High Mega Jugular Bulb Presenting with Facial Nerve Palsy and Severe Headache

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ABSTRACT

We present a rare case of a 50-year-old female patient with symptomatic high mega jugular bulb requiring surgery. We review her medical file, preoperative and postoperative imaging, audiograms, and surgical report. High jugular bulb was diagnosed with computed tomography and magnetic resonance imaging. Symptoms of facial nerve palsy and headache were abolished after surgical procedure. Headache and facial nerve palsy can be caused by high mega jugular bulb. Surgery is indicated in such symptomatic cases and leads to relief of signs and symptoms of disease.

KEYWORDS: High jugular bulb, facial nerve palsy, headache, surgery, jugular foramen

High mega jugular bulb is a vascular abnormality of the internal jugular vein (IJV) at the jugular foramen. The position and size of the jugular bulb are extremely variable. High jugular bulb (HJB) is diagnosed if the cranial margin of the jugular bulb extends over the inferior surface of the bony annulus, protruding into the middle ear or over the basal turn of the cochlea. In one histological study, HJB was found in 3.5% of 815 temporal bone samples.¹ If the jugular bulb is large in cross-diameter, it is called *mega jugular bulb*, regardless of its position. In two-thirds of the population, the jugular bulb and the jugular vein on the right side are significantly larger than the ones on the left side.² A jugular bulb diverticulum is a localized expansion of the jugular bulb wall, with a potential of causing pressure to the surrounding structures that leads to various symptoms.³

In most cases, HJB remains asymptomatic, but sometimes symptoms of ear disease are present.⁴ The symptoms related to a jugular bulb diverticulum depend on its localization. A lateral protrusion toward the tympanic cavity leads to symptoms of conductive hearing

loss and pulsatile tinnitus, whereas a medial localization toward the petrous apex can cause vertigo, sensorineural hearing loss, and tinnitus.⁵ HJB can even mimic Ménière's disease with severe acute vertiginous attacks, as reported in a case study of six patients.⁶ So far, three patients were described in the literature who had various forms of facial nerve involvement ranging from facial twitching and recurrent facial paralysis to episodic hemifacial spasm.⁷⁻⁹ In regard to this rare clinical entity, we add a fourth case and evaluate the presentation and management considerations.

CASE REPORT

In this report, we present a 50-year-old female patient, who was referred with persisting headache lasting for 28 months despite various medical treatments. Her main symptom was an incapacitating headache starting behind the right ear and extending to the right side of the face. The headache was present in a laying or bending position, usually for a period of several hours.

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Figure 1 Patient presenting with right-sided facial nerve palsy limited to the corner of the mouth (with patient approval).

With each attack, the headache was becoming more intense. Soon it was so severe that the patient was neither able to perform her daily activities nor do her job and was constantly on narcotic analgesics. One month after the headache started, she developed an ipsilateral lower facial asymmetry on resting and smiling. Two months later, she reported right-sided hearing loss and tinnitus.

Head and neck examination revealed right peripheral facial nerve palsy limited to the corner of the mouth (Fig. 1). Audiological testing demonstrated tympanometry and stapedial reflexes within the normal range and a mild right sensorineural hearing loss of 34 dB. Needle electromyoneurography of the musculus zygomaticus and musculus orbicularis oris showed signs of chronic neural lesion, registered as a significantly reduced motor conduction velocity and reduced amplitude of the compound muscle action potential in the right facial nerve, as compared with the opposite side. The patient underwent high-resolution computed tomography (CT) scan with the standard radiological protocol for the inner ear (Fig. 2), magnetic resonance imaging (MRI) scan (Figs. 3 and 4), and 3-D angiography and postcontrast venography (Fig. 5). Imaging demonstrated a high-positioned mega jugular bulb on the right side with a diverticulum at its cranial curvature reaching the vestibular aqueduct (Fig. 5). Additionally, intimate contact of the HJB with the mastoid segment of the facial nerve was shown (Fig. 4).

The patient was advised to undergo a surgical exploration of the jugular foramen area with exposure of the facial nerve and removal of the jugular bulb anomaly. A retroauricular skin incision with extension to the neck was used to expose the mastoid and the IJV in the neck. Complete mastoidectomy was performed, exposing the sigmoid sinus and the facial nerve in the mastoid and the

Figure 2 CT axial scan: high jugular bulb (B) protruding into tympanic cavity, with close contact to the mastoid segment of facial nerve (N VII).

