

Case study:

Dehiscent Jugular Bulb in a case of Thalassemia: case study and brief review

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Abstract:

Dehiscence of the floor of the middle ear with protrusion of the jugular bulb into the cavity in an uncommon entity .It is an important cause of bluish mass behind the tympanic membrane and bleeds dramatically when incised.Tomography demonstrates the characteristic knuckle of jugular bulb extending into middle ear cavity ¹.

This entity was first described by Pages in 1914 and 10 cases were alluded to by Uffenorde in 1942.Plain film radiologic findings were described in 1946 and Gejrot and Lauren described venographic findings in high bulb position with protrusion in 1964.

INTRODUCTION:

Dehiscence of the floor of the middle ear with protrusion of the jugular bulb into the cavity in an uncommon entity .It is an important cause of bluish mass behind the tympanic membrane and bleeds dramatically when incised.Tomography demonstrates the characteristic knuckle of jugular bulb extending into middle ear cavity ¹.

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Overton and Ritter described histologic study of 257 temporal bones,13 of which had apex of jugular bulb extending above the inferior margin of the bony drum annulus into the middle ear cavity ³.Of these 13 specimens,12 had atleast some bone between the bulb and middle ear cavity and only one had a a truly dehiscent middle ear cavity floor.They found the prevalence of high jugular bulb to be 6%.In these cases the height of jugular bulb above the inferior margin of the bony annulus varied from 0.6mm to 4mm.

P.Vachata,P.Petrovicky and M.Sames described the relationship between jugular bulb and internal auditory canal in 200 temporal bones on high resolution CT scans and alcohol fixed skull bases of adults ⁴.The average distance between the internal auditory canal and jugular bulb was 7.5 ± 2.3 mm.Jugular bulb was was found to be higher on right side than its companion in 53.3% of patients.When the Jugular bulb reached or exceeded the floor of the IAC (16.5%) it was defined as High Jugular bulb.61% of HJB was found in females and bilateral HJB was found in 0.5% of patients.

The retrospective review conducted by Friedmann et al. concluded that jugular bulb abnormalities erode the vestibular aqueduct most often (25 patients),followed by the facial nerve (5 patients) and posterior semicircular canal (4 patients) ⁵.

CASE REPORT:

An 18 year old male presented to us with chief complaint of left earache which was insidious,intermittent and resolving spontaneously since last 3 years.Patient also had history of left tinnitus since last 2 years which was persistent,pulsatile,aggravated in silent surroundings and improved subjectively on applying digital pressure over internal jugular vein.It was associated with bilateral decreased hearing since 3 years which was insidious,progressive , more in left ear and interfered with patient’s day to day activities.There was history of conductive hyperacusis i.e transmission of sound of walking steps and eating meals into the ear.There was no history of ear discharge,headache,giddiness,facial deviation.

Patient was a diagnosed case of Thalassemia intermedia since last one and half year for which patient had taken regular blood transfusions.There was no history of taking chelation therapy. Patient’s younger sister 15 year old is also a diagnosed case of thalassemia .

On general examination patient had typical thalassemia facies comprising of maxillary hyperplasia,flat nasal bridge and frontal bossing .(Ref fig 1)



Fig 1: Thalassemia facies

On examination of right ear patient had bulging tympanic membrane with bluish hue suggestive of haemotympanum. On the left side patient had bulging tympanic membrane with bluish hue suggestive of haemotympanum, pulsation in postero-inferior tympanic membrane. Pulsation changed pattern on holding respiration. (Ref fig 2a&b)



Fig 2 A Right Tympanic membrane findings suggestive of Haemotympanum



Fig 2b: Left Tympanic membrane findings suggestive of haemotympanum and pulsation of Tympanic membrane in postero-inferior quadrant

On Tuning fork tests patient had negative Rinne's for 256 Hz,512 Hz and 1024 Hz.Weber's was centralized and absolute bone conduction was bilaterally decreased.Rest of ENT examination was within normal limits.There was no cranial nerve deficit.Pure tone Audiometry of the patient was suggestive of moderately severe mixed type of sensorineural hearing loss.Impedance audiogram showed bilateral 'B' type of curve suggestive of fluid in middle ear. High Resolution Computed Tomography of temporal bone was suggestive of absence of bony sigmoid plate between middle ear and jugular bulb suggestive of dehiscent jugular bulb on the left side.The sigmoid plate between jugular bulb and middle ear was maintained on right side.HRCT also showed soft tissue opacification of bilateral middle ear which could be attributed to bilateral haemotympanum , widened diploic space with coarse trabecular pattern which was consistent with osteoporotic changes seen in thalassemia .(Ref fig 3a & b)



Fig 3a: Axial tomogram showing dehiscent sigmoid plate and jugular bulb above the level of tympanic annulus on left side



Fig 3b: Axial tomogram showing normal jugular bulb on right side with intact sigmoid plate

Patient was counseled for Hearing Aid Trial for his sensorineural type of hearing loss and also counseled for tinnitus masker .It was thought judicious not to surgically intervene in this patient as firstly, his tinnitus was not incapacitating secondly, taking into consideration high risk of surgical complications due to thalassemia and finally,given the success rate of surgical interventions in this cases is variable ⁸ .

