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Vestibular schwannoma clinically presenting with CSF rhinorrhea: A case report

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Abstract

Background: CSF rhinorrhea is leakage of CSF from the intracranial cavity through osseous & dural defects of the skull base into the paranasal sinuses or the nasal cavity. Common causes of this entity are trauma and iatrogenic skull base defects. Rarely, presence of large intracranial tumors may also be associated with CSF rhinorrhea.

Case Presentation: A 58-year-old female presented with complaint of intermittent headache and episodes of watery nasal discharge since 6 months. Endoscopy and biochemical examination confirmed the presence of CSF rhinorrhea. CT images of head and paranasal sinuses of this patient showed large right CP angle mass lesion, along with hyperpneumatisation of sphenoid sinuses as well as aberrant arachnoid granulations. MR images confirmed the findings of CT images and additionally showed features of intracranial hypertension and small sphenoid encephalocele.

Conclusions: Large intracranial tumours may rarely present with CSF rhinorrhoea, especially in cases, where paranasal sinuses are hyperpneumatised or arachnoid granulations are abnormal large. Hence, in non-traumatic cases of CSF rhinorrhea, imaging of brain should be advised to rule out any space occupying lesion.

Keywords: CSF, arachnoid granulations, hyper pneumatized, vestibular schwannoma, encephalocele

Introduction

Background: CSF rhinorrhea is described as cerebrospinal fluid (CSF) leakage from the intracranial cavity through osseous & dural defects of the skull base into the paranasal sinuses or the nasal cavity. Clinically, it presents as intermittent, often positional, discharge of clear fluid from the nose [1].

CSF rhinorrhea can be primary or secondary based on underlying cause. Among secondary types, common causes are trauma and iatrogenic skull base defects. One of the rare causes of such secondary CSF leaks are presence of large intracranial tumors, which in turn may cause raised intracranial pressure and bone erosion during the course of many years [2]. One such rare case we have encountered in our hospital, which, we are going to present here, along with review of literature.

Case presentation: A 58-year-old woman presented to otorhinolaryngology outpatient department with complaint of intermittent headache and episodes of watery nasal discharge since 6 months. She also gave history of right sided hearing loss for 1 year, for which she was never investigated properly. On endoscopic examination in otorhinolaryngology department, CSF rhinorrhea was suspected and further confirmed on biochemical study.

For further evaluation of the cause of CSF rhinorrhea, computed tomography of skull base and paranasal sinuses was performed.

A. Findings on CT images of skull-base and paranasal sinuses

First striking finding on the CT images was an incidental note of a large, well-defined, round to oval mass lesion at right cerebello-pontine angle which appeared to be beaking and extending into the right internal acoustic meatus (Figure 1a). Lesion was also causing widening of meatus (Figure 1b), with overall appearance of "ice-cream cone" (Figure 1c). It was causing mass effect in the form of effacement of right cerebellar folia and partial effacement of fourth ventricle, leading to mild upstream supratentorial ventricular dilatation (Figure 1d). Mild cerebellar tonsillar herniation was also noted (Figure 1e). Based on these imaging features, provisional diagnosis of vestibular schwannoma was made and contrast enhanced MRI was planned.

