

CASE REPORT

Jugular Bulb Diverticulum Mimicking Meniere's Disease: Case Report

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The position and size of the jugular bulb is incredibly variable. High mega jugular bulb is a vascular abnormality of the internal jugular vein at the jugular foramen. Mostly, high jugular bulb remains asymptomatic although ear diseases are present in some cases. The position of jugular bulb diverticulum determines the kind and degree of related symptoms. High jugular bulb could even mimic Meniere's disease with severe acute vertigo episodes, sensorineural hearing loss, and tinnitus. We present a rare case of a 17-year-old male patient with symptomatic high mega jugular bulb with diverticulum requiring surgery along with literature review.

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Introduction

Jugular bulb, for its variable anatomical positions, may extend laterally to the tympanic membrane or external auditory meatus, and medially to the petrous apex or inner ear. If the dome of the jugular bulb projects above the inferior aspect of the bony annulus or the basal turn of the cochlea, it is considered as a high jugular bulb. If the jugular bulb is large in cross-diameter, it is called mega jugular bulb regardless of its position. In addition, a jugular bulb diverticulum is characterized by its location inside the expansion of the jugular bulb wall potentially causing pressure to the surrounding structures that leads to various symptoms.^[1,2]

Most cases of high jugular bulb are asymptomatic while symptomatic cases have also been reported.^[3] Reported symptoms vary according to the location of jugular bulb diverticulum; vertigo, sensorineural hearing loss, and tinnitus may be caused in medial location.^[4] High jugular bulb may mimic Meniere's disease showing severe acute vertiginous attacks, pulsatile tinnitus, and hearing loss.^[5]

In this case, surgical treatment can relieve the symptoms. We experienced one case of Meniere's disease mimicking jugular bulb diverticulum with vertigo that was treated surgically; therefore, we would like to report this case with literature review.

Case Report

17-year-old male visited our outpatient clinic complaining intermittent headaches, vertigo, and seizure that were not improving by previous treatment. He was already diagnosed with and treated for Meniere's disease three years ago at another hospital. Given treatments were Meniett (Meniett, Medtronic Inc., USA) wearing and diuretics (dichlozid) for more than six months. Prior to the diagnosis of Meniere's disease, he was treated with anti-seizure medication at the department of pediatrics and neurology in another hospital. When he visited our outpatient clinic, his both tympanic membrane was normal, hearing test was within normal range for the left and 40dB for the right which was low frequency hearing decline, and no response from glycerol test. Since he was

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already treated with diuretics, he was followed up at outpatient clinic for six months to observe the response. During follow-up observation, there was fluctuation in hearing ability. Low frequency hearing was further decreased to 60dB, and vertigo became worse. Glycerol test was repeated for worsened vertigo. There was no change in the threshold of pure tone audiometry while the threshold of speech reception test showed an improvement from 40dB to 25dB; therefore, the result was positive. During vertigo attacks, no nystagmus was observed on VNG, and both slow harmonic acceleration and caloric test were normal. His vertigo, hearing declination and tinnitus on the right side, and headache were worsening extent to which he was unable to perform everyday activities; thus, immediate treatment was urgently required. According to diagnostic criteria of

Meniere's disease published by AAO-HNS in 1995, this patient falls on definite Meniere's disease, and he was stage three considering hearing threshold.^[6] The report standard of treatment effect for vertigo was not significant enough to classify this patient since follow-up time period after diuretics administration was only for six months. We were supposed to follow-up the patient eighteen more months observing for the effect of diuretics. However, considering the young age and rapidly worsening symptoms of the patient, we decided to perform endolymphatic sac decompression. Pre-operative computerized tomography of temporal bone showed high jugular bulb with diverticulum which was compressing vestibular aqueduct (Fig. 1). In order to release the compression on the vestibular aqueduct applied by jugular bulb diverticulum, we decided to treat this patient

