. There was no clinical or laboratory evidence of systemic vasculitis.

The response to treatment in Cogan's syndrome varies with the target organ involvement and duration of disease before starting the treatment. Ocular manifestations usually respond well to corticosteroid. Vestibular component of the vestibuloauditory dysfunction is generally reversible while hearing loss is progressive in most cases. However, early treatment may prevent deafness. The clinical course of cranial neuropathy is not well defined due to a small number of cases. From our experience in this case, early treatment with high dosage of corticosteroid resulted in improvement of the ocular, otologic and neurological manifestations.

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ความผิดปกติของเส้นประสาทสมองในกลุ่มอาการโคแกน

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ผู้ป่วยหญิงอายุ 20 ปีมาด้วยอาการปวดตาและตาแดงสลับข้าง เป็น ๆ หาย ๆ มา 4 เดือน 1 เดือนก่อนมาโรงพยาบาลเริ่มรู้สึกว่าหูทั้ง 2 ข้างได้ยินไม่ชัด อาการเป็นมากขึ้นเรื่อย ๆ ร่วมกับมีตามัวทั้ง 2 ข้าง ใบหน้าทั้ง 2 ซีกขยับไม่ได้เนื่องจากเส้นประสาทสมองเส้นที่ 7 เป็นอัมพาต และสูญเสียการรับความรู้สึกที่ใบหน้าจากเส้นประสาทสมองเส้นที่ 5 เสีย จากอาการทางตาที่ตรวจพบว่ามีการอักเสบที่กระจกตาและการมีความผิดปกติทางหู ทำให้ได้รับการวินิจฉัยเป็น กลุ่มอาการโคแกน ผู้ป่วยรายนี้ได้รับการรักษาด้วยสเตอรอยด์พบว่าอาการทั้งหมดดีขึ้นอย่างรวดเร็ว

คำสำคัญ : กลุ่มอาการโคแกน, เส้นประสาทสมอง, กระจกตาอักเสบ

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