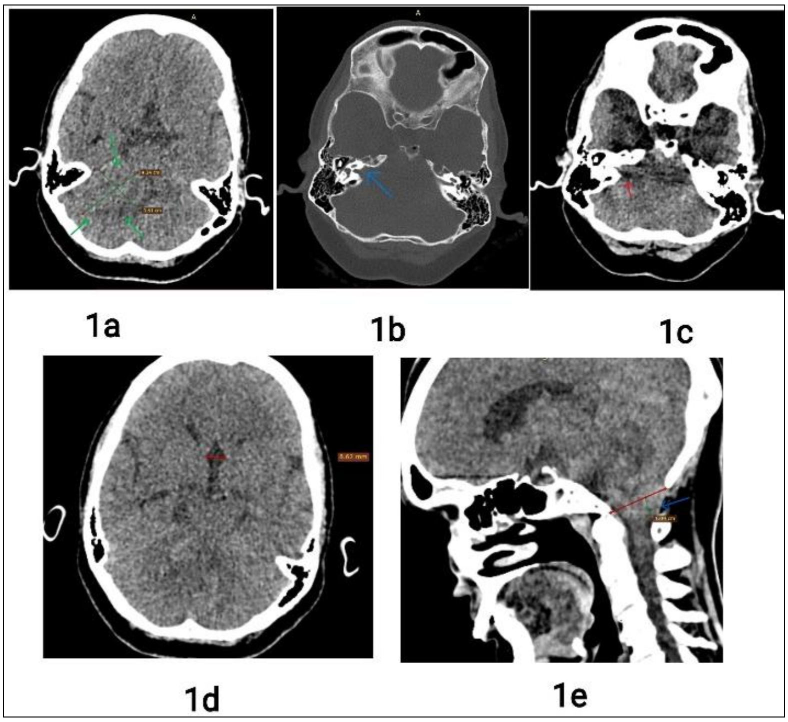

Fig 1: Non-contrast CT head images- axial images of CT head in brain window showed large, well-defined, round to oval mass lesion at right cerebello-pontine angle (1a, green arrow). Widening of internal acoustic meatus (1b, blue arrow) and resultant ice-cream cone appearance (1c, red arrow) are also seen. Also note the mild dilatation of third ventricle (1d, red arrow) and tonsillar herniation (1e, blue arrow)

Other findings included extensive pneumatization of sphenoid bone on both sides including the lateral recesses (left>right) with severe thinning of roof of sphenoid sinus (Figure 2a, 2b). Numerous circumscribed, irregular, lytic lesions were noted in greater wing of sphenoid and sphenoid body, with thin sclerotic margins and no sign of aggressive growth (Figure 2c). Imaging morphologies were in favor of aberrant arachnoid granulations.

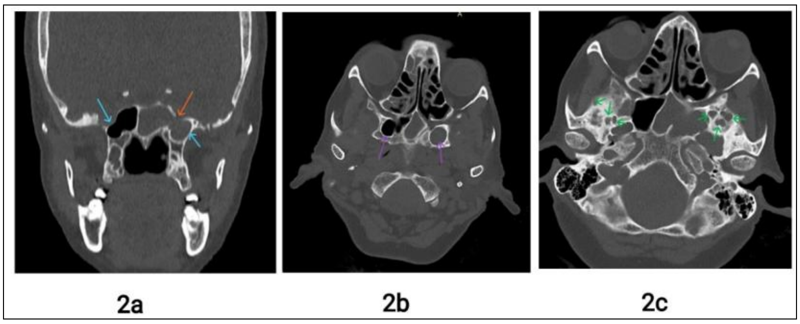

Fig 2: Bone window of CT head images shows extensive pneumatization of sphenoid bone on both sides including the lateral recesses (cyan/purple arrows) with severe thinning of roof of sphenoid sinus (orange arrow) (2a, 2b). Also note the numerous circumscribed

Multifocal small bone defects are noted at left lateral aspect of body of sphenoid bone in the region of lateral recess (Figure 3). Fluid to soft tissue density content was seen in continuity from left temporal lobe to left sphenoid sinus- explaining the cause of CSF leakage. Another similar smaller bony defect was seen at roof of left sphenoid sinus in para-midline location.

B. Contrast-enhanced MRI of brain and paranasal sinuses

The lesion in right CP angle appeared iso-hypointense on T1-weighted images, hyperintense on T2-weighted images and showed intense nearly-homogeneous enhancement on post-contrast sequences (Figure 4). Beaking and extension of the lesion is noted into the right internal acoustic meatus causing its widening and giving overall appearance like ice-cream cone. The lesion showed some internal non-enhancing areas which represent cystic/necrotic changes. Few blooming foci were also seen on susceptibility-weighted images, suggesting microbleeds (Figure 4). CSF cleft was seen at the interface of the lesion with the cerebellar parenchyma suggesting that extra axial location of the lesion. Overall features were more in favor of right vestibular schwannoma.