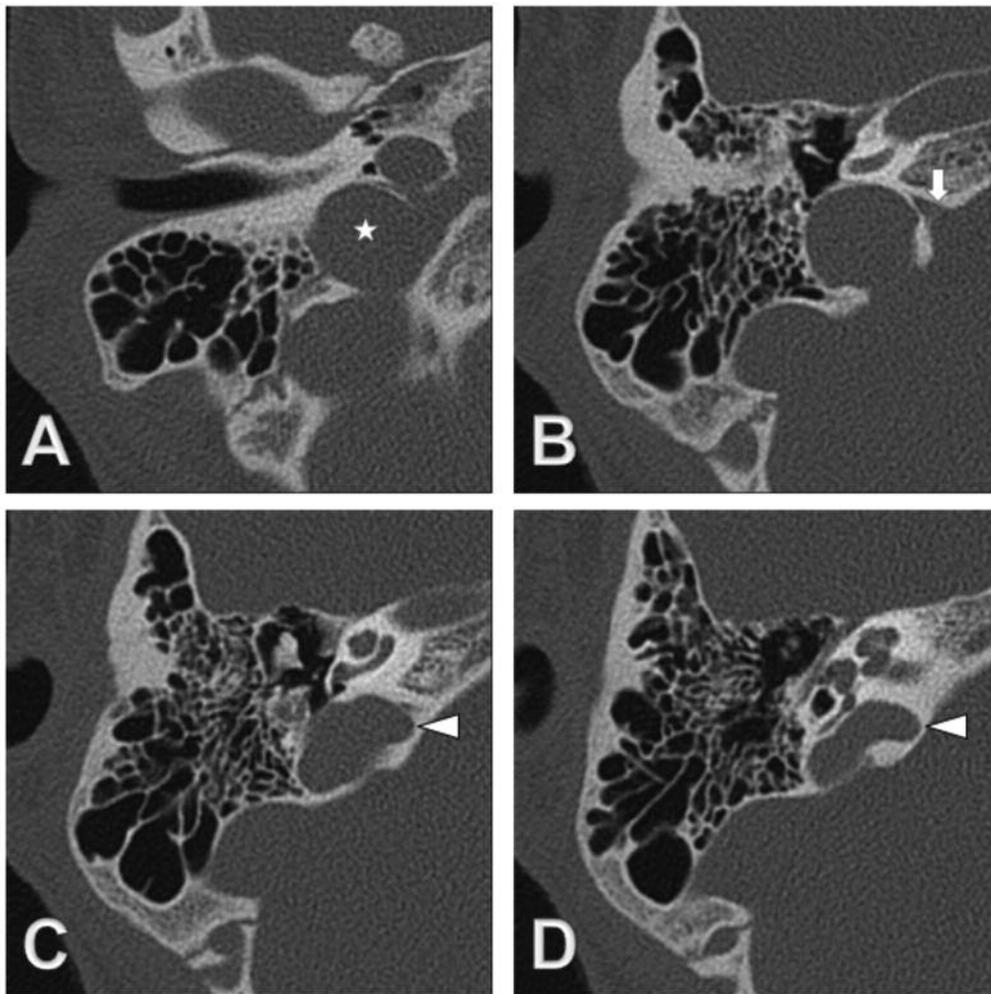


Figure 1. Temporal bone CT(axial view), **A:** high jugular bulb(asterisk), **B:** vestibular aqueduct(white arrow), **C,D:** jugular bulb diverticulum(white arrow head) compressing vestibular aqueduct

surgically under general anesthesia. We performed venography to evaluate the function of internal jugular vein in opposite site which turned out good. A retroauricular skin incision to the neck was done to expose the mastoid and the internal jugular vein in the neck. Complete mastoidectomy was performed exposing the sigmoid sinus, the facial nerve in the mastoid, and the stylomastoid foramen. The jugular bulb was exposed by subfacial, and infralabyrinthine approaches. After we expose the dura mater of the posterior cerebral fossa, the lateral and posterior walls of the jugular bulb were exposed. The size of the jugular bulb was large, and its location was high extending to the level of the posterior semicircular canal very close to the sigmoid sinus. Since the endolymphatic sac was not observed, approaching to the endolymphatic sac was impossible (Fig. 2). The sigmoid sinus was extraluminally packed with Surgicel(Surgicelâ, Ethicon. Inc., NY, US). The jugular bulb and inferior petrosal sinus was progressively compressed with Surgicel(Surgicelâ, Ethicon. Inc., NY, US). The emissary vein was cut and the internal jugular vein was ligated at the neck (Fig. 3). Since he was showing no significant problems for three postoperative days, he was discharged. Three months after the operation, pure tone audiometry was performed. The hearing ability on the right side was 40dB on average, and the low frequency and threshold of speech reception audiometry was 30dB; the result showed improvement in hearing. During seven months follow-up period of the patient, vertigo was progressively improved extent to which he was able to perform everyday activities with no difficulties including going to school. We are still following up the patient, and he has no other symptoms except tinnitus.

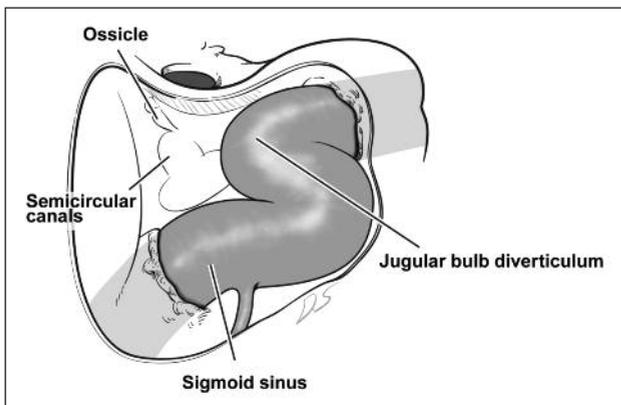


Figure 2. Surgical view after complete mastoidectomy exposing sigmoid sinus and high jugular bulb.

