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Neoplasm

Exophytic choroid plexus papilloma of the fourth ventricle presenting with cerebrospinal fluid rhinorrhea: a case report

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Abstract

Background: Choroid plexus papillomas are rare benign neoplasms that arise from the ventricular choroid plexus and are considered to be of ependymal origin. They are slow-growing lesions, are commonly associated with hydrocephalus, and are prone to hemorrhage spontaneously. Most of them are located in the posterior fossa in adults, the fourth ventricle being the most common location.
Case Description: We report a case of a 61-year-old male patient with a fourth ventricular exophytic choroid plexus papilloma extending caudally into the foramen magnum causing obstructive hydrocephalus. In February 2005, he presented with spontaneous CSF rhinorrhea to an ENT surgeon and underwent an endoscopic transnasal repair. Six months later, he came to us with progressive loss of vision due to raised intracranial pressure.
Conclusion: The lesion may not be detected on CT scans and MRI scan is the imaging modality of choice. Excision of the tumor takes precedence over any attempt to repair the fistula, as many a time, the CSF leak may stop.

Keywords: Choroid plexus papilloma; Hydrocephalus; CSF rhinorrhea

1. Introduction

Choroid plexus papillomas are benign, slow-growing tumors. They account for 0.4% to 1% of all intracranial neoplasms in all age groups [12,13], mostly occurring in children with an incidence of 1.5% to 4%, with a peak in the first 2 years of life. They are less frequent in adults comprising 0.3% to 0.89% of intracranial tumors. In adults, they are mostly located in the fourth ventricle and can occasionally extend to an extraventricular space such as the foramen magnum. Tumors in such locations may reach a large size and can obstruct CSF flow and cause hydrocephalus. The prolonged raised intracranial pressure can cause bony and dural defects resulting in CSF rhinorrhea. It is difficult to diagnose choroid plexus papilloma preoperatively when the main portion of the tumor is not located in the fourth ventricle, as the lesion may be missed on a CT scan. This is the purpose of presenting this case—to highlight the importance of an MRI scan of the brain to rule out a posterior fossa lesion with extension into the foramen magnum in patients with spontaneous CSF rhinorrhea.

2. Case report

A 61-year-old man presented to us with history of a spontaneous clear watery discharge from the left nostril since February 2005. He consulted an ENT surgeon for the same, and a CT cisternogram was done, which showed a bony defect in the anterior cranial fossa base in the region of the

Abbreviations: CPP, choroid plexus papilloma; CSF, cerebrospinal fluid; CT, computed tomography; ENT, ears, nose, and throat; ICP, intracranial pressure; MRI, magnetic resonance imaging.

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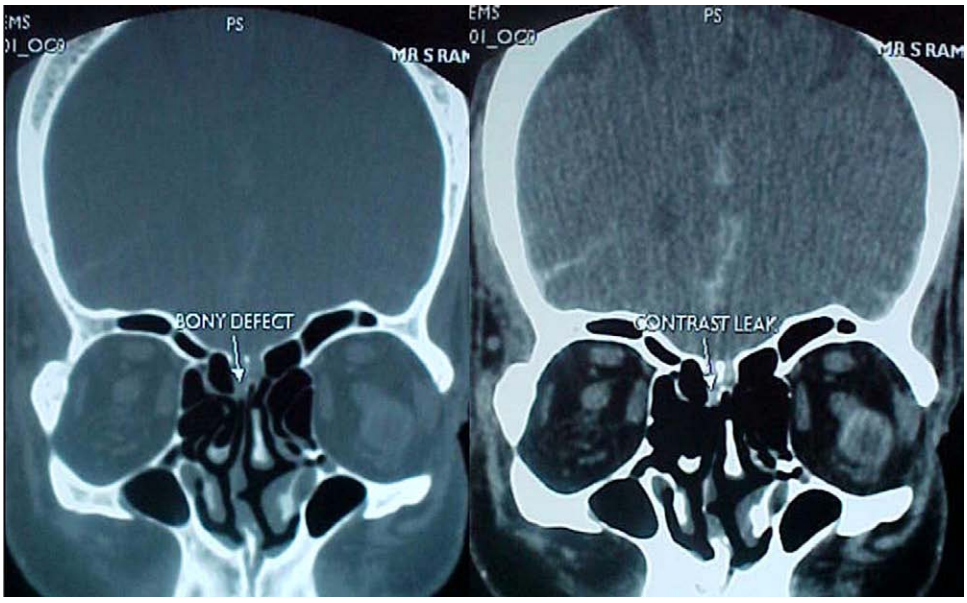

