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CASE REPORT OF GRADENIGO SYNDROME

Otorhinolaryngology

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ABSTRACT

Background: Back in 1900's, Gradenigo reported a series of patients with suppurative otitis media presenting with headache, unilateral eye pain and restricted lateral gaze of ipsilateeral side. Chronic ear infections spreading to temporal bone extends to the petrous apex which is the hub for 5th and 6th cranial nerves.**Case:** We present one such case of a 19 year old male with history of chronic otitis media who presented with acute onset of retro-orbital pain, headache and diplopia.**Conclusion:** patient with chronic ear discharge who present with cranial nerve involvement, one must suspect intracranial extension. Management with appropriate antibiotics can change the course of the condition.

KEYWORDS

otitis media, Gradenigo syndrome, cranial nerves, abducens nerve, trigeminal nerve

INTRODUCTION

The trigeminal ganglion lies close to the petrous apex in the Meckel's cave and is separated only by Dura mater. Adjacent and medial to the trigeminal ganglion lies the abducens nerve in the Dorello's canal. In patients with suppurative otitis media, infection may spread to the petrous apex of the temporal bone, via pneumatised air cell tracts, or vascular channels, or as a result of direct extension through fascial planes giving rise to apical petrositis. This results in a classical triad of symptoms consisting of otitis media, VIth nerve palsy and retro-orbital pain (area supplied by the ophthalmic branch of the trigeminal nerve), collectively known as Gradenigo's syndrome [1,3].

This classical triad was first described in 1900's by Guisseppe Gradenigo [1, 2]. Since then many atypical presentations of Gradenigo syndrome have been reported. However presence of apical petrositis is common in all. Neuroimaging modalities i.e. Computed tomography scan (CT scan) and Magnetic Resonance Imaging (MRI), are used to diagnose this condition.

Previously, surgical management was mainstay of this condition. However, with advent of improved antibiotics, conservative management with appropriate antibiotic coverage is being employed in selected cases [4].

Case Report

Case History

A 19 year old male patient presented to OPD of Department of ENT, Deccan College of Medical Sciences, Hyderabad, with complaints of left ear discharge, diplopia, left eye pain and headache of 4 days duration. The headache was holocranial but predominantly in frontal region and along left ear area. Patient was also complaining of left eye retro-orbital pain, double vision with both eyes and blurring of vision with left eye. The retro-orbital pain was aggravated with eye movements.

1 month ago, patient had presented to the OPD with complaints of left ear discharge of 3 months duration. He was also experiencing recurrent upper respiratory tract infections. Ct scan of temporal bones and paranasal sinuses was taken which had revealed fluid in the left middle ear cavity, mastoid antrum and air cells suggestive of oto-mastoiditis and an 'S' shaped nasal septum with bony spur on the right side of the septum. In lieu of deviated nasal septum and chronic sinusitis, patient underwent mastoid exploration and functional endoscopic sinus surgery (FESS) along with septoplasty and was planned for tympanoplasty of left ear after 2 months.

Examination

On general examination, the patient's vital signs were normal. His higher mental functions were normal. On cranial nerves examination, patient had restricted abduction of left eye which was suggestive of left abducens nerve palsy. All other extra ocular movements were normal. The rest of CNS examination was normal. His visual acuity was 6/6 in both eyes with normal color perception. Pupils were equally reactive to light bilaterally and fundus examination was normal. On otoscopic examination of the left ear, two polypoidal masses were seen in the roof of external auditory canal along with purulent discharge. The tympanic membrane couldn't be visualized due to polypoidal masses. Right side external auditory canal and tympanic membrane were normal. The rest of the systemic examination was unremarkable.

Investigations

All routine baseline investigations were normal. MRI of brain (plain and contrast) was done which revealed fluid signal completely filling the left middle ear cavity and mastoid space, along with expansion of the fluid signal in left sided petrous apex. Adjacent enhancing granulation tissue was present on the medial side of petrous apex in the cerebello - pontine angle cistern. Marrow edema was noted in the left half of the clivus and base of the skull. Post contrast imaging reveals prominent peripheral enhancement of the fluid in the petrous apex, along with enhancement in the left middle cavity and mastoid cells. Involvement of the left fifth and sixth nerves due to petrous apex pathology was noted. There was no evidence of lepto-meningeal or cerebral parenchyma involvement. These radiological findings were suggestive of petrous apicitis.

Our patient's left petrous apex enhancement on MRI, coupled with his sub-acute presentation of left temporal headache, diplopia and left eye pain, 6th cranial nerve involvement in form of inability to abduct left eye, along with history of CSOM led to the diagnosis of Gradenigo syndrome.

Treatment

The polypoidal masses in left external auditory canal were excised and the ear was packed. In lieu of petrous apicitis, patient was started on intravenous ceftriaxone 1 g twice a day and clindamycin 600 mg twice daily. Headache subsided by 3rd day and restricted eye movement improved by 10th day of treatment. Patient was advised to undergo mastoidectomy after infection subsided.

DISCUSSION

Chronic suppurative otitis media is associated with many intracranial complications of which Gradenigo's syndrome is one such complication which is very rare. It was first recognized by Gradenigo as a condition presenting with unilateral facial pain, unilateral lateral gaze palsy and otorrhea. Due to the intricate anatomical architecture of intracranial structures, this syndrome has the classic triad of petrous apicitis has fallen out of picture. The present given literature has a spectrum of atypical presentations. However, the patient reported in this study has the presented with classical triad of features.

Petrous apicitis is mostly caused due to spread of ear infection to temporal bone and ultimately reaching the petrous apex. Other causes of Gradenigo's syndrome that have been reported are neoplasms like nasopharyngeal carcinoma, metastatic non-Hodgkin lymphoma; infections like tuberculosis. Irrespective of etiology, the common pathology is the presence of petrous apicitis and involvement of 5th and 6th cranial nerves [5,8].

Treating the patient with appropriate antibiotic which can penetrate blood brain barrier can change the course of this almost reversible condition. Nonetheless, surgery is the mainstay of treatment. Management of apical petrositis should include permanent drainage and ventilation of the apical cells while also attempting to preserve hearing. This goal can be achieved through a trans mastoid infralabyrinthine suprajugular approach, depending on the anatomy of the region.

Fig 1: MRI brain

Fig 2: Lateral gaze palsy of left eye

Scott *et al*. [9] reported a diabetic patient with chronic ear discharge and cranial nerve involvement who on CT imaging was found to have petrous apicitis. The patient reported by them had made remarkable recovery with appropriate antibiotic therapy.

Safia *et al*. [3] had reported an atypical presentation of a 55-year-old male who presented with unilateral headache, dysphagia and hoarseness (IX and X cranial nerve involvement), and diplopia with lateral gaze palsy (VI nerve involvement) in the absence of trigeminal neuralgia or a history of otitis media. The patient had made complete recovery after intravenous antibiotics.

Parekh, *et al*. [10] reported a 71 year old male diabetic with candida mastoiditis who presented with initially with 6th and partial 3rd nerve palsy, later developed Vernet syndrome progressive dysphagia, dysarthria (9th, 10th and 11th cranial nerve paralysis), right otalgia, and hearing loss. This patient was managed by surgical debridement and a mastoid biopsy, cultures of which had revealed *Candida parapsilosis*. He was started on antibiotics and antifungals, but died after few weeks.

CONCLUSION

Although rare, petrous apicitis should be considered in the differential diagnosis of any patient with chronic ear drainage or pain that is not responding to conventional treatment, or in anyone with a cranial nerve dysfunction in the setting of acute or chronic otitis media, so as to avoid life threatening complications.

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