

Otitis complicated by Jacod's syndrome with unusual facial nerve involvement: Case report and review of literature

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Abstract

Otitis media is a well-known condition and its infra-temporal and intracranial complications are extremely rare because of the widespread usage of antibiotic treatment. We report a case of 63-year-old female with complaints of right-sided facial pain and diplopia. She had a history of acute otitis media before 4 months of admission to our neurology unit. Neurological examination showed that total ophthalmoplegia with ptosis, mydriasis, decreased vision and loss of pupil reflex on the right side. In addition, there was involvement of 5th and 7th cranial nerves. Neurological and radiological follow-up examinations demonstrated Jacod's Syndrome with unusual facial nerve damage and infection in aetiology. Sinusitis is the most common aetiology, but there are a few cases reported Jacod's Syndrome originating from otitis media.

Keywords: Orbital apex, Petrous apex, Otitis, Cranial nerve, Facial nerve, Infection.

Introduction

Otitis media (OM) is a well-known condition and its infratemporal and intracranial complications are extremely rare because of the widespread usage of antibiotic treatment. In some OM cases, the infectious and inflammatory disease can spread by following the route from apex petrositis to cavernous sinus and then other structures in the vicinity of the peri or retroorbital area.¹ Orbital canal, superior orbital fissure and cavernous sinuses are located in the retroorbital area and anatomically related to each other. The infectious or inflammatory processes can spread from one area to another and incidents that affect these areas may cause similar clinical findings. These overlapping signs may cause difficulty in diagnosis. Tumours are the most common aetiology in younger and inflammatory and infectious aetiologies are the most common ones in older patients with JS.²⁻⁴ We present a rare case of Jacod's Syndrome (JS) following OM and sinusitis.

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Case Report

A 63-year-old female was diagnosed as acute OM before 4 months of admission to our neurology unit in March 2013. Despite antibiotic treatment, her complaints progressed. She was hospitalised with a diagnosis of orbital cellulitis. She had been treated with Ceftazidime 3g/day and Vancomycin 1000 mg/day for 15 days. Cranial and orbital magnetic resonance imaging (MRI) studies performed during those days revealed a high T2 signal intensity in the right orbital area and contrast enhancement within periorbital region with maxillary and sphenoidal sinusitis. Temporal bone computed tomography (CT) examination findings were consistent with mastoiditis on the right side. On the 9th day of antibiotic treatment, right-sided facial paralysis developed. After neurology consultation, prednisolone 500mg/day was given. After regression of the periorbital cellulitis, she was referred to neurology clinic with preliminary diagnosis of cavernous sinus syndrome. Venography was normal. The patient had a history of longstanding poorly-controlled diabetes, hypertension and hyperlipidaemia. Her neurological examination



Figure-1: Contrast-enhanced fat-saturated axial T1-weighted image shows exophthalmos and massively enhancing pre and postseptal orbital structures with scleritis in the right orbit, suggestive of orbital cellulitis. Patient can easily present with orbital apex syndrome since inflammatory changes are more prominent at the orbital apex.

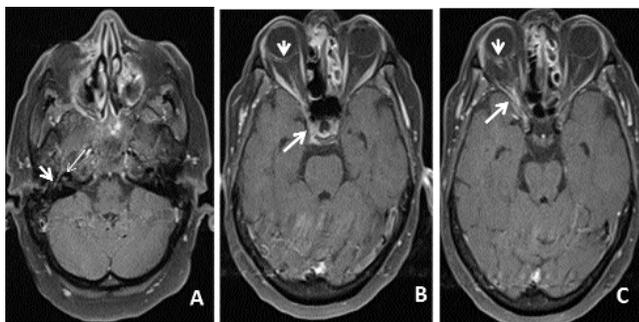


Figure-2: In a patient following the treatment of orbital cellulitis and petrous apicitis, peripheral facial paralysis, labyrinthitis and orbital apex syndrome with optic neuropathy are still present on the right side. On contrast-enhanced fat-saturated axial T1-weighted images (A, B, C) show that tympanic segment of 7th nerve (short arrow on A) and cochlea (thin arrow on A) are asymmetrically enhancing on the right compared to their counterparts. Right orbital inflammation with exophthalmus has greatly resolved although scleral and periscleral inflammation (short arrows on B, C), orbital apex infiltration (long arrow on C), asymmetric cavernous sinus enhancement (long arrow on B) are still going on. Note that superior orbital fissure anatomically continues with the cavernous sinus itself.

revealed right-sided complete ophthalmoplegia with mydriasis, absence of corneal sensory and pupil reflex on the right eye. She also had vision loss, ptosis and peripheral typed facial paralysis on the right side. There was hypoesthesia along the distribution of the first and second divisions of the right trigeminal nerve. The patient was put on 60mg of oral prednisolone treatment for 1 month and then it was stopped.

