UNUSUAL CASE- REVISITED- GRADENIGO'S SYNDROME

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HOW TO CITE THIS ARTICLE: Mani S, Rekha A. Unusual case- revisited- Gradenigo's syndrome. J. Evolution Med. Dent. Sci. 2017;6(73):5274-5275, DOI: 10.14260/Jemds/2017/1144

PRESENTATION OF CASE

A 25-year-old male presented with 5 days history of double vision and a 4 days history of ear discharge and hearing loss. Patient was apparently normal 5 days ago after which he developed unilateral headache, throbbing type of pain localises to right frontal and retro-orbital area associated with right ear discharge and hard of hearing. On examination, patient had right lateral rectus palsy. (Figure-1) Otoscopic examination revealed a postero-superior perforation in the right ear tympanic membrane. Clinical findings were compatible with the classic triad of Gradenigo's syndrome. In pure tone audiogram, there was mild conductive type of hearing loss in right ear. Magnetic Resonance Imaging revealed hyperintensity in T1 axial flair and T2 weighted images in both mastoid air cells and petrous apex involving Dorello's canal and Meckel's cave area on right side. (Figure-2) Gram stained revealed few pus cells and gram-positive cocci in pair, but no growth was found in culture.

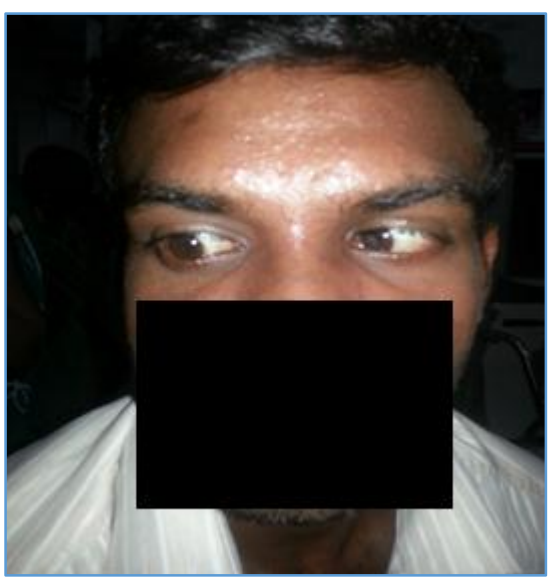

Figure 1. Right Lateral Rectus Palsy

Financial or Other, Competing Interest: None.
Submission 21-06-2017, Peer Review 25-08-2017,
Acceptance 01-09-2017, Published 11-09-2017.
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DOI: 10.14260/jemds/2017/1144

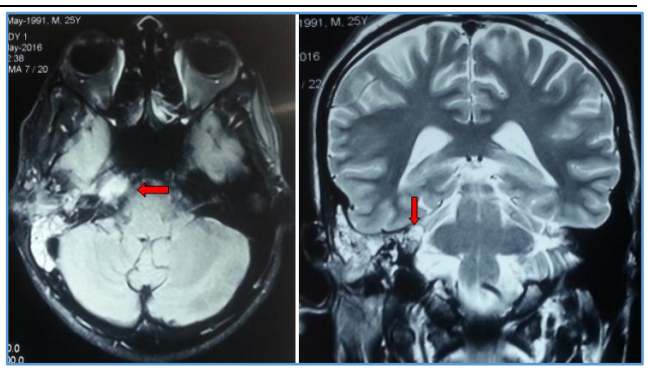

Figure 2. MRI T1 Axial showing Petrous Apicitis (MRI T2 Coronal showing the Petrous Apicitis)

Figure 3. Post-Operative

Pre-Operative Post-Operative

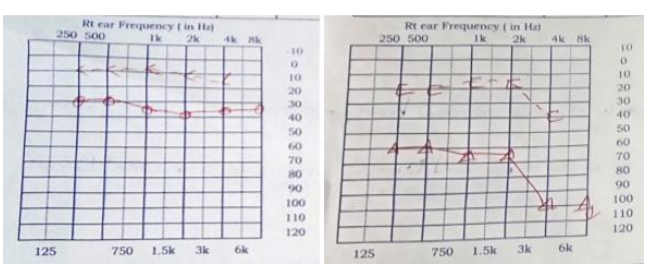

Figure 4. Pure Tone Audiometry

DIFFERENTIAL DIAGNOSIS

- Cerebellopontine angle tumour.
- Petrous apex lesion- Cholesterol granuloma, Cholesteatoma, Meningioma, Metastatic carcinoma, Arachnoid cysts, Chondroma, Chondrosarcoma and Chordoma.

