



Rare Case of Bilateral Facial Paralysis with Labrynthine Fistula in A Patient Diagnosed As Takayasu Syndrome

KEYWORDS

Takayasu's, LMN facial palsy, fistula

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ABSTRACT Complications of unsafe chronic otitis media are on decline since the advent of newer antibiotics & with the early referral to the specialists & the higher centres. The known complications are meningitis, sigmoid sinus thrombophlebitis, brain abscesses, labyrinthine fistula, mastoiditis, facial nerve paralysis. Here we are citing a case having bilateral facial palsy with bilateral labyrinthine fistula in the patient having Takayasu's syndrome.

Takayasu's disease is arteritis of unknown etiology. It is said to be secondary to tuberculosis. Mostly the young adult females are affected. Patient had history of tuberculosis ten years back. Then she was diagnosed as having Takayasu's syndrome too. She was operated for ear pathology. Now she developed facial paralysis on both sides & also complaining of giddiness.

Introduction-

Chronic suppurative otitis media is of two types, safe & unsafe. The safe type is always secondary to upper respiratory tract infection. Unsafe is due to the blockage of Eustachian tube leading to negative middle ear pressure which causes retraction of the tympanic membrane. The natural cleansing mechanism is hampered. Thus developing retraction pocket which grows slowly. Enlarging pocket may lead to pressure necrosis & also due to enzymatic activity the bone gets eroded. Spread of the disease may also be through preformed pathways or through blood. If the dural plate is eroded there is first formation of subperiosteal abscess and then different brain abscesses may develop. If sinus plate is eroded, subperiosteal abscess develops. Then sinus thrombophlebitis develops. When the bone of semicircular canal is eroded patient gets vertigo due to pressure changes across the fistula. The lateral semicircular canal being anatomically closer to mastoid antrum and aditus gets eroded commonly.

The facial nerve canal may also get eroded in its horizontal or vertical part leading to facial nerve paralysis. Our patient presented with bilateral cholesteatoma with bilateral labyrinthine fistula and bilateral facial paralysis. Other causes of bilateral facial paralysis are as under-

Trauma Skull fracture, Parotid surgery, Mastoid surgery.

Infection Postinfluenza, Infectious mononucleosis, HIV infection, Lyme disease, Banwarth's syndrome, Guillain-Barre syndrome, Syphilis, Brainstem encephalitis HTLV-1 infection, poliomyelitis

Metabolic Diabetes, Acute porphyria

Neoplastic Acute leukemia, Acoustic neuroma

Autoimmune Sarcoidosis, Amyloidosis

Neurological Multiple sclerosis, Pseudobulbar and bulbar palsy, Parkinson's disease

Idiopathic Bell's palsy

Takayasu arteritis commonly occurs in woman younger than age 50 years and can manifest as isolated, atypical, and/or catastrophic disease. It can involve any or all of the major organ systems. The disease has been reported in all parts of the world, although it appears to be more prevalent in

Asians.

Pathophysiology:

Takayasu arteritis is an inflammatory disease of large- and medium-sized arteries, with a predilection for the aorta and its branches. Advanced lesions demonstrate a panarteritis with intimal proliferation.

Lesions produced by the inflammatory process can be stenotic, occlusive, or aneurysmal. All aneurysmal lesions may have areas of arterial narrowing. Vascular changes lead to the main complications, including hypertension, most often due to renal artery stenosis or, more rarely,

Case report-

A 51 year old female patient came with the complaints of vertigo since last 6 mths, pain in both the ears with foul smelling discharge since 2 mths & facial weakness from one month. She was operated for right cholesteatoma 15 years back. Same old case papers were showing that she had pulmonary kochs that time. The old reports also revealed that she was diagnosed as having aortoarteritis.

General examination- masked facies, incomplete eye closure on both sides.

Pulse in both the upper limbs was not felt. Lower limb pulse was 78/min, Her carotid pulsations were felt prominently & she had a carotid bruit on right side.

BP in lower limbs was 160/66 mm Of Hg Bell's phenomenon was positive in both the eyes.(Fig1)



Fig.1: Bells phenomenon – preoperative

Findings on local examination-both the ears were full of cholesteatoma flakes.

S/O LMN facial palsy was present on both sides.

Fistula sign was also positive bilaterally.

Haematological investigation showed only raised ESR.

X-ray chest –old healed kochs.

ECG- Borderline LVH PTA- bilateral moderate to severe conductive deafness.

Angiography-irregularity of aortic lumen, dilated ascending aorta, irregularity with proximal narrowing of right innominate artery. Total occlusion of right carotid artery immediately after its origin. There was reformation of both carotids indicating development of collaterals.

She had optic atrophy and diminished vision.

Her ANA was negative.

CT temporal bone showed erosion of lateral semicircular canals & the fallopian canal.

