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Vocal Cord Paralysis and Dysphagia as Sequelae of Gradenigo Syndrome

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ABSTRACT

Objectives: To present a case of vocal cord paralysis and dysphagia developing in Gradenigo syndrome and to discuss its clinical presentation, differential diagnosis and therapeutic approach.

Methods:

Design: Case Report
Setting: Tertiary Government Hospital
Patient: One

Results: A 54-year-old lady was admitted with a six-month history of left-sided otorrhea, cheek and jaw pain, three months otalgia, and recent-onset hoarseness, dysphagia and diplopia on a background of mastoidectomy at age six. Otoscopy revealed granulation tissue and chlolesteatoma occupying the left external auditory canal. There was left vocal cord paralysis with pooling of saliva in the pyriform sinus, left lateral gaze paralysis and left facial nerve paralysis. CT scan revealed sclerosis of the left petrous apex and leptomeningeal enhancement on the left temporal lobe. Chronic suppurative otitis media with cholesteatoma and Gradenigo syndrome was diagnosed, and canal wall down mastoidectomy was performed. Postoperatively, the otalgia and pain over the left jaw diminished in intensity while hoarseness and left lateral gaze palsy remained.

Conclusion: Gradenigo syndrome is known for its triad of retro-orbital pain, lateral gaze paralysis, and chronic middle ear infection due to petrous apicitis. Although rare, vocal cord paralysis and dysphagia may develop when infection traverses and encroaches on the jugular foramen where cranial nerves IX, X, and XI are lodged. Knowledge of the syndrome should not be limited or confined to the classic triad. Practicing ear specialists should be vigilant and cognizant of the clinical manifestations and sequelae of chronic middle ear infection. Prompt surgical intervention is crucial while resolution of the disease may vary for different individuals.

Keywords: *chronic otitis media, Gradenigo syndrome, vocal cord paralysis, petrous apicitis*

Dealing with chronic suppurative otitis media requires utmost attention and care because of the clever and sometimes elusive nature of the disease. Life threatening complications include meningitis, brain abscess, lateral sinus thrombophlebitis and petrous apicitis which may manifest with Gradenigo's syndrome.

A recent Grand Rounds discussion of Gradenigo syndrome in a 17-year-old patient with nuchal rigidity and anisocoria was reported in this journal.[1] The literature has several reports about the syndrome manifesting with the classic triad[1-10] but vocal cord paralysis and dysphagia are not usually mentioned sequelae.[2]

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CASE REPORT

A 54-year-old lady from Pampanga was admitted because of diplopia. Six months prior, she experienced left ear discharge which was treated with oral antibiotics and otic drops. Three months later, she experienced left jaw and left eye pain radiating to the left frontal and occipital areas. She was diagnosed with trigeminal neuralgia but steroid injection in the left lower alveolar area afforded no relief. Two weeks before admission, she developed hoarseness, difficulty of swallowing (especially liquids) and doubling of vision prompting this consult.

Review of history disclosed that a left facial palsy developed after mastoidectomy at age six, with occasional ear problems thereafter. Otoscopy showed a dry 30% central tympanic membrane (TM) perforation on the right while no tympanic membrane was appreciated on the left because of granulation tissue and cholesteatoma occupying the left mastoidectomy cavity. *(Figure 1)* Videolaryngoscopy revealed left vocal cord paramedian paralysis with pooling of saliva in the left pyriform sinus. *(Figure 2)* There was lateral gaze palsy of the left eye *(Figure 3)* with left facial nerve paralysis House-Brackmann Grade V. Topognostic exam showed loss of taste on the anterior 2/3 of the left side of the tongue and decreased lacrimation of the left eye on Schirmer's test suggesting pathology in the mastoid segment of the facial nerve.

Plain and contrast cranial and temporal bone CT scans showed evidence of previous left mastoidectomy with mastoiditis, cholesteatoma formation, abnormal enhancement of the meninges in the left temporal region and sclerosis of the left petrous apex. *(Figures 4 and 5)* Pure tone audiometry revealed profound hearing loss on the left and moderate mixed hearing loss on the right.

