Recurrent spontaneous CSF leak in a patient with idiopathic intracranial hypertension

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Cerebrospinal fluid (CSF) leaks can appear anywhere in the cranio-spinal tract. According to the literature, CSF leaks can be divided into two forms: traumatic (most frequent, 90%) and spontaneous.

Traumatic etiologies include both accidental trauma and iatrogenic cranio-spinal injuries. Non-traumatic CSF leaks include tumoral causes, hydrocephaloceles, congenital abnormalities and focal atrophy. Spontaneous CSF leaks can be normal and high-pressure types. High-pressure leakages are related to long-term increases of the intracranial pressure (ICP) and they appear in 45% of spontaneous CSF leaks, while other patients have CSF leak associated with a normal ICP.

The main symptom of a cranial CSF leak is clear rhinorrhea. Anosmia is a common symptom that appears when the cribriform plate is affected. Meningitis is the most important risk that arises in 25-50% of untreated traumatic CSF leaks. The presence of recurrent meningitis could be the only sign of a non-traumatic CSF leak, as they have a 10% risk of meningitis per year. The pathogen agent most frequently involved in the etiology of meningitis in this group of patients is Streptococcus pneumoniae.

The therapeutic plan for patients with a CSF leak involves a correct diagnosis and localization of the dural defect. The standard imaging technique is high-resolution CT scan with fine coronal and axial sections and bone window settings. Recent studies have shown that MRI is also a very useful method, used for identification of the defect site. Patients with non-traumatic CSF leaks can be sometimes hard to diagnose and it is indicated to perform a radionuclide cisternography, along with nasal pledgets and different techniques for elevating intracranial pressure (head position and straining). In the cerebrospinal fluid, a specific marker exists, b2-tranferrin, used as a diagnostic tool for a CSF leak, as it has levels of almost 35 times higher in the cerebrospinal fluid, than in blood serum.

It is essential to identify the exact site of CSF leak in any case when we need to perform a surgical approach. After having a diagnosis of CSF leak, a really useful technique is nasal endoscopy, which is able to indicate the localization of the fistula, allowing also to realize an obliteration of the fistula, at the time same time of the endoscopic investigation.

Treatment methods must take into consideration the causes, the severity of the CSF leak, as they also rely on the moment and way of presentation to the doctor.

The initial therapy of patients with traumatic CSF leaks usually includes general methods (bed rest, elevation of the head), with a resolution in 70-85% of cases after seven days. Surgical approach is applied in case of traumatic CSF fistulas that do not have a proper response to non-operatives measures. Surgical procedures can be realized by intracranial or extracranial manoeuvres. Extracranial approaches allow access only to the paranasal sinuses and anterior cerebral fossa. The appearance of endoscopic methods allowed enlarged indications, with better functional results, in association with different extracranial approaches of CSF fistulas, with successful results in more than 90% of the cases. Unlike post-traumatic fistulas, spontaneous CSF leaks tend not to be resolved by non-operative interventions. The purpose of the treatment in this case is to identify the localization of the fistula and to treat the cause that determined this pathology.

We present a case of a 5-year-old patient that is known with idiopathic intracranial hypertension and has two episodes of meningitis in history. The patient was diagnosed with ethmoidal CSF leak, operated in May 2014 in another ENT Department, by endoscopic approach, when the defect was obliterated with autologous tissue with fibrin glue. It is also notable that in February 2015, the patient experienced an episode of acute meningitis with Streptococcus pneumoniae, as well as sepsis, for which he was hospitalized in a Clinical Infectious Diseases Unit. He was admitted in our clinic.
for the recurrence of rhinoliquorea at about one month postoperatively. We must note that the intracranial hypertension was poorly treated with acetazolamide; despite the current treatment, the patient presented numerous episodes of high intracerebral pressure.

The cranio-facial CT scan performed in March 2015 revealed enlarged right olfactory bulb, with an anterior-posterior diameter of 20 mm, prolapsing caudally, laterally from the insertion of the middle nasal concha, with a polylobulated outline. Caudally, it is continued with a fine dehiscent trajectory, immediately adjacent to the lamina papyracea, on the lateral part of the anterior pole of the superior concha; it also showed many arachnoid cysts in different regions of the brain. The MRI scanning from March 2015 was also evocative for an active CSF fistula, originating from the right side of a defect in the cribiform lamina (Figure 1).

After admission in our clinic, under general anaesthesia IOT, we performed a nasal endoscopic examination that revealed a CSF leak in the posterior third of the nasal septum, through two pores corresponding to the projection area of the ethmoid perpendicular plate and its insertion on the cribiform plate, a second appearance of CSF at the posterior third of the right superior nasal concha. The site of CSF leakage was obliterated with temporal muscle fascia, reinforced with fibrin glue and Surgicel®.

Figure 1 CT scan (A) and MRI (B) - enlarged right olfactory bulb, prolapsing caudally, laterally from the insertion of the middle nasal concha, with a polylobulated outline; fine dehiscent trajectory, adjacent to lamina papyracea, on the lateral part of the anterior pole of the superior concha

Figure 2 Nasal endoscopy – CSF leak in the posterior 1/3 of the nasal septum (A) and the posterior 1/3 of the right superior nasal concha (B)
Postoperative evolution was encumbered by the occurrence of an incident - voluntary release of nasal packing at approximately 4 hours after surgery, followed by the replacing of nasal packing and 2 episodes of vomiting. Despite these incidents, we have not found signs of meningeal irritation or nasal right CSF rhinorrhea. Nasal packing was released 48 hours postoperatively.

Postoperative nasal endoscopic examinations performed at 7 days, one month and two months did not reveal any obvious sign of CSF leak (Figure 3), aspect that was also confirmed by a cerebral MRI made at the 2-month follow-up.

Nevertheless, given the idiopathic intracranial hypertension, it is possible that the long-term evolution is unfavourable if the main condition is not treated properly with different methods of reducing intracranial pressure.

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