Mass effects due to the lesion were similar to the seen on CT images. Marked indentations and displacements of right cerebellar hemisphere, pons and medulla were noted. Fourth ventricle was approximately 10 mm displaced towards left, with deformed shape.

There were prominent subarachnoid spaces around bilateral optic nerve with vertical tortuosity (Figure 5a, 5b). Mild flattening of posterior sclera, prominent bilateral Meckel's cave, partially empty sella and mild cerebellar tonsillar herniation were also noted. Optic nerve sheath diameter was approx. 5.8 mm on each side, at 3 mm distance from posterior scleral margin. These findings favored the possibility of raised intracranial pressure.

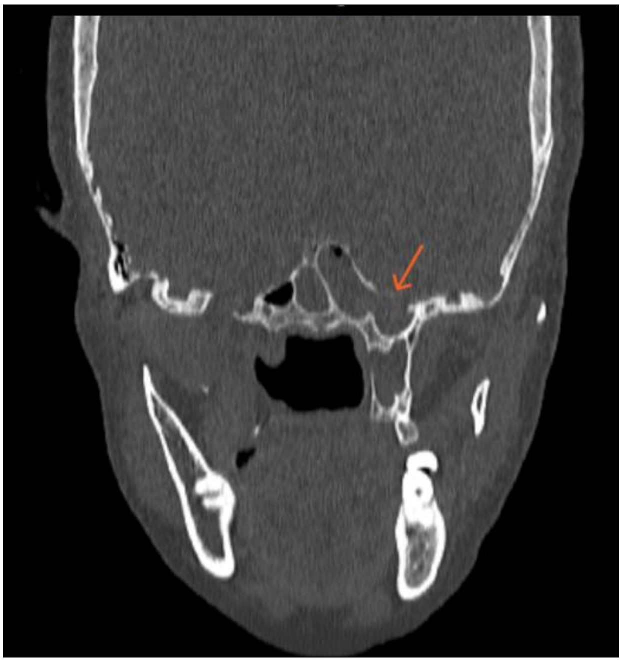

Fig 2: Bone window of coronal images of CT head depicts bone defects at left lateral aspect of body of sphenoid bone in the region of lateral recess (orange arrow).

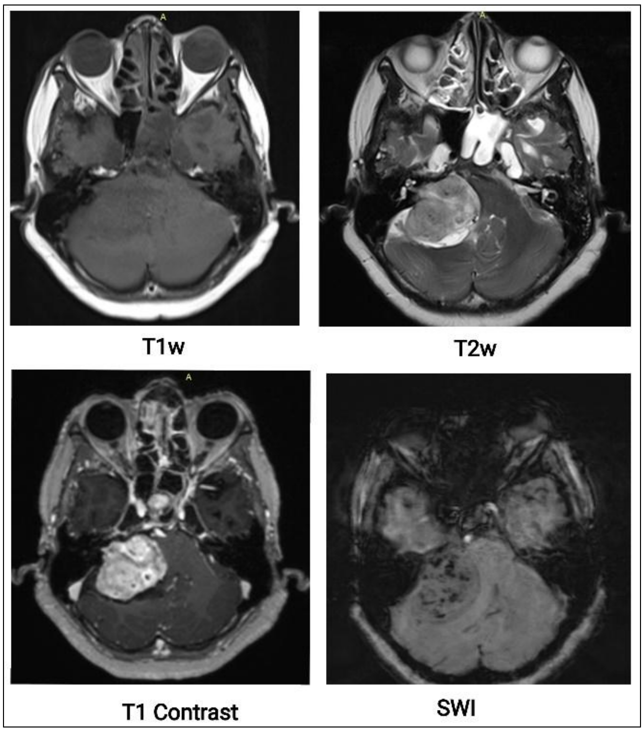

Fig 3: MR images of same patient showed lesion in right CP angle as iso-hypointense on T1-weighted images, hyperintense on T2-weighted images and showed intense nearly- enhancement on T1-contrast image. Few blooming foci were also seen on susceptibility-weighted images.

