Aplasia of Sigmoid Sinus with High-Lying Jugular Bulb

Ziad L. Deeb,1 Dov Rotenberg,2 and F. Owen Black3

A patient was seen for investigation of a retrotympanic mass associated with pulsatile tinnitus. A high-lying jugular bulb and congenital aplasia of the ipsilateral sigmoid sinus were diagnosed. Review of the world literature failed to reveal any prior such report.

Case Report

A 25-year-old woman was admitted with a 20 year history of pulsatile tinnitus. She had been aware of "buzzing" in her right ear since she was a child. There was no history of otalgia, hearing loss, nor ear discharge.

The physical examination revealed a blue hypotympanic mass medial to an intact right tympanic membrane. Increasing the air pressure in the external canal failed to cause a blanching of the mass (Brown sign). There was no audible bruit. An audiogram showed a minimal low-toned ipsilateral conductive loss. Electro-nystagmography, posturography, and rotation testing were normal.

Petrosal pyramid tomography showed a large right jugular foramen but without any obvious bony erosion. CT and cerebral angiography failed to reveal any mass lesion or abnormality. Venous films were not obtained at angiography.

Retrograde jugular venography showed obstruction of the right sigmoid sinus at the level of the jugular bulb (fig. 1A). There seemed to be good cross-filling through the cavernous sinus and parasellar venous plexus to the contralateral side (fig. 1B).

An exploratory tympanotomy was attempted. Severe bleeding when elevating the flap made this impractical. The bleeding was controlled and a posterior tympanotomy was performed. A high blind-ended jugular bulb was encountered and after exenteration of the entire mastoid air cell system, no sigmoid sinus could be found proximally.

Discussion

The venous drainage of the neurocranium arises from the fusion of the capillary network (surrounding the fetal neural tube) into two cardinal veins. With further skull development, these vessels perforate the skull through foramina that are destined to become the jugular foramina. The superior part of these veins, above the jugular foramen, develop into the sigmoid sinus while the inferior part becomes the internal jugular veins.

Fig. 1.—A, Retrograde jugular venogram. Complete obstruction at jugular bulb. Inferior petrosal sinus fills (arrow). B, Cross filling to contralateral jugular system. Sigmoid sinus was never visualized. Negative shadows of carotid arteries in cavernous sinuses are demonstrated (white arrows). Catheter (black arrow) seen with tip in jugular bulb region.
Page [1] was the first to report massive bleeding at myringotomy due to a superiorly located jugular bulb. Overton and Ritter [2] subsequently serially sectioned a large number of temporal bones and found a 6% incidence of high-lying jugular bulbs. Of 189 specimens dissected, nine had the right side abnormal, four the left side, and one bilaterally. Since then, there have been sporadic reports and descriptions of both normal and abnormal morphology of the sigmoid sinus and jugular bulb [2–7] in its relation to otologic surgery. The incidence on the right side is higher, as expected, because the transverse and sigmoid sinuses are larger on the right in 75% of the people [8].

Aplasia of the sigmoid sinus is also rarely encountered. Kalbag and Woolf [9] in discussing cerebral venous thrombosis state that the sigmoid sinus may be absent or narrowed and venous drainage accomplished through other pathways, mainly via the mastoid foramina. A thorough search of the world literature failed to reveal any previous report of congenital aplasia of the sigmoid sinus in association with a high-lying jugular bulb.

We believe that aplasia of the sigmoid sinus should now be included in the differential diagnosis when the sigmoid and/or transverse sinus system fails to fill on angiography, especially on jugular venography, and when a tumor is suspected.

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REFERENCES