DISCUSSION:

High riding jugular bulb and dehiscent jugular bulb are 2 different entities.A high riding jugular bulb is distinguished from an asymmetrically large jugular bulb by its dome(roof) reaching above the internal acoustic meatus ⁶.A high riding jugular bulb has an intact sigmoid plate-a thin plate of bone separating the jugular bulb from the middle ear cavity.This can only be appreciated on thin slice bone algorithm CT.

If the sigmoid plate is deficient,the bulb is free to protrude into the middle ear cavity and is then known as a dehiscent jugular bulb and is a common cause of retrotympanic vascular mass ⁶ .

A high position of jugular bulb is a congenital variant associated with a large jugular fossa and usually is not associated with a dehiscent floor ⁷.High incidence is noted in cases of cranio-facial dysostosis like Crouzon's syndrome and Apert's syndrome . A dehiscent floor can occur with a normal jugular bulb size and position.The condition is usually acquired secondary to infection,trauma,aneurysm or tumou erosion.Although the term protruding jugular bulb implies an accompanying dehiscence, a dehiscence usually is not accompanied by bulb protrusion.

Tinnitus due to high jugular bulb is a venous tinnitus associated with an unusually located jugular vein .A high riding jugular bulb is a common vascular anomaly found in 2.4-7% of temporal bones.The jugular bulb is not present at birth,but develops over time.The size and location is somewhat dependent on pneumatization of the mastoid bone.

When present,this structure can be associated with bleeding during surgery,it can cause tinnitus,hearing loss,a conductive hyperacusis and Meniere's disease type symptoms.Tinnitus is attributed to direct pressure wave from the jugular.Conductive hyperacusis i.e increased awareness to somatosounds is due to increased compliance of the inner ear,in the similar way as is seen in superior canal dehiscence and fenestration surgery patients.

Diagnosis is mainly via MRI/MRA with contrast or similar techniques that show blood flow in relation to the skull.High resolution CT angiography is the method of choice. Internal jugular vein ligation has been used as surgical treatment for incapacitating pulsatile tinnitus due to venous pathology .Rouillard et al reported that ligation of the jugular vein usually do not produce long term successful results and blood shunting with occurrence of symptoms in the other ear ⁸.The results of this procedure have been very inconsistent,so that some reports have indicated recurrence of pulsatile tinnitus and the development of intracranial hypertension.Therefore degree of annoyance and daily life impact should be evaluated before surgical management of pulsatile tinnitus and it is necessary to spend sufficient time in observing the patient.Few studies have suggested surgical management using multilayer middle ear floor reconstruction(using bone dust,perichondrium and tragal cartilage) for management of incapacitating tinnitus in this patients ⁹.But the results of such studies are preliminary and reconstruction was found to be associated with high risk of sigmoid sinus thrombosis .

REFERENCES:

- 1) Y.Hertzanu,Dianne B.Mendelsohn,M.Waner,R.T. Gregor :Protruding jugular bulb presenting as middle ear mass:A case report.S A Mediese Tydskrif 1983;63:900-1.
- 2) Gejrot T,Lauren T :Retrograde jugulography in diagnosis of abnormalities of the superior bulb of the internal jugular vein,Acta otolaryngol(stockh) 1964;57:177-18
- 3) Overton SB,Ritter FN :A high placed jugular bulb in middle ear,A clinical and temporal bone study.Laryngoscope. 1973;83(12):1986-1991.
- 4) P.Vachata,P.Petrovicky,M.Sames:An anatomical and radiological study of the high jugular bulb on high-resolution CT scan and alcohol-fixed skull of adults. J Clin Neurosci 2010 ;17(4):473-8.
- 5) Friedmann DR,Eubig J,Winlata LS,et al :A clinical and histopathologic study of jugular bulb abnormalities,Arch Otolaryngol Head Neck Surg 2012;138(1):66-71.
- 6) Dehisent Jugular Bulb Ch.18 Pg 61-62 by Sachin Gujar : Temporal Bone Imaging edited by Ellen G. Hoeffner , Suresh K. Mukherji, Dheeraj Gandhi, Diana Gomez-Hassan
- 7) Frank W. Farrell and Oscar Hantz :Protruding jugular bulb presenting as a middle ear mass : Case report and brief review,AJR Am J Roentgenol 1977;128(4):685-7
- 8) Hyeog-Gi Choi et al : Clinical efficacy of Internal Jugular vein ligation in treatment of vascular tinnitus :Case report,Int. Adv Otol 2009;5(2):255-60
- 9) El-Begermy MA,Rabie AN :A novel surgical treatment for management of tinnitus due to high dehiscent jugular bulb,Otolaryngol Head Neck Surg 2010;142(4):576-81.