Discussion

Endolymphatic hydrops has been well known as the pathophysiology of Meniere's disease.^[7] In many animal studies, symptoms similar to the ones of Meniere's disease were caused by either decreasing endolymph resorption, or increasing endolymph secretion in endolymphatic duct or sac.⁸ Also in anatomical and tomographic studies carried by Wadin and Wilbrand, possibility of Meniere's disease occurrence was considered to be high in the patients who had high jugular bulb that was in contact with the distal portion of vestibular aqueduct.^[2,3] Cases of Meniere's disease with pulsatile tinnitus were reported to be associated with high juglar bulb in many literatures.^[9-12] Similar condition was found in our patient. His jugular bulb diverticulum was preventing the vestibular aqueduct from reabsorbing endolymph due to compression (Fig. 1); thus, this condition was considered to cause Meniere's disease. However, since the bony erosion in vestibular aqueduct was not observed directly in surgical field, there was a possibility of which the juglar bulb disrupted the venous drainage of endlymphatic sac.

Meniere's disease is predominantly found in adults. The average age of onset is from the fourth to the seventh decades of life. In the present study, the prevalence ranged from 9 cases per 100,000 in patients under 18 years of age.^[13] Although the pediatric cases of Meniere's disease, according to previous case reports, have known to well respond to diuretics,^[14] our patient did not respond to diuretics at all, and his age was 14 which was considered to be young. His condition was worsening more rapidly than typical Meniere's disease cases. This condition is thought to have a lot of possibility to be

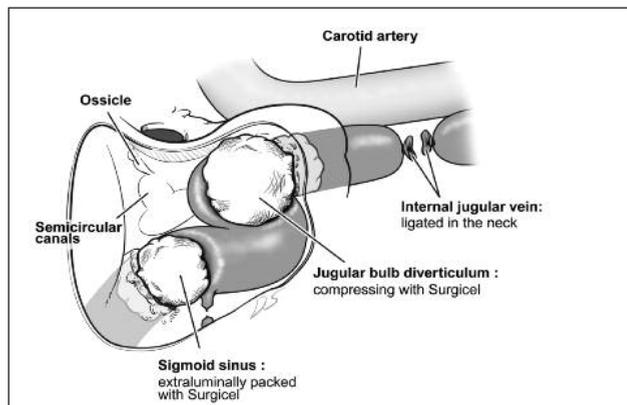


Figure 3. Surgical view; sigmoid sinus and jugular bulb were compressed with Surgicel and the internal jugular vein was ligated in the neck.

associated with the development and characteristic of patient's jugular bulb.^[15] Jugular bulb is a dynamic structure that is not present at birth, and it starts forming after 2 years of age and continues until adulthood. In addition, high jugular bulbs are not anatomically stable, and progressively modeled by centrifugal hemodynamic forces.^[15,16] As this force applies on the thin wall of jugular bulb in which tunica adventitia is absent, jugular bulb expands to form diverticulum at jugular dome.^[3] Such expansion is normally located in the pneumatized region of the petrous bone in which mechanical resistance is low. A progressive increase in the volume and height of high jugular bulb has been observed in patients followed for several years.^[17,18] Taking these conditions into account, as this patient grew from adolescence to adulthood, venous blood flow dynamics affected the size or position of jugular bulb which turned high jugular bulb into diverticulum resulting in suppressing or eroding adjacent structures.

Bozorg Grayeli et al. reported a case in which they treated surgically eight cases of jugular bulb diverticulum with vertigo mimicking Ménière's disease.^[18] Then, they published about surgical lowering of the high jugular bulb as treatment for fifteen patients of Ménière's disease with pulsatile tinnitus. They reported, in most of the cases, that vertigo attacks were successfully managed, hearing ability remained unchanged compared to pre-operative condition, and tinnitus was significantly improved.^[19] Although the cases of intracranial hypertension or a tear of the dura mater in the posterior cerebral fossa, have been reported, the morbidity of these complications is lower than the one occurs with labyrinthectomy and vestibular neurectomy.^[19-21]

Diagnosis of Meniere's disease is based on AAO-HNS guideline published in 1995^[6]. The guideline covers only clinical symptoms such as vertigo with more than twenty minutes of spinning nature, hearing disturbances. Clinicians, although this is due to the pathophysiology of Meniere's disease that has not been fully understood, tend to overlook radiologic study in clinic. Since our patient was young in age, and symptoms were worsening rapidly despite of diuretics therapy, it was inevitable to treat him surgically instead of waiting for eighteen to twenty four months to check the response of diuretics; then, the outcome was successful. The symptoms of the patient would have been worse if the follow-up time of diuretics was

prolonged. In addition, the prognosis may have been better if radiologic study was performed even earlier. Recently, the diagnosis rate of Meniere's disease has been increasing, for more information about the symptoms of Meniere's disease is becoming available making more people to visit their doctors for Meniere's disease. However in the clinic, symptoms are not sufficient enough to make a diagnosis for Meniere's disease. Therefore, considering the limitation in diagnosing Meniere's disease, performing preceded radiology study such as computerized tomography is thought to be helpful. Furthermore, more efficient, accurate, newer guidelines for diagnosing Meniere's disease may be suggested if multicenter radiologic study is carried.

