

Primary Spontaneous Transclival Cerebrospinal Fluid Leak

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ABSTRACT : Cerebrospinal fluid leaks through the clivus are usually post-traumatic or occur secondary to lesions such as chordomas. The clivus is an uncommon site for a primary spontaneous CSF leak, with a PubMed search carried out in August 2014 revealing only fourteen such reported cases. We report our experience with two cases of primary spontaneous CSF leaks through the clivus, managed via endoscopic transsphenoidal approach.

Key Words : Cerebrospinal fluid leak, Spontaneous, clivus, endoscopic repair .

INTRODUCTION : Cerebrospinal fluid (CSF) leaks may be classified on the basis of etiology, anatomical site or intracranial pressure. Spontaneous CSF leaks are those for which no cause can be found. Some authors have used the term 'non-traumatic' instead of 'spontaneous' as they are of the opinion that there is no CSF leak that is truly 'spontaneous' in nature. Common sites of spontaneous CSF leaks are cribriform plate and the sphenoid sinus, especially in a pneumatized lateral recess¹. The clivus is an uncommon site for a primary spontaneous CSF leak, with a PubMed search carried out in August 2014 revealing only fourteen such reported cases²⁻⁷. We report our experience with two cases of primary spontaneous CSF leaks through the clivus, managed via an endoscopic trans-sphenoidal approach. **Case 1:** A 68-year-old obese, female presented with clear watery discharge from the left nostril since 4 months. There was no past history of head trauma. Clinical examination revealed no positive findings other than the watery discharge from the left nostril, which was confirmed to be CSF by a positive test for beta-2 transferrin. Computed tomographic (CT) cisternography revealed an empty sella and fluid in the left sphenoid sinus. A detailed reading of the scans however, revealed the site of leak to be in the clivus, but not from a single point (Figure 1). The patient was operated via an endoscopic trans-sphenoidal approach. On opening the sphenoid sinus, it was found to be filled with CSF, which was seen oozing out through the clivus, from multiple points, giving it an appearance of a 'watering can'. The defects were successfully repaired using a fascia lata graft and fibrin glue. This was further supported by oxidized cellulose and absorbable gelatin sponge, held in place by polyvinyl alcohol sponge pack. The CSF leak was thus successfully sealed and there has been no recurrence after 8 years followup.

Case 2: A 72-year-old obese, hypertensive female presented with clear watery discharge from the left nostril since 5-6 months. There was no past history of head trauma. The nasal fluid was confirmed to be CSF by a positive test for beta-2 transferrin. A CT Scan revealed a defect in the clivus with a small pseudomeningocele protruding out through the defect

(Figure 2). An empty sella was found to be present on magnetic resonance imaging (MRI). An endoscopic trans-sphenoidal approach was used to repair the defect. On entering the sphenoid sinus, the clival defect was visualized in the posterior wall, with a protruding pseudomeningocele and CSF leaking out. The pseudomeningocele was cauterized, the edges of the defect were defined, and a raw area created around the defect by baring the bone. The defect was then plugged using fat in a 'bath plug' fashion, held in place with fibrin glue, and oxidized cellulose and absorbable gelatin sponge. A non-absorbable polyvinyl sponge pack was placed to reinforce the repair. A post-operative CT scan revealed the fat plug in situ with successful sealing of the defect and leak. Long-term follow-up, however, could not be obtained in this patient as she succumbed to a myocardial infarction about a week after surgery. **DISCUSSION :** The incidence of primary spontaneous CSF leaks has been reported to be between 6%-40% of all CSF leaks^{8,9}, and are less common as compared to traumatic leaks¹⁰⁻¹¹. Various explanations have been put forth about the pathophysiology of spontaneous CSF leaks. These are usually seen in obese middle-aged females, with a raised body mass index, and are known to be associated with benign intracranial hypertension¹²⁻³. High-pressure CSF pulsations and chronic hydrostatic forces acting on thinned out skull base bone may result in defects, giving rise to CSF leaks. Empty sella, broadly attenuated skull base and arachnoid pits have also been found to be associated with spontaneous CSF leaks¹³. Menopause with its associated decrease in bone density and osteoporosis are other predisposing factors¹⁴. Transclival CSF leaks secondary to skull base lesions such as chordomas are known¹⁵⁻⁶. Primary spontaneous CSF fistulas through the clivus are however, very rare²⁻⁷. Various theories have been postulated to explain the occurrence of spontaneous clival CSF fistulas. The skull base including the clivus is formed via endochondral ossification. At birth the clivus consists of partially ossified components of the basiocciput and sphenoid body separated by the sphenoccipital synchondrosis. The age at which the sphenoccipital synchondrosis fuses is variable, although closure usually occurs by the