Two months after discharge, her complaints were still present and she was referred to our hospital with an increase of her facial pain. Neurological examination showed similar findings as reported above. Total blood count, erythrocyte sedimentation rate (ESR), serum electrolytes and fasting glucose level, C-reactive protein (CRP), antinuclear antibody, angiotensin-converting enzyme (ACE), cytoplasmic antineutrophil cytoplasmic antibodies (C-ANCA), perinuclear antineutrophil cytoplasmic antibodies (P-ANCA), serology for human immunodeficiency virus (HIV) and treponema pallidum results were normal. Cranial MRI, neurography and venography examinations were evaluated. Neuroradiodiagnostic examinations showed that orbital cellulitis had greatly reduced into inflammation in the orbital apex, superior orbital fissure and cavernous sinus on the right side (Figure-1, 2). Tympanic segment of 7th nerve (short arrow on 2A) and cochlea (thin arrow on 2A) were asymmetrically enhancing on the right compared to their counterparts. The diagnosis of JS and peripheral 7th nerve palsy was made. Because of actively contrast enhancement, Prednisolone (P.O.) 80mg/day was added to the treatment. During follow-up period of 2 months,

her facial pain, her facial paralysis, and her eye movements especially related to the 3rd cranial nerve improved partially.

Discussion

The mastoiditis, labyrinthitis, facial nerve palsy and petrous apicitis are infratemporal complications of acute OM. Infectious process can spread to the intracranial compartment ending up with meningitis, intracranial abscess, venous sinus thrombosis and death when the infection is not treated effectively.¹ The complications in adult cases are rare and most of them have an underlying immune suppression such as diabetes, renal failure, malignancies, positive serology for HIV, long-term usage of immuno-suppressant agents.²⁻⁹ In our patient, mastoiditis and facial nerve involvement were present as infratemporal complications of acute OM. She was an old-aged woman and had diabetes. OM may cause infections of cavernous sinus, but the infections of cavernous sinuses, superior orbital fissure or orbital apex are usually originated from para-nasal sinuses.⁴ Time interval between the onset of OM and cranial nerve involvements is usually between 1 week to 2-3 months.⁵ Similarly, cavernous sinus problem followed by sinusitis were diagnosed in our patients with JS and time interval between the onset of OM infection and JS was just 2 months.

Third, 4th, 5th and 6th cranial nerve problems are seen in not only JS but also superior orbital fissure syndrome (also known as Rochon-Duvignaud Syndrome) and cavernous sinus syndrome.^{3,6} The most common complaints of these three syndromes are retroorbital pain and all are characterised by ophthalmoplegia and ptosis.^{3,6} Cavernous sinuses contain internal carotid artery and its sympathetic plexus, so orbital congestion and proptosis are seen in cases with cavernous sinus syndrome. Maxillary branch of trigeminal nerve and sympathetic plexus involvements are important for differential diagnosis.^{3,6} In JS, the patients complain of visual loss in addition to clinical findings related to superior orbital fissure involvement. This syndrome is also commonly associated with a tumour. As mentioned above, retroorbital pain, ophthalmoplegia, ptosis, decreased corneal sensation and visual loss are seen. Optic neuropathy may be seen and optic atrophy may develop over weeks to months.^{3,6} Petrous apicitis (Petrous apex syndrome) is another and rare complication of OM. The petrous part of the temporal bone is pyramidal in shape and the apex has a neighbourhood with clivus and cavernous sinus.^{7,8} The spread of the infection can also occur by direct extensions via facial planes, vascular channels and through bone.^{2,7} In cases with petrous apex syndrome, facial nerve paralysis is

common and also other cranial nerve (II-X) deficits may occur because of the inflammatory process and oedema when the infection spreads to the skull base or cavernous sinus.^{2,7,8} While managing these syndromes, including JS, the target is underlying cause. Differential diagnosis between inflammatory, infectious and infiltrative aetiologies is difficult and they may all be responsive to corticosteroids.³ Radiotherapy and immuno-suppressant agents such as methotrexate or azathioprine is useful in selected patients.³

JS consists of signs of involvement of all the neural structures traversing the optic foramen and the superior orbital fissure. Thus, signs of optic neuropathy coexist with other cranial nerve symptoms.^{9,10} Visual loss secondary to 2nd nerve involvement was the main point in differential diagnosis of JS. Considering infectious aetiologies, sinusitis is the most common one but there are a few cases reported JS originating from OM.^{3,4} Our patient had both OM and sinusitis. After parenteral antibiotic therapy and oral methylprednisolone therapy within 6 months, her facial pain, vision, eye adduction and facial paralysis partially improved.

Conclusion

Our case is the first case revealing JS with facial nerve involvement. The present case shows that the infectious

processes causing JS may not be bordered and may spread into nearby structures such as cavernous sinus and petrous apex.

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