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References

1. Filipović B, Gjurić M, Hat J, Glunčić I. High mega jugular bulb presenting with facial nerve palsy and severe headache. *Skull Base*. 2010; 20(6):465-8.
2. Wadin K, Wilbrand H. The jugular bulb diverticulum. A radioanatomic investigation. *Acta Radiol Diagn (Stockh)*. 1986; 27:395-401.
3. Wadin K, Thomander L, Wilbrand H. Effects of a high jugular fossa and jugular bulb diverticulum on the inner ear. A clinical and radiologic investigation. *Acta radiol Diagn (Stockh)* 1986; 27:629-636.
4. Presutti L, Laudadio P. Jugular bulb diverticula. *ORL J Otorhinolaryngol Relat Spec* 1991; 53:57-60.
5. Bozorg Grayeli A, Bouccara D, Julien N, Rihane S, Chaigne P, Sterkers O . Surgical treatment of vertigo induced by jugular bulb diverticulum. *Rev Laryngol Otol Rhinol Bord* 1995; 116:31-37.
6. Committee on Hearing and Equilibrium guidelines for the diagnosis and evaluation of therapy in Ménière's disease. American Academy of Otolaryngology-Head and Neck Foundation, Inc. *Otolaryngol Head Neck Surg*. 1995; 113:181-5.
7. Merchant SN, Rauch SD, Nadol JB. Ménière's disease. *Eur Arch Otorhinolaryngol* 1995; 252:63-75.

8. Feldman AM, Brusilow SW. Effects of cholera toxin on cochlear endolymph production: model for endolymphatic hydrops. *Proc Natl Acad Sci USA* 1976; 73:1761-1764.
9. Dilenge D. The jugular "notch". *J Can Assoc Radiol* 1977; 28:274.
10. Graham MD. The jugular bulb: its anatomic and clinical considerations on contemporary otology. *Laryngoscope* 1977; 187:105-125.
11. Jahrsdoerfer RA, Cail WS, Cantrell RW. Endolymphatic duct obstruction from a jugular bulb diverticulum. *Ann Otol Rhinol Laryngol* 1981; 90:619-623.
12. Pappas DG Jr, Hoffman RA, Cohen NL, Holliday RA, Pappas DG Sr. Petrous jugular malposition (diverticulum). *Otolaryngol Head Neck Surg* 1993; 109:847-852.
13. Harris JP, Alexander TH. Current-day prevalence of Ménière's syndrome. *Audiol Neurootol*. 2010;15:318-22. Epub 2010 Feb 18.
14. R. Filipo, M. Barbara. Juvenile Ménière's disease J. *Laryngol. Otol.* 1985; 99:193-6.
15. Friedmann DR, Eubig J, McGill M, Babb JS, Pramanik BK, Lalwani AK. Development of the jugular bulb: a radiologic study. *Otol Neurotol*. 2011; 32:1389-95.
16. Beyer RA, McCarty GE. High jugular and high carotid canal first observed as intracranial bruit. *Arch Neurol* 1983; 40:387.
17. Buckwalter JA, Sasaki CT, Virasongse C, Kier EL, Bauman N. Pulsatile tinnitus arising from jugular megabulb deformity: treatment rationale. *Laryngoscope* 1983; 93:1534-1539.
18. Couloigner V, Grayeli AB, Bouccara D, Julien N, Sterkers O. Surgical treatment of the high jugular bulb in patients with Ménière's disease and pulsatile tinnitus. *Eur Arch Otorhinolaryngol* 1999; 256:224-229.
19. Bigelow DC, Hoffer ME, Sclakman B. Angiographic assessment of the transverse sinus and vein of Labbé to avoid complications in skull base surgery. *Skull Base Surg* 1993; 3:217.
20. Haid CT, Gjuric M, Wigand ME (1996) Vestibular nerve section and neuro-vascular decompression of patients with Ménière's disease. In: Vesterhauge S, Katholm M, Mikines P (eds) Ménière's disease. 16th Danavox symposium. Scanticon, Denmark, p 317.
21. Magnan J, Bremond G, Chays A, Gignac D, Florence A. Vestibular neurectomy by retrosigmoid approach: techniques, indications, and results. *Am J Otol* 1991; 12: 101-04.