CLINICAL DIAGNOSIS

Gradenigo's syndrome.

PATHOLOGICAL DISCUSSION

Gradenigo's syndrome is characterised by a classic triad of discharging ear, retro-orbital pain, abducens nerve paralysis causing diplopia. Gradenigo's syndrome, also called Gradenigo-Lannois syndrome and petrous apicitis, is a complication of otitis media and mastoiditis involving the apex

of the petrous temporal bone.[1] Infection and inflammation of petrous apex involves 6th cranial nerve at the Dorello's canal and 5th cranial nerve in the Meckel's cave. Retro-orbital pain is caused due to the involvement of trigeminal ganglion (Gessarian ganglion) at the level of Meckel's cave.[2] Abducens nerve passes through Dorello's canal to enter the middle cranial fossa.

The relationship between the sixth nerve, Gruber's ligament, the carotid artery and the anterior clinoid process, is necessary to understand the three dimensional structure of the canal.[3]

CT and MRI scans are useful in identifying the lesions around the petrous apex which can differentiate the other possible diagnosis, also tells about the mastoid pneumatisation. So radiological evaluation plays a key role in the diagnosis and management of Gradenigo's syndrome.[4]

Acute otitis media is a common disease in our country, complications are rare, but should be suspected always in association with abnormal symptoms. Awareness of the condition coupled with prompt investigations is required for early recognition of complications, which can prevent further morbidity and mortality.

Gradenigo's syndrome is rarely seen after the introduction of broad spectrum antibiotics and wide availability of health care system. Still it is a potentially fatal complication secondary to acute otitis media and acute mastoiditis because of intracranial spread of infections.[5] This syndrome is most commonly seen with chronic suppurative otitis media in adult population and in children with acute otitis media. On the contrary our case was an adult, having acute otitis media presented with petrous apicitis.[6,7]

The delay between otologic symptoms and cranial nerve involvement varies from 1 week to 3 months.[7] In our case, the time between the onset of the initial symptoms and of the abducens palsy was less than a week. As we see most of the Gradenigo's syndrome cases usually present with diplopia and ear discharge, patient usually will give importance for vision and goes to ophthalmologist first rather than otorhinolaryngologist, that is where the treatment delays, so it is the duty of the ophthalmologist who should immediately refer the case without delay to prevent complications.

Microbiological studies of Gradenigo's syndrome are difficult to perform and cultures are often negative. Most common organisms include Staphylococcus species, Pseudomonas aeruginosa, Streptococcus species and Mycobacterium tuberculosis.[8] In our case also, organism does not grow in the culture but well responded to ceftriaxone therapy. The organism could be a streptococcal species by considering factors like gram stain, which showed gram positive cocci in pairs, acute otitis media and its response to ceftriaxone. Both CT and MRI plays an important role in identifying the disease process. MRI gives information concerning inflammatory changes in the petrous apices and plays a vital role in ruling out the other causes of Gradenigo's syndrome, which includes tumours at the petrous apex and other rare causes such as traumatic intracranial aneurysm.[9]

DISCUSSION OF MANAGEMENT

Patient was started on Intravenous Ceftriaxone 1 G twice a day and taken up for mastoid exploration. Patient underwent Extended Cortical Mastoidectomy with exenteration of retrolabyrinth air cells. Patient had drastic improvement after surgery and reported complete resolution of his retro-orbital pain, headache and diplopia within 5 days of surgery (Figure-3). Pure Tone Audiogram was performed, which showed conductive hearing loss (Figure-4).

Gradenigo's syndrome has been mostly treated by radical surgery, but there are some reports managing conservatively. Burston et al reported two cases of Gradenigo's syndrome, which were managed without the need for radical surgery.[6] Marianowski et al reported a case of a 6-year-old child, which was totally cured by myringotomy and antibiotic therapy. Rest of the cases were treated surgically.[7] Most of the surgical approaches leads to permanent hearing loss, because of sclerotic mastoids in chronic otitis media. In acute disease with well pneumatised bone gives us an easy approach to petrous apex without disturbing the bony labyrinth. We approached with less aggressive surgery in view to preserve the hearing, but it may not be applicable in all cases.

FINAL DIAGNOSIS

Gradenigo's syndrome is a rare complication of acute otitis media. It should always be suspected in the presence of unilateral headache and diplopia (Abducens nerve palsy) with or without ear discharge. Awareness of the condition coupled with prompt investigations is required for early recognition. Choice of Antibiotics should cross the blood brain barrier to prevent complications secondary to petrous apicitis. Hearing preservation surgeries totally depends on pneumatisation of temporal bone.

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