Hidden Connection: Unusual Case of Vertigo as a Result of Sinus Pericranii

Introduction

Sinus pericranii (SP) is an unusual vascular abnormality that involves an anomalous connection between intracranial and extracranial venous systems.[1] This connection is established through transosseous emissary veins and can be either congenital or acquired.[2] Although it is typically diagnosed in infants and young children, cases have been reported in patients of all ages.[3] Interestingly, some researchers have suggested that it may result from transient increases in intracranial venous pressure during late embryonic development.[4] The clinical presentation varies widely, making the diagnosis challenging. While some patients may remain asymptomatic, others may present with a palpable scalp mass or a pulsatile bruit. Additional symptoms may include headache, dizziness, and neurological deficits, such as visual disturbance or seizures. Symptomatic SP commonly manifests with headaches, which can have a gradual onset and fluctuating pattern over several months or have an acute onset.[5] While the exact cause of SP-related headaches remains unclear, intracranial hypertension (IIH) is thought to play a role. Hereby, we report an elderly lady who presented with vertigo managed adequately with repositioning maneuvers but was found to have asymptomatic IIH and SP on imaging.

Description of the Case

A 65-year-old lady presented with complaints of intermittent episodes of internal vertigo (particularly during head movements or in changing posture, which used to subside spontaneously or after taking betahistine) for 2–3 years, bilateral pulsatile tinnitus for 5 months, increased frequency of vertigo episodes for 3 months, and intermittent occipital headache and imbalance while walking for 3 weeks. Her supine rollover test was positive and had right horizontal canal Benign paroxysmal positional vertigo (BPPV), initially unresponsive to the Barbecue manoeuvre (BBQ) maneuver. However, significant improvement was achieved with the Gufoni maneuver. In between vertigo episodes, the patient was completely symptom-free. At the presentation, the general physical examination was noncontributory. Higher mental status including speech was normal. Her best corrected visual acuity was 6/6 in both eyes. The fundus examination showed bilateral papilledema. Visual field charting showed a mildly enlarged blind spot. B scan done revealed a small amount of subarachnoid fluid in the bilateral optic sheath (left > right). The head impulse test was negative. The rest of the cranial nerve and motor and sensory examinations were normal. Deep tendon reflexes were normal, and plantars were bilaterally flexor. The patient had mild swaying toward the right side while walking, difficulty in taking turns, and could not perform tandem gait, but no other cerebellar signs were observed. Considering the long history of intermittent postural internal vertigo, the patient seems to have developed persistent postural perceptual dizziness (PPPD) in the current episode. Audiology evaluation showed bilateral hearing sensitivity within normal limits. Routine investigations including hemogram and biochemical parameters were normal. Cerebrospinal fluid (CSF) opening pressure was 150 mm of CSF. Magnetic resonance imaging (MRI) brain [Figure 1] revealed tortuous optic nerves with empty sella and dilated Meckel's cave suggestive of IIH. MR venogram showed a hypoplastic left transverse–sigmoid sinus junction. SP was seen with the anomalous connection between intra- and extra-axial venous systems. Considering raised intracranial pressure, the patient was treated with acetazolamide 250 mg thrice daily. On follow-up after 3 months, the patient had almost complete resolution of her symptoms.

Discussion

The term "Sinus pericranii' was initially introduced by G. Stromeyer in 1850 to describe a blood-filled reservoir situated on the skull bones, which establishes communication between the dura mater sinuses via the diploic veins.[6] Even after more than a century of its description, consensus regarding the diagnosis, classification, and management of SP remains elusive, primarily due to the relative rarity of the condition. SP forms during the later stages of embryonic development, primarily as a consequence of venous hypertension caused by occlusion of venous outflow.[7] Additionally, it is frequently observed in individuals with craniosynostosis.[8] It can arise as a consequence of trauma indicating the multifaceted nature of its etiology.[7] The anomalous connection between the intra- and extracranial venous systems results in several

