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Case Report

Fourth Ventricle Choroid Plexus Papilloma presenting with CSF Rhinorrhoea

Brajesh Kumar[1]* and Anita Kumari[2]

[1]*Assistant Professor, Department of Neurosurgery, Indira Gandhi Institute of Medical sciences, Sheikhpura, Patna, Bihar, India*

[2]*Associate Professor, Department of O&G, NSMCH, India*

***Corresponding Author:** Brajesh Kumar, Assistant Professor, Department of Neurosurgery, Indira Gandhi Institute of Medical sciences, Sheikhpura, Patna, Bihar, India.

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Abstract

Background: Cerebrospinal fluid (CSF) when comes out from the nasal cavity is called CSF Rhinorrhoea which can either be spontaneous or non-spontaneous. Trauma (Accidental and Surgical), tumors and exposure to radiation therapy to the base of skull are the most common causes of Non-spontaneous CSF Rhinorrhoea [1]. Congenital anatomical defects are the commonest cause of spontaneous CSF Rhinorrhoea which are not very common and has been reported, less than 4% [2].

Objective: To discuss the case of a Fourth Ventricle Choroid Plexus Papilloma in a 28-year-old female with 5 month amenorrhea which presented with spontaneous CSF rhinorrhea as an isolated complain. She was treated with primary management (microsurgical excision) of the tumor. CSF rhinorrhea was secondary to raised ICP (Intra cranial pressure) which caused the dehiscence of the cribriform plate.

Method: We planned for microsurgical excision of fourth ventricle choroid plexus papilloma with a thought that in next stage we will do the surgery for CSF rhinorrhea if that not resolve with tumor excision.

Keywords: CSF Rhinorrhea; Choroid Plexus Tumour; Intracranial Pressure

Key Message

Microsurgical excision of the fourth ventricle choroid plexus papilloma in a 28-year-old female resolved the spontaneous CSF rhinorrhea which was her isolated complain for last six months. Treatment of primary pathology of 4th ventricular resolved the CSF leak which was looking like second pathology.

Introduction

Cerebrospinal fluid (CSF) Rhinorrhoea can spontaneous or non-spontaneous. Spontaneous or non-traumatic is a less common causes of CSF Rhinorrhoea. Which include congenital anatomical defects of temporal and frontal skull base, or duramater [1]. Non-spontaneous are the most common cause of CSF Rhinorrhoea due to Traumatic injury to skull base (surgical and accidental trauma), tumors, or e post radiation therapy to base of the skull [1]. Early diagnosis and effective management are needed to prevent the life-threatening complications of CSF rhinorrhea, including bacterial meningitis and brain abscesses. We present a case of CSF rhinorrhea as an isolated complain for six months, associated with fourth ventricle choroid plexus papilloma in a previously well 28-year-old female having 5 month amenorrhea.

Case Presentation

A 28-year-old pregnant female presented with history of six-month of progressive Rhinorrhoea from the both nostrils. CSF rhinorrhea was aggravated on forward bending, coughing and

straining. The amount and frequency of rhinorrhea increased over time and by the time of admission water used to start dripping through her nose whenever she sits with forward bending. Initially she was having history of occasional headaches for last 10 months, which was more in the morning and occasionally associated with blurring of vision and nausea. These symptoms resolved once the CSF rhinorrhea started 6 months back. At the time of admission to the hospital there were no complaints of headache. There was no any other known medical co-morbidity. She was having congenital squint of right eye. Regular obstetric call was given throughout the stay for fetal wellbeing.

On examination, clear fluid poured from the both nostril when the patient moved from the supine to prone position. On fundoscopy, there was normal cup-to-disc ratio with the absence of papilledema. The 'Handkerchief test' was in favour of CSF Rhinorrhoea. The biochemical analysis of the nasal fluid confirmed the presence of CSF.

CT cisternography was performed at ENT center showed 7mm X 6mm sized defect in the region of greater wing of sphenoid with active leakage of contrast filled CSF into Right sphenoid Sinus extending through nostrils. There was no hydrocephalus or any other lesion reported (Figure 1).

Figure 1: CT cisternography was performed at ENT center showed 7 mm X 6 mm sized defect in the region of greater wing of sphenoid with active leakage of contrast filled CSF into Right sphenoidal Sinus extending through nostrils.

We advised for the Magnetic Resonance Imaging (MRI) Brain and paranasal sinuses with gadolinium Contrast revealed (Figure 2A, 2B and 2C).

- There was tracking of CSF from B/L cribriform plate which shows continuity with subarachnoid space.
- CSF fistula in right sphenoid sinus through defect in inferolateral wall of sphenoid sinus.
- There is a lobulated soft tissue lesion of size approx. 3.2×2.8×3.5 centimetres, noted in posterior fossa with mass effect, lesion was posterior-inferior to medulla and inferior to cerebellum. The lesion was heterogeneous hyper intense on T2W, T1W and no restriction on DWI.

Figure 2A: There is tracking of CSF from B/L cribriform plate which shows continuity with subarachnoid space.

Figure 2B: CSF fistula in right sphenoid sinus through defect in inferolateral wall of sphenoid sinus.