Fig. 1. Computed tomographic cisternogram showing site of CSF leak.

cribriform plate on the left side (Fig. 1). A CT scan of the brain plain and contrast study was also done, and there was no evidence of an intracranial space-occupying lesion or hydrocephalus (Fig. 2). He underwent an endoscopic transnasal repair of the CSF fistula with a fascia lata graft in March 2005. The surgery was done by an ENT surgeon. Postoperatively, the CSF leak stopped. Six months later, in September 2005, he presented to us with complaints of progressive deterioration of vision in his right eye, with visual obscurations, occipital headaches, and vertigo for 3 months. On examination, he had papilloedema; his visual acuity in the right eye was only perception of light with normal vision in the left eye. He had no other cranial nerve deficits, no long tract signs, and no cerebellar signs. There were no neurocutaneous markers of neurofibromatosis. An MRI scan of the brain showed a well-defined, irregular-shaped, exophytic lesion of the fourth ventricle, descending down through the fourth ventricle. It measured 40 × 29 × 21 mm. The lesion was hypointense on T1-weighted images, heterogeneously hyperintense on T2-weighted images, and homogeneously enhancing with contrast (Fig. 3). The lesion was displacing the cerebellum superiorly, and the inferior vermian region was effaced. The cervicomedullary junction was pushed anteriorly and compressed against the clivus with hyperintense signal changes seen in the cord. Hydrocephalus was present. In prone position, a midline suboccipital craniectomy, removal of the posterior arch of the atlas, and total excision of the lesion was done. The lesion was growing exophytically out of the fourth ventricle and extending inferiorly through the foramen magnum. Macroscopically, it resembled a choroid plexus papilloma. The postoperative period was uneventful. Postoperative MRI scan confirmed total excision of the tumor (Fig. 4). Histopathology report was choroid plexus papilloma.

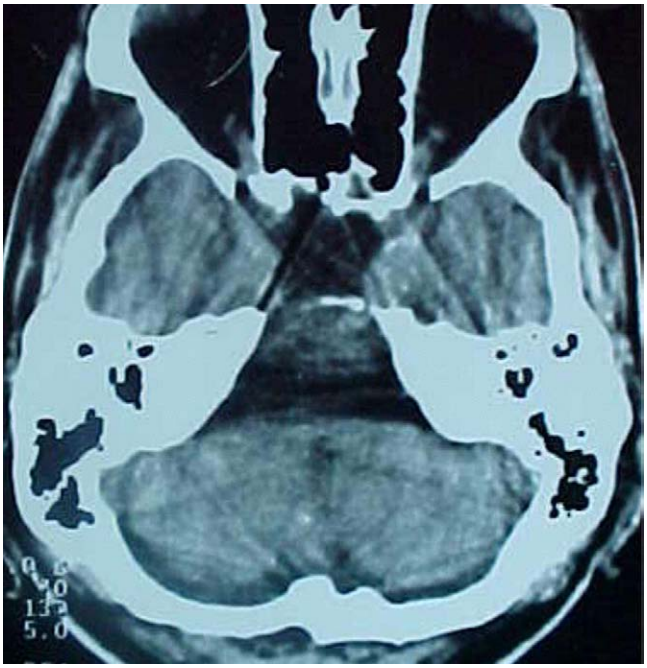

Fig. 2. Computed tomographic scan showing no lesion in the fourth ventricle.

3. Discussion

Choroid plexus papilloma was described by Guerard in 1832, and Perthes described the first successful removal in 1919. They occur less frequently in adults; most are located in the posterior fossa and arise from the fourth ventricle. Its extraventricular location and extension toward the foramen magnum is extremely rare. Ventricular enlargement is seen in