Patient was taken for right sided radical mastoidectomy with exploration of the facial nerve under general anaesthesia. On opening the mastoid antrum, we could see that the mastoid antrum was filled with cholesteatoma. The fallopian canal

was eroded & bare facial nerve was seen. There was a small labyrinthine fistula and the dural plate seen was also eroded. After removing the disease completely labyrinthine fistula was covered with the fascia graft. After a week other mastoid was explored. That also revealed a labyrinthine fistula and the naked facial nerve. Fistula was closed with fascia graft. Post operatively patient was given physiotherapy also with oral steroids.

During the postoperative period we could see the improvement in facial palsy. (Fig 2)



Fig.2: Postoperative recovery

Our case was a rarest combination of multiple complications of cholesteatoma with Takayasu's disease. Whether takayasu's has predisposed her for multiple complications?

REFERENCE

- Hata A, Noda M, Moriawaki R, Numano F. Angiographic findings of Takayasu arteritis: new classification. *Int J Cardiol.* Aug 1996;54 Suppl:S155-63. [Medline]. | 2. Aggarwal A, Chag M, Sinha N, Naik S. Takayasu's arteritis: role of Mycobacterium tuberculosis and its 65 kDa heat shock protein. *Int J Cardiol.* Jul 5 1996;55(1):49-55. [Medline]. | 3. Soto ME, Vargas-Alarcón G, Cicero-Sabido R, Ramírez E, Alvarez-León E, Reyes PA. Comparison distribution of HLA-B alleles in mexican patients with takayasu arteritis and tuberculosis. *Hum Immunol.* May 2007;68(5):449-53. [Medline]. | 4. Hall S, Barr W, Lie JT, Stanson AW, Kazmier FJ, Hunder GG. Takayasu arteritis. A study of 32 North American patients. *Medicine (Baltimore).* Mar 1985;64(2):89-99. [Medline]. | 5. Jain S, Kumari S, Ganguly NK, Sharma BK. Current status of Takayasu arteritis in India. *Int J Cardiol.* Aug 1996;54 Suppl:S111-6. [Medline]. | 6. Park MC, Lee SW, Park YB, Chung NS, Lee SK. Clinical characteristics and outcomes of Takayasu's arteritis: analysis of 108 patients using standardized criteria for diagnosis, activity assessment, and angiographic classification. *Scand J Rheumatol.* Jul-Aug 2005;34(4):284-92. [Medline]. | 7. Siglock TJ, Brookler KH. Sensorineural hearing loss associated with Takayasu's disease. *Laryngoscope.* Jul 1987;97(7 Pt 1):797-800. [Medline]. | 8. Andrews J, Mason JC. Takayasu's arteritis—recent advances in imaging offer promise. *Rheumatology (Oxford).* Jan 2007;46(1):6-15. [Medline]. | 9. Andrews J, Al-Nahhas A, Pennell DJ, Hossain MS, Davies KA, Haskard DO, et al. Non-invasive imaging in the diagnosis and management of Takayasu's arteritis. *Ann Rheum Dis.* Aug 2004;63(8):995-1000. [Medline]. [Full Text]. | 10. Maksimowicz-McKinnon K, Hoffman GS. Takayasu arteritis: what is the long-term prognosis? *Rheum Dis Clin North Am.* Nov 2007;33(4):777-86. vi. [Medline]. | 11. McIntosh WE, Brenner JF, Aschenbrenner JE. Bilateral facial paralysis | as the sole presenting feature of sarcoidosis: report of a case. *J Am Osteopath Assoc.* 1987;87:245-7. | 12. George MK, Pahor AL. Sarcoidosis: a cause for bilateral facial palsy. *Ear Nose Throat J.* 1991;70:492-3. | 13. Haydar A, Hujairi NM, Tawil A. Bilateral facial paralysis: what's the cause? *Med J Am.* 2003;179:553. | 14. Steenerson RL. Bilateral facial paralysis. *Am J Otol.* 1986;7:99-103. | 15. Gevers G, Lemkens P. Bilateral simultaneous facial paralysis—differential | diagnosis and treatment options. *Acta Otorhinolaryngologica Belg.* | 2003;57:139-46. | 16. Teller D, Murphy T. Bilateral facial paralysis: a case presentation and | literature review. *J Otolaryngol.* 1992;21:44-46. | | 17. Stahl N, Ferit T. Recurrent bilateral peripheral facial palsy. *J Laryngol | Otol.* 1989;103:117-9. | | 18. Haydar A, Hujairi NM, Tawil A. Bilateral facial paralysis: what's the cause? *Med J Am.* 2003;179:553. | | 19. Steenerson RL. Bilateral facial paralysis. *Am J Otol.* 1986;7:99-103. | | 20. Gevers G, Lemkens P. Bilateral simultaneous facial paralysis—differential | diagnosis and treatment options. *Acta Otorhinolaryngologica Belg.* | 2003;57:139-46. | | 21. Teller D, Murphy T. Bilateral facial paralysis: a case presentation and | literature review. *J. Otolaryngol.* 1992;21:44-46.