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CASE REPORT

Spontaneous CSF Leaks Caused by Double Skull Base Defects: Case Report

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Problems: This is a case report of a double spontaneous cerebrospinal leak. A 49-years-old female came to our attention for an intermittent clear rhinorrhea, with no other associated symptoms and a history of previous surgery for an anterior skull base leak.

Methodology: The rhinorrhea was considered at first as a consequence of the previous surgical repair, but the intraoperative fluorescein test showed another unexpected site of leak in the floor of the middle cranial fossa of the temporal bone.

Results: The bone defect was thus surgically repaired with success through a transmastoid approach.

Conclusions: This case-report offers matter of discussion about the possibility of double spontaneous cerebrospinal leak in the skull base, which should be always investigated with preoperative imaging both in the anterior and lateral skull base as to avoid erroneous diagnosis of site. The surgical options to repair anterior and lateral skull base defects are discussed as well.

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Introduction

Cerebrospinal fluid (CSF) leakages occur when there is a communication between the subarachnoid spaces and the skull base. Clear rhinorrhea or otorrhea can be triggered by skull trauma, surgical intervention, infection, or neoplastic invasion. Between 6% and 40% of all leaks occur spontaneously, in the absence of any apparent cause[1].

We report a case of a double skull base defect. To date, this is the seventh case of multiple location of spontaneous CSF leak reported in literature in the individual patient. Most of them originate from the anterior skull base. Though multiple spontaneous defects are unusual, the risk of misdiagnosis is not rare[2].

Radiological investigation is diagnostic in most of cases, but sometimes the anterior skull base defects are not evident and intraoperative fluorescein test is the option. The aim of our study is to add some information regarding the management of this rare pathology[3].

Case report

The case is about a 49-years-old female who came to our attention for intermittent clear rhinorrhea, with no other associated symptoms, obese, affected by

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thalassaemia minor and severely oligophrenic because of a neonatal meningitis of unknown pathogenesis.

Two years previously she had been affected by clear rhinorrhea and an anterior skull base CT scan had detected an ethmoidal roof bone defect which had been confirmed at intraoperative fluorescein test. The patient had been surgically treated with endoscopic technique in our Department and was asymptomatic for a couple of year. A new, presenting symptom of rhinorrhea was supposed to be a recurrence of the anterior skull base leak and was referred to our Department. A new CT scan was not repeated, due to the difficult clinical situation of the patient, that was claustrophobic and would have required a further general sedation. The patient was thus brought straight to surgery. Intraoperatively, sodic fluorescein test did not show any evidence of rhinoliquorrea from the anterior cranial fossa floor, nor from the site of the previous cranio-plasty, with a well continent ethmoidal graft. Conversely, a fluorescein leak was detected from the right tubaric ostium; the intraoperative micro-otoscopy showed CSF in the right middle ear. In this situation the patient could have been awakened, gone for a temporal bone CT scan then planning surgical repair of the temporal bone defect. Considering her clinical situation, including mental disorders and obesity, after discussing with the patient's parents the option of two procedures of anesthesia to obtain imaging and then surgery, we agreed to go straight for surgery. A retroauricular mastoidectomy was performed and after drilling the cortical bone, a multiple bone defect was found, extending from the antrum to the epytimpanum. Arachnoid exposure with CFS leak was evident in different sites, since dura was disrupted in different areas with irregular margins. The residual bone among the sites of leak was drilled and removed in order to obtain a single, wide bone defect with clear margins. The ossicular chain was left in situ, though this made the exposure of the tegmental bone more difficult. Dura was replaced with a wide shell of autologous fascia lata which was inserted under the bony margins of tegmen. Bone paté was harvested with fibrin glue and fixed the fascia lata from below. Another shell of fascia lata was placed overlay, extending on the esocranic bony surface of the tegmen. Fibrin glue and readsorbed packing was placed to reinforce the plasty. The cavity of mastoidectomy was closed like in a canal wall up technique. Nasal packing and bed rest were mantained for two days. No lumbar drain was placed but diuretic drugs were administered.

Discussion

In our case a wrong preoperative diagnosis was made. One year after the first operation on the ethmoidal roof leak, the second episode of spontaneous leak was supposed to be a recurrence of the anterior skull base defect. A new CT and MRI would have been necessary since multiple spontaneous leaks are rare, but if not properly diagnosed, inefficacious surgery in the wrong site would be performed[1,2]. We managed this case differently from the routine practice, since the disability of the patient and the need to perform each examination under general anesthesia made everything more difficult and influenced our conduct. We renounce to perform new preoperative imaging, as we based on the misleading belief of a recurrent leak from the site which had been repaired two years before.

We brought the patient straight to surgery. No leak was evident from the anterior skull base but intraoperative Fluorescein was localized in Eustachian tube, though it did not added more precise details about the site of the temporal bone defects [4,5]. Again, a whole skull base CT scan should have been performed before the decision to perform surgery in the temporal bone, in order to assess properly the site and size of the defect, rule out herniation and plan the surgical repair. We discuss with the family the opportunity to wake the patient up and study the case with proper imaging under general anhesthesia or to continue surgery and go straight in the temporal bone to understand and solve the problem. We decided to do an immediate surgical exploration of the tegmen antri, tegmen tympani and, if necessary, to repair immediately the defect.

Although not conventional and patient related, our decision making was efficacious for the good outcome of the disease. The large bone defect at the temporal bone was repaired and the follow up of the patient showed no recurrence of the disease after one year. The disability of the patient was misleading in our management and strongly influenced our wrong preoperative diagnosis, nevertheless the possibility of

a double, spontaneous CFS leak in the skull base is a very rare event. This gave us the opportunity to investigate and review the management of multiple spontaneous leaks of the skull base.

To date, six cases of anterior and lateral skull base defect occurring in the same patient have been described [6]. A combination of congenital and acquired factors have been proposed to explain the aetiology of these multiple lesions, such as the presence of aberrant arachnoid granulations at the temporal bone, congenital focal atrophy in the anterior skull base, increased intracranial pressure, radiological empty sella, obesity and age over the seventies[7,8]. The skull base defect may develop for all these reasons in the thinnest bone walls. Our patient suffered from meningitis some years before rhinorrhea started: inflammatory dural changes might relate to the multiple sites of skull base defect.

In case of spontaneous CSF leak, a CT scan of the whole skull base is mandatory and more than one defect should be searched. Fluorescein test should be adopted routinely[4,5] first to confirm the site of a spontaneous CSF leak to repair, then in case of recurrence, to assess the real site of origin of rhinorrhea, and to rule out that no other defects were eventually missed at the CT. The longest time interval reported between recurrent spontaneous leak presentation is four years; therefore, long and careful clinical and radiological follow-up is necessary.

The best and nowadays more diffuse approach to repair the anterior skull base defect is the endonasal endoscopic technique[5,9-11]. It offers advantages if compared to craniotomy, allowing a