There was also note made of protrusion of left medial temporal lobe into left sphenoid sinus, through a small bony defect, that was suggestive of sphenoid encephalocele (Figure 6). Herniated left medial temporal lobe showed gliotic changes.

Fluid intensity opacification was also noted in left sphenoid sinus and the fluid was tracking into ethmoid sinuses and nasal cavity, explaining the patient's complaint of rhinorrhea.

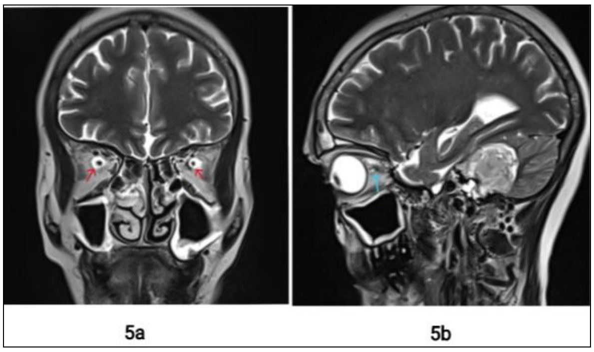

Fig 4: T2-weighted coronal and sagittal images of orbit shows prominent subarachnoid spaces around bilateral optic nerve (5a) with vertical tortuosity of optic nerves (5b).

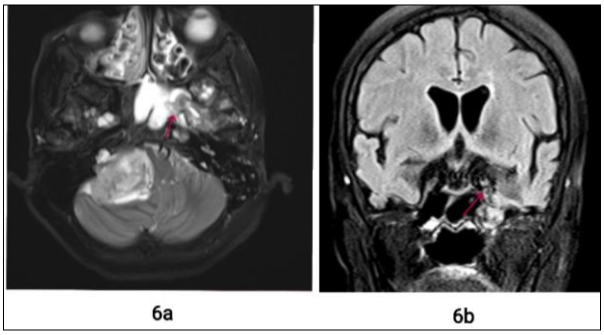

Fig 5: Note the protrusion of left medial temporal lobe into left sphenoid sinus, through a small bony defect (red arrow) - suggestive of sphenoid encephalocele

C. Conclusion of the imaging

Overall imaging findings concluded that right cerebello-pontine angle mass lesion, which most probably represents vestibular schwannoma, may have caused raised intracranial tension. This, raised intracranial pressure, may have eroded already thinned-out left lateral wall of sphenoid body due to aberrant arachnoid granulations and pneumatized lateral recess. Hence, sphenoid bony defects and raised intracranial pressure further precipitated sphenoid encephalocele and CSF leakage.

Discussion

Cerebrospinal fluid (CSF) rhinorrhea is passage of CSF through direct fistulous communication between nasal extracranial space and the subarachnoid space. It can occur due to congenital or acquired causes. Congenital causes include persistent craniopharyngeal canal, cribriform plate defect, or cephaloceles. Acquired causes of CSF rhinorrhea are divided into spontaneous, traumatic, inflammatory and neoplastic categories. Spontaneous and traumatic causes are most common reasons of CSF rhinorrhea, whereas later two are rarely cited as causes of CSF leaks [3].

Spontaneous CSF leaks are considered as primary CSF rhinorrhea and are commonly seen in middle-aged, obese women. Such cases are usually associated with idiopathic intracranial hypertension. Postulated hypothesis in such cases is that, long standing raised intracranial pressure causes bony erosion and attenuation of the adjacent investing membranes at the base of the skull [4]. The cribriform plate and lateral sphenoid walls are most common vulnerable sites for development of such fistulous communications. Trivial head trauma, forceful coughing and sneezing can complete the process in an already focally thinned out area of the skull base [4].