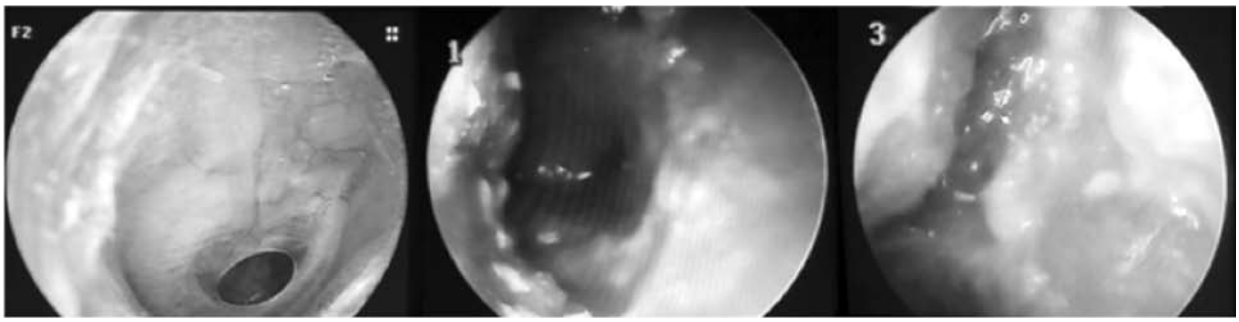

Figure 1. (Leftmost photo): right tympanic membrane with 30% central perforation, (middle photo) left ear granulation tissue and (right photo) cholesteatoma occupying the middle ear.

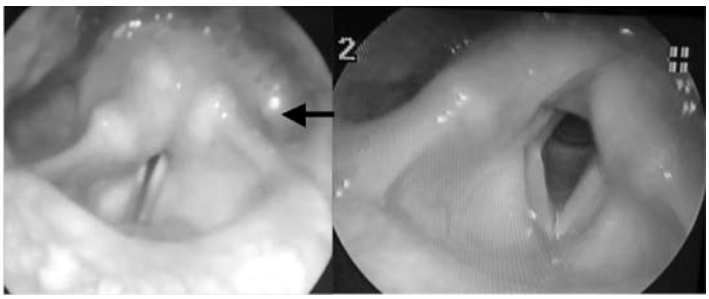

Figure 2. Left vocal cord paralyzed in paramedian position with pooling of saliva in the left pyriform sinus (left photo) in phonation, (right photo) in inspiration.

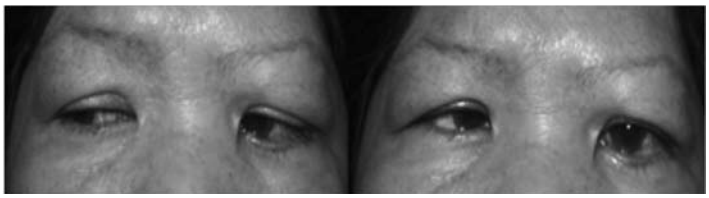

Figure 3. Extraocular muscle movements, (left photo) on right gaze, and (right photo) on (L) gaze, showing left lateral gaze palsy of the left eye.

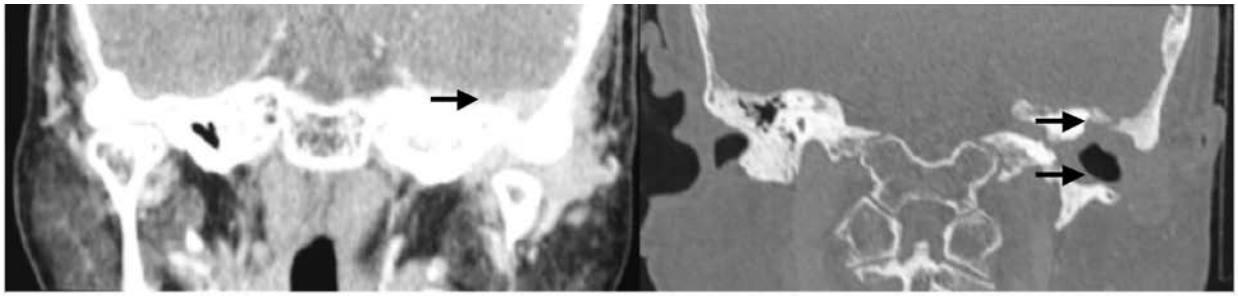

Figure 4. CT scan showing (left) moderate enhancement of the meninges in the left temporal lobe (arrow) adjacent to (right) the cortical defect in tegmen tympani (upper arrow) and mastoidectomy defect in the left mastoid with soft tissue density representing cholesteatoma and granulation tissue (lower arrow).