Figure 2C: Lobulated soft tissue lesion of size approx. 3.2×2.8×3.5 centimeters, noted in posterior fossa with mass effect, lesion was posterior-inferior to medulla and inferior to cerebellum. The lesion was heterogeneous hyper intense on T2W.

We thought for two stage surgery. In first stage we planned for microsurgical excision of tumor and second stage surgery for repair of fistula either through endoscopic approach or through open approach if it does not resolve as per our belief that it was secondary to raised ICP. Intraoperatively tumor was whitish in color, firm in consistency, attached with pedicle of blood vessel, hanging in the cavity of fourth ventricle.

We could do the complete microsurgical excision of the tumor, the Dura repaired with help of Duragen^{R} (Artificial Dural product). Post -operative period was uneventful and CSF rhinorrhea stopped from 2nd post-op days. Stitch was removed on 11th day and patient observed for 15th post-op days and discharged on 15th post-op day. Post -operative MRI done to confirm complete excision of the tumor (Figure 3).

Figure 3: Post-operative MRI of brain.

Histopathology of the tumor was choroid plexus papilloma (WHO grade 1) (Figure 4 A and B).

A

B

Figure 4: A and B: Histopathological examination of the tumor.

Discussion

There is a communication between the subarachnoid space and defect (Dura and bone) in the skull base which causes leaking of CSF through the nasal cavity [1]. Spontaneous or non-traumatic CSF Rhinorrhoea is uncommon and has been reported less than 4% [2]. Spontaneous CSF rhinorrhea has been associated with raised intracranial pressure (ICP) [3,4]. Previous studies have hypothesized that prolonged Intra Cranial Hypertension (ICH) may lead to defects in the skull base over time though the pathogenesis of CSF Rhinorrhoea is not clear.

Presence of prolonged ICH in presence of defect of Dura or bone can cause herniation of the Dura mater into the bony defects. This causes further weakening the Dura mater. These herniated Dura are more prone to tears and leads to Dural-mucosal fistula [5]. This patient in the present case was having symptoms of raised intracranial pressure (morning headache, vomiting and blurring of vision).

Beta-2 Transferrin or Beta-2 trace protein [6] is the best test for detecting the presence of CSF. Beta-2 transferrin is exclusively found in CSF, peri- lymphatic fluid, and the vitreous humor of the eye, with a reported sensitivity of 100% and a specificity of 95% [6]. In our patient, we have not done Beta-2 Transferrin test. We did comparative blood glucose concentrations of the nasal fluid to the blood. Glucose is absent in the nasal secretions. The presence of glucose in the nasal secretions points toward the presence of CSF but this is not recommended as a confirmatory test due to low diagnostic specificity and sensitivity. There can be false-negative results in the presence of bacterial contamination or false-positive results can be there in the diabetic patients [6]. Therefore, for the confirmation there should be clinical and radiographic evidence to support the diagnosis of CSF Rhinorrhoea [7].

High-resolution CT and MRI scans can accurately differentiate between spontaneous and non-spontaneous CSF Rhinorrhoea. This can help in the localization of leaks, can demonstrate the fractures of bone and presence of the tumour but do not demonstrate the leakage itself [2]. CT/MR cisternography is the gold standard for the detection of CSF leaks as it can identify the size, location, and quantity of the leak but is an invasive procedure and thus considered unnecessary if the diagnosis is supported by both the clinical presentation and imaging findings on CT and MRI [6,7].

Treatment of CSF Rhinorrhoea depends upon the type of pathology. It may be conservative or surgical. The main goal of the Conservative treatment is toward decreasing the ICH/ICH, Decreasing CSF Production by using Acetazolamide, Bed rest with elevation of the head with measures to decreasing Coughing and straining [9]. Surgical intervention includes either an Endoscopic or extra cranial approach or an intracranial approach. Intracranial approach carries increased morbidity and failure rates of 20-40%, whereas an endoscopic approach has less associated morbidity and a success rate of 90-100% [10,11]. Intracranial approach has the advantage of working in the large area where leaks can be seen directly and repaired. Currently the endoscopic repair is the Gold standard and initial treatment of CSF leaks. Intracranial repair is reserved if indicated or if the endoscopic repair has failed [12].

In our case, we thought for two stage surgery. In first stage we planned for microsurgical excision of tumor and second stage surgery for repair of fistula either through endoscopic approach or through open approach if it does not resolved. We performed the first stage surgery and excised the tumor and luckily CSF rhinorrhea stopped from Post-operative Day-1. Post- operative period was uneventful; patient was observed for 15 days in ward and followed up in OPD after 2 weeks. Fetal wellbeing was monitored regularly.

Conclusions

Choroid plexus papilloma of fourth ventricle is a rare tumor in adults. When patient present with CSF Rhinorrhoea as the only complain, it creates confusion in diagnosis and management. But in our case CSF rhinorrhea was due to raised intracranial pressure caused by choroid plexus papilloma of fourth ventricle. Once microsurgical excision of the tumor was done, the CSF flow was normalized and there was resolution of raised ICP and CSF Rhinorrhoea. So we should first treat the primary pathology and second stage surgery can be done if resolution of symptom does not occur.

Conflicts of Interest

Nil.

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