Presence of aberrant arachnoid granulations may be an independent causative factor of CSF rhinorrhea. It can also facilitate the osteo-dural fistula formation in cases with raised intracranial pressure. Arachnoid granulations, also called as pacchionian bodies, are villous structures, invaginating into the dural sinuses through a congenitally formed dural opening (porus villous arachnoidale), whose functions are to drain the CSF into the venous system. Normally, they are present in large number of clusters in adults [5].

Cranial AGs are usually situated close to the posterior half of the superior sagittal sinus and its adjacent venous lacunae, less commonly in contact with the transverse, petrosal, sigmoid, cavernous, and occipital sinuses. With increasing age, they cause absorption of the bone (compressive bone thinning) and thus produce the depressions on the inner table of the skull [5].

Sometime, during development of the CSF system, few of these granulations do not reach a venous target, and are called as aberrant arachnoid granulations. These are seen along the paramedian occipital squama, greater wing of sphenoid in middle cranial fossa, petromastoid plate, tegmen tympani, tegmen antri, tegmen mastoidea, and sigmoid sinus plate. When adjacent to the pneumatized parts of the skull, these aberrantly located arachnoid granulations may be responsible for spontaneous adult CSF leak or pneumocephalus [6, 7]. This hypothesis is supported by review of temporal bone series as well as several case reports by Gacek [8].

Gacek [8] also put a theory that progressive enlargement of these arachnoid granulations with increased age and CSF pressure is responsible for bone rarefaction or erosion, leading to the communication of the CSF with the pneumatic system of the temporal bone. Other factors that modulate CSF flow dynamics and pressure such as a physical effort, jugular venous pressure and subject's posture can also contribute in bone erosion around these arachnoid granulations.

Our case also had presence of such aberrant arachnoid granulations in greater wing and lateral wall of body of sphenoid bone. Few of such AGs on left lateral wall of sphenoid body may have progressed to osseous-dural defects due to long standing raised intracranial pressure, leading to the onset of CSF rhinorrhea.

Presence of large vestibular schwannoma in our case may have led to raised intracranial pressure. As vestibular schwannoma has usually slow growing nature, hence patient would have raised intracranial pressure from long time, which may have exaggerated the bone thinning and erosion around the aberrant arachnoid granulations at sphenoid body, and thus precipitated the CSF leakage through sphenoethmoid recess.

Large vestibular schwannoma can raise the intracranial pressure by three mechanisms- local mass effect, obstructive hydrocephalus due to compression of fourth ventricle, and communicating hydrocephalus due to excessive secretion of proteins by tumor cells, that blocks the arachnoid villi and reduces the CSF reabsorption [9]. The ventricular obstruction by large CP angle tumor may sufficiently raise intracranial pressure, without necessarily enlarging the ventricles beyond the normal range. The obstruction may be intermittent, but still be frequent and severe enough to produce features of raised ICP.

However, in our case, feature of raised ICP were quite prominent as evidenced by the degree of optic nerve tortuosity and significantly dilated Meckel’s cave as well as left sphenoid encephalocele. Hence, we hypothesized that, arachnoid villi blockage due to excessive protein secretion by tumor may had contributed to intracranial pressure, by reducing the CSF reabsorption.

Conclusions

CSF rhinorrhoea, though commonly associated with traumatic and iatrogenic causes, may rarely be associated with intracranial mass lesions. Hence, in cases of CSF rhinorrhoea, particularly with absent history of trauma or any operative intervention, imaging of brain should be advised to rule out any space occupying lesion. In present case, large right CP angle mass led to raised intracranial pressure, which created bony defects at hyperpneumatised sphenoid sinuses and aberrant arachnoid granulations. Leakage of CSF was noted through these bony defects explaining the patient's complaint.

List of abbreviations

CSF: Cerebrospinal fluid
CP: cerebello-pontine
CT: Computed tomography
MRI: Magnetic resonance imaging.

Conflict of Interest

Not available

Financial Support

Not available

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