Commentary

Intrasphenoidal encephaloceles are subdivided by location into medial perisellar and lateral sphenoid recess types.[1] Perisellar encephaloceles within the sphenoid sinus (SS) are considered more common, whereas basal encephaloceles limited to the lateral SS are rare. Intrasphenoidal encephaloceles are usually congenital in nature and occur supposedly due to presence of cricopharyngeal canal, which was described way back in 1888 by Sternberg.[2]

Lateral cricopharyngeal canal also known as Sternberg canal is formed due to incomplete fusion of greater wing with body of sphenoid bone. In presence of lateral recess, Sternberg canal can communicate with SS after its pneumatization and act as a site of origin of congenital neningoceles or cerebrospinal fluid (CSF) fistulas when there is sufficient SS pneumatization. Baranano *et al*. contradicts the above theory as they found presence of only one Sternberg canal in 1000 computed tomography (CT) scans examined.[3] They found that the development of arachnoid pits in the lateral recess of the SS, probably from underlying intracranial hypertension, was the major cause of intrasphenoidal encephaloceles.

Most patients present during childhood and abnormalities of face usually coexist with transsphenoidal encephaloceles, corresponding to median face-cleft syndrome. Cases diagnosed in adult life are rare and classically present with CSF rhinorrhea, emphasizing the importance of pneumatization of SS in the pathogenesis.[4] Nearly 45% of CSF leaks are high pressure because of associated hydrocephalous. Asymptomatic patients are usually diagnosed during imaging studies for other problems. Authors described similar case of intrasphenoial encephalocele who presented as a case of minor head injury and were diagnosed incidentally on CT examination.[5]

CT cisternography, 3D reconstructed multislice CT scan, and magnetic resonance imaging (MRI) provide excellent three-dimensional definition of the lesion and is useful for both diagnosis and surgical planning.[6] Intermittent or inactive CSF leaks are usually associated with a high incidence of false negative CT cisternography results and MRI may be a better choice in these patients. Endocrine assessment should be done in every patient as hypothalamic pituitary dysfunction is often found in these cases and deficiency of Antidiuretic hormone and growth hormone are the most common findings.[7]

Asymptomatic adult patients with true transsphenoidal meningoencephalocele should not undergo surgery, in view of complex anatomy of SS and very slow progression of the lesion. The treatment of symptomatic lesion is solely surgical and is indicated by the presence of persistent CSF rhinorrhoea, epipharyngeal respiratory obstruction, and progression of neurological deficient.

Both transcranial, endoscopic or combined approaches have been used for repair of these lesions. Transcranial route is preferred in CSF leaks involving the lateral recess of widely pneumatized SS to directly visualize the defect. Fronto-temporal craniotomy provides excellent access for middle cranial fossa. Access to the bony defect, particularly in the posterior portion, is difficult and may require sacrifice of prolapsed gliotic cerebral tissue. Perisellar lesions with small defects may be successfully obliterated by using traditional microscopic or endonasal transsphenoidal or transethmoidal approaches.

Initial attempts at repairing skull base defects in the lateral SS via a midline microsurgical transsphenoidal approach were associated with a high rate of failure, because of poor visualization of these defects. In most cases, the laterally based defect is situated beyond the line of sight of the speculum-based exposure. In

addition, a complete removal of mucosa of lateral recess of SS is not possible with midline approaches leading to failure in sinus obliteration. Wider exposures can be obtained with LeFort I osteotomy. However, these may be associated with postoperative malocclusion or traction neurapraxia of the infraorbital nerves as well as injury to the hard palate and upper alveolar region affecting the neurovascular supply to the teeth.

Endonasal transpterygoid approach through the posterior wall of maxillary sinus is a better technique in these cases as it allows exploration of most lateral aspect of SS, complete removal of sinus mucosa in lateral recess, and direct multilayered repair.[8]

Because of the underlying intracranial hypertension, direct repair without long-term CSF diversion would pose a high risk of postoperative CSF leakage. An initial ventriculoperitoneal shunt insertion is, thus preferred followed by endoscopic repair of the CSF leak and skull base defect via an endonasal transpterygoid approach. This technique is technically challenging and not completely sterile as it traverses the dirty nasal and paranasal cavities, nevertheless it offers avoidance of craniotomy and brain retraction, excellent panoramic visualization, improved illumination, and is preferred technique in hands of skilled neurosurgeons.

Overall determination of surgical approach is based on various factors including the degree of lateral sphenoid pneumatization, size and location of bony defect and ability to perform an adequate skull base repair through given exposure and although the endoscopic transpterygoid approach is probably the best technique, transcranial approaches are an optimal alternative when a transnasal approach had failed.

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