Figure 3 MRI T1 axial scan (equivalent plain): contrast-enhanced, HJB (B) protruding into tympanic cavity, mastoid segment of facial nerve (N VII).

Figure 4 MRI T1 3-D contrast-enhanced, oblique reconstruction image: mastoid part of the facial nerve (N VII) in contact with the HJB (B).

Figure 5 Dynamic contrast-enhanced angiography: arterial and venous phase, right internal carotid artery (ACI), HJB diverticulum (D).

stylomastoid foramen. The sigmoid sinus was extraluminally packed with Surgicel (Ethicon, Neuchatel, China) and the IVJ ligated in the neck. The bony canal of the facial nerve was found eroded by the pressure of the HJB in a short segment inferior to the posterior semicircular canal toward the level of the digastric ridge. The denuded facial nerve was separated from the HJB, which was then resected, keeping its medial wall preserved. Patching of the bone defect and multilayer wound closure concluded the procedure.

Immediately after surgery, there was a dramatic drop in the intensity of the headache. Right facial nerve palsy, which developed in the early postoperative period, resolved completely after 2 months. The hearing loss remained unchanged and the tinnitus diminished shortly after the operation. One year after surgery, the patient is well and working again. From time to time, she experiences some pain, which is different in character and due to vertebral discopathy (C3–C7). The patient is at present undergoing physical therapy for this problem.

DISCUSSION

Anatomic variations and anomalies of the IJV at the jugular foramen are common but mainly asymptomatic. If present, symptoms are related to the jugular foramen mass and lead to unilateral hearing loss, aural fullness, tinnitus, and vestibular symptoms. Otalgia and mi-

grainlike headache are possible in patients with different vascular anomalies, whereas symptoms of cranial nerve involvement are extremely rare.¹⁰ Jahrsdoerfer et al, in the review of literature on jugular bulb diverticula, mentioned one patient with facial twitching.⁸ Pappas et al reported a case of a young woman with a history of recurrent facial paralysis since age 16 years and episodic hemifacial spasm between episodes of paralysis.⁹ Like our patient, she also suffered from frequent parietal headaches, and the facial paralysis was also limited to the lower face. The patient did not undergo surgery. Gal et al presented a 33-year-old woman with jugular diverticulum and facial paralysis who underwent transmastoid surgery with posterior tympanotomy.⁷ Facial nerve decompression led to recovery of its function.

In a patient with clinical symptoms of jugular foramen mass, it is mandatory to rule out the presence of an expanding tumor (cholesteatoma, neurilemmoma, meningioma, or paraganglioma) and the existence of vascular anomalies (enlarged jugular bulb or aberrant internal carotid artery). The correct diagnosis is made from an imaging study including high-resolution CT scan and MRI scan. Vascular pathology is evaluated with CT, magnetic resonance angiography, and digital subtraction angiography. In our patient, CT scan demonstrated a wide HJB that protruded into the tympanic cavity and reached the external auditory canal, which was dehiscence. There were no signs of osteolysis or destruction of surrounding pneumatic cells. Contrast enhancement of the lesion was homogenic; MRI scan showed hypointense signal in the T1 and T2 sequences (flow signal void), and postcontrast enhancement of the lesion was homogenic, consistent with the CT scan. The HJB was in close contact with and compressed the mastoid part of the facial nerve canal. There was no contrast enhancement of surrounding structures that would suggest additional anomaly or tumor. 3-D time-of-flight magnetic resonance angiography showed a normal aspect of internal carotid artery and other arteries. Postcontrast venography showed the characteristic pattern of HJB.

Management of high mega jugular bulb must be individualized. Asymptomatic cases are discovered while evaluating unrelated pathology and do not require treatment. The appreciation of the existence of HJB is of extreme importance if surgical procedures are planned in the ear. Rare cases of symptomatic cranial nerve involvement merit surgical consideration. A high index of suspicion and a reasonable degree of plausibility of symptoms possibly explained by the existence of HJB or jugular diverticulum must be present. Competency of the contralateral venous drainage must be established prior to surgery. Surgery itself is a standard lateral skull base procedure with minimal morbidity and high chances of success in properly evaluated cases.

CONCLUSION

The presence of HJB can cause severe headache and facial palsy. Diagnosis is confirmed with CT, MRI, and vascular imaging studies. In such symptomatic cases, surgical management is reasonable and has high chances of success in alleviating the symptoms and problems.

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