Fig-1: CT Cisternography (Patient 1) showing dye leaking through the clivus (white arrow). The arrowhead points towards the empty sella.

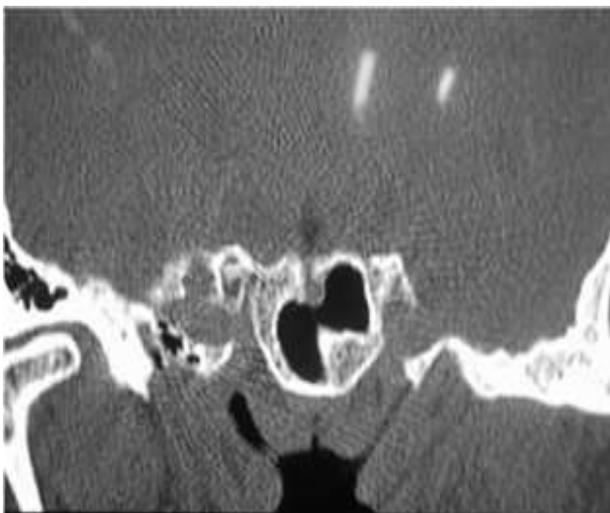


Fig-2 : CT Scan (Patient 2) showing a defect in the clivus with a protruding meningocele.

age of 17 years¹⁷. A developmental defect in this area could lead to a future area of dehiscence². It is suggested that spontaneous clival CSF leaks could occur due to erosion of the clivus secondary to repeated pulsations of the basilar artery. Repetitive increase in intracranial pressures acting on a congenitally thin bone, could eventually lead to a dehiscence and pseudomeningocele formation with a CSF fistula². Both our patients were post-menopausal, obese women with a demineralized, thinned out clivus. Also, hypertension in one of these patients could have resulted in increased arterial pressure, which over a period of time could have resulted in a bony defect. Both our patients had an empty sella, a common association with spontaneous CSF leaks. Investigation protocols for a patient with a spontaneous CSF leak are similar to those with any other type of leak and include detection of beta-2 transferrin or beta-trace protein in nasal fluid, high resolution CT scans, CT/MR

cisternography and intra-thecal fluorescein use in selected cases. In addition to this, bone density scans could be performed in patients with spontaneous leaks, to look for osteoporosis. Spontaneous CSF leaks are unlikely to heal with conservative measures, and usually require a surgical repair. All the previously reported spontaneous clival leaks have been repaired via a trans-sphenoidal route²⁻⁷. Adequate knowledge about the clival anatomy is necessary during surgery in this area, due to the close proximity of major blood vessels such as the vertical segment of the internal carotid artery. Also, a thick clivus can hold significant venous channels. The clival venous plexus extends along the basiocciput to the foramen magnum. Bone removal in this area should be done slowly as significant hemorrhage can occur. Occasionally, a drill may be required for bone removal. Both our patients underwent an effective repair of the clival defects via an endoscopic trans-sphenoidal approach.

CONCLUSION : Spontaneous clival leaks though rare, can be managed effectively via an endoscopic trans-sphenoidal approach. In case of CSF leaks within the sphenoid sinus, if the defect cannot be localized in the roof or the lateral recess, the clivus should be looked into as a possible site of leak.

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