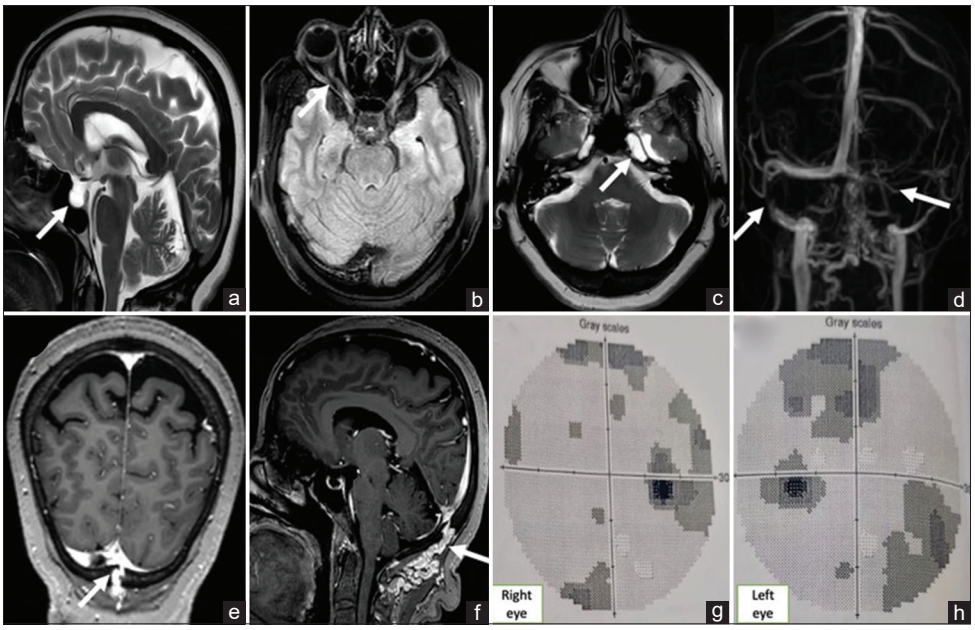

Figure 1: (a) Sagittal T2-weighted image showed empty sella. (b) Axial FLAIR image showed bilateral tortuous optic nerves with prominent perioptic CSF spaces. (c) Axial T2-weighted image showed bilateral prominent Meckel's caves with diffuse prominence of CSF spaces. (d) Magnetic resonance (MR) venogram sequence showed abrupt narrowing of the right transverse sigmoid junction noted. Left transverse and sigmoid sinuses are narrow in caliber. (e and f) (Coronal and sagittal, respectively) T1-weighted MPRAGE sequence showing extracranial tortuous veins connecting with intracranial torcula (confluence of superior sagittal sinus, straight sinus, and occipital sinus) forming sinus pericranii. (g and h) An enlarged blind spot in both eyes

pathophysiological alterations. As the blood from these sinuses is drained into extracranial veins, intracranial venous pressure reduces, leading to impaired cerebral venous drainage and raised intracranial pressure. Additionally, the diversion of blood away from the intracranial venous sinuses can induce a decrease in cerebral perfusion, which can trigger cerebral ischemia.

In our patient, though the MRI showed features of IIH, CSF opening pressure was normal, probably SP was compensating for the raised intracranial pressure. SP usually occurs in the frontal region and is typically found in the median or paramedian area.[5] Occipital location is extremely rare.[4] Unlike usual pediatric cases, our case is presented in old age.[3] While most cases are asymptomatic, patients may exhibit varied symptoms. Mild symptoms can include local pain, nausea, headache, and dizziness. In more severe cases, symptoms may involve increased intracranial pressure, bradypnea, bradycardia, ataxia, hearing loss, and seizures.[9] Giddiness as a symptom of SP has been reported in a few reports.[4,9] Because the SP and its associated probable IIH (probable IIH because CSF opening pressure was normal, but MRI brain had telltale signs of IIH) cannot explain the patient's symptoms of "unsteadiness during walking with an impaired tandem walk, lateralized sway to one side, and difficulty in turning," there is a possibility of concurrent PPPD. The only other central pathology that could have explained these symptoms is tonsillar herniation due to IIH, but this was not found on the MRI of this patient. The association between IIH and SP is rare, but the pathophysiological link between them seems a logical explanation for their association. For treatment, most reports suggest it to be a benign condition and the primary concern often revolves around its cosmetic appearance. Gandolfo *et al*.[8] in 2007 presented a useful classification of SP into dominant and accessory SP that can aid in the treatment decision. They highlighted that while dominant SP cannot be treated due to their crucial role as a major venous outflow channel to the intracranial compartment, accessory SP is treatable since they only carry a small portion of intracranial venous outflow.[8]

Earlier reports have encompassed various treatment modalities, including surgical intervention, endovascular embolization, and conservative management approaches.[8,9] In cases where treatment for cosmetic purposes is pursued or when there is a risk of potential complications such as hemorrhage and embolism, the endovascular approach has demonstrated both safety and efficacy, particularly in managing accessory SP.[10,11] Overall SP is associated with a favorable prognosis, as the likelihood of traumatic or spontaneous hemorrhage is very low. Despite its rarity, SP poses an intriguing challenge for clinicians and requires careful evaluation and management.

In conclusion, SP is a rare vascular anomaly that can be challenging to diagnose and manage. In our case we observed, vertigo can be the primary manifestation of SP and it can present in the elderly. Our case expands the spectrum of this unusual clinical entity. Knowledge of this condition helps in appropriate diagnosis, management, and prognostication.

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Informed consent

Formal consent was taken from the patient.

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Conflicts of interest

There are no conflicts of interest.

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