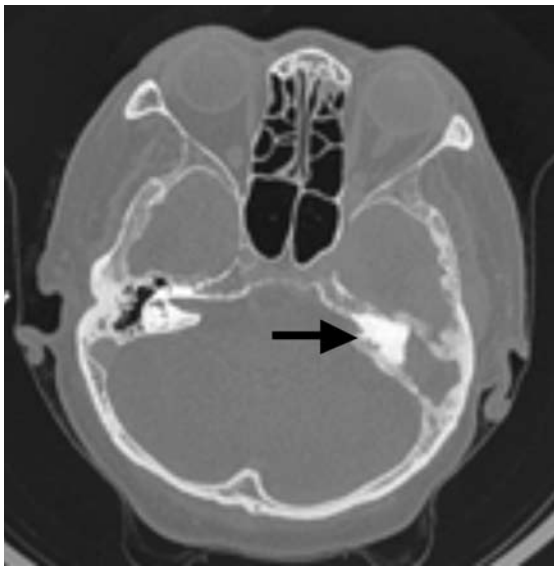

Figure 5. CT scan axial bone window, showing sclerotic left petrous apex.

The clinical impression was post-mastoidectomy recurrent chronic suppurative otitis media with cholesteatoma formation and Gradenigo syndrome. Intravenous ceftriaxone was started and a canal wall-down mastoidectomy with translabyrinthine approach to the petrous bone was commenced but difficulty drilling the sclerotic bony labyrinth hindered access to the petrous apex. Postoperatively, the left jaw, cheek and retro-orbital pain diminished within 24 hours but the hoarseness and lateral gaze palsy remained.

DISCUSSION

Although rarely encountered, complications of chronic suppurative otitis media continue to develop, perhaps because of neglect, complacency, poor patient education or inadequate treatment. Dreaded intracranial complications include brain abscess, meningitis, lateral sigmoid sinus thrombophlebitis, cavernous sinus thrombosis and petrous apicitis.

Petrous apicitis may be defined as an extension of infection from the mastoid air cell tract into a pneumatized anterior or posterior petrous apex.[3] With involvement of cranial nerves located near the vicinity of the petrous apex, Gradenigo syndrome may develop which includes the clinical triad of acute inflammation of the middle ear, pain in the temporal and parietal areas and paralysis of the abducens nerve.[1-10] These three cardinal features were observed during the course of illness of our patient. The left vocal cord paralysis and dysphagia are unusual, and need to be explained.

Petrous apicitis rarely occurs because infection in sclerotic or petrous apices containing marrow is uncommon and the prevalence of pneumatization is low. When it occurs, infection in the petrous apex may persist even if mastoid disease responds to treatment.[3]

Petrous apicitis may develop due to direct extension of acute otitis media into a pneumatized petrous apex. When drainage from the petrous apex to the middle ear is compromised, it promotes growth of microorganisms. Common etiologic agents include *Streptococcus pneumoniae, Haemophilus influenzae, and Staphylococcus aureus*[11] while tuberculous and fungal petrous apicitis have also been reported.[12]

Because of proximity of the petrous apex to the venous sinuses, the risk of developing venous sinus thrombosis is high. When infection reaches Dorello's canal which transmits the abducens nerve (CN VI) and the gasserian ganglion (CN V), the patient may manifest with lateral rectus palsy and hemifacial pain ipsilateral to the involved ear.[12] This explains the pain in the left eye, cheek and jaw which may mimic trigeminal neuralgia. A discerning clinician should evaluate and analyze the pain alongside other signs and symptoms. The dysphagia felt for two weeks when hoarseness and diplopia developed was accompanied by coughing, choking sensations and aspiration. Despite this, she was able to tolerate feeding with some difficulty and had no significant weight loss for the past three months. Her dysphagia maybe classified as neurogenic and may be secondary to involvement of the IX and X cranial nerve.

While symptoms of acute petrositis are subtle and may be attributed to sudden obstruction of the pneumatized petrous apex air cell system resulting from acute mastoid inflammation, chronic apicitis follows a more indolent course. Patients with chronic apicitis may not appear acutely ill compared to those suffering from acute petrositis.[12] As mentioned earlier, the patient had a childhood mastoid operation because of the intermittent ear infections spanning 50 years, may have developed chronic petrositis.

Involvement of the abducens nerve and gasserian ganglion may be secondary to local pachymeningitis as a consequence of acute petrous apicitis.[5] Other possible routes include spread of infection via pneumatized air cell tracts and vascular channels, or via direct extension beneath fascial planes.[6]

Symptoms of petrous apicitis may be attributed to middle ear and mastoid infection, and symptoms coming from the apex which include deep or retro-orbital pain (from irritation of the trigeminal ganglion in Meckel's cave), paralysis of the abducens nerve as it passes through Dorello's canal abutting the petrous apex, dysfunction of cranial nerves VII and VIII, or labyrinthitis.[1-10] In a study of 22 patients by Chole and Donald, otalgia was the most consistent symptom followed by deep pain and headache, and otorrhea.[2] Only one patient presented with CN IX and X involvement because of its proximity to the petrous apex.[2]

The patient met the diagnostic criteria for Gradenigo syndrome but the left vocal cord paralysis and dysphagia need further explanation. Was this a case of Gradenigo versus Vernet syndrome? The latter occurs

when lesions affect the jugular foramen where cranial nerves IX, X, XI are lodged. A stroke or cerebrovascular event was unlikely with no history of hypertension and negative cranial CT scan results.

The proximity of various venous sinuses to the petrous apex has been implicated as the cause of various complications following Gradenigo syndrome such as venous sinus thrombosis,[8] meningitis, intracranial abscess, involvement of CN IX, X, XI (Vernet Syndrome), involvement of the sympathetic plexus around the carotid sheath (Horner Syndrome), and prevertebral and parapharyngeal abscesses.[6] This is the possible mechanism by which the patient developed hoarseness and dysphagia after manifesting the Gradenigo triad.

The diagnostic procedure of choice are CT scans which show details of the petrous apex and provide important anatomical blueprints for surgical access.[13] If the CT scans indicate potential apicitis, magnetic resonance imaging (MRI) may add details about the nature of the fluid or tissue within the apex while a gallium bone scan may show high signal intensity on the side of the apicitis[3] Ideally, combination of MRI and CT scans are necessary to evaluate normal anatomic variations and to eliminate other disease entities.

Diagnostic findings of petrous apicitis on CT scan include opacification of the mastoid air cell system and the petrous apex, enhancement of the cavernous sinus and bony erosion within the petrous apex. With contrast media, cavernous sinus enhancement may be seen.[12] Contrary to acute petrous apicitis which may appear as an expanding lesion with irregular margins, chronic petrous apicitis may demonstrate hypopneumatization and sclerosis.[14] Sclerosis of the mastoid and petrous bone may be secondary to lack of development in which no pneumatization occurs or it may be due to new bone formation in a previously pneumatized area as a result of middle ear infection.[15]

Because of the complex anatomy and the need to dissect around delicate structures such as the labyrinth, cranial nerves and carotid artery, diseased petrous air cells cannot be totally removed by surgery and establishment of drainage and antibiotics are essential components of the treatment regimen. Indications for surgical drainage include failure to respond to antimicrobial therapy, cranial nerve deficits, brain abscess, and development of life-threatening complications.[12] A transmastoid, infralabyrinthine, suprajugular approach has been advocated to provide drainage and ventilation of the petrous apex with preservation of hearing.[7]

Prolonged postoperative antibiotics are recommended for 2-3 weeks after surgery, and for patients that may have accompanying osteomyelitis, 3-6 weeks of IV antibiotics may be required.[12] Pain caused by trigeminal nerve irritation usually resolves within one week of intravenous antibiotic therapy.[9] Other reports showed resolution of cranial nerve palsies within 3-4 weeks when adequately treated. Rapid resolution of cranial deficits can result if surgery is combined with antibiotics than with antibiotics alone.[12]

Gradenigo syndrome is well known for its triad of retro-orbital pain, lateral rectus (CN VI) paralysis and chronic middle ear infection due to petrous apicitis. Although rare, vocal cord paralysis and dysphagia may develop when infection traverses and encroaches on the jugular foramen where cranial nerves IX, X, and XI are lodged. Knowledge of the syndrome should not be limited or confined to the classic triad. Practicing ear specialists should be vigilant and cognizant of the clinical manifestations and sequelae of chronic middle ear disease. Prompt surgical intervention is crucial while resolution of the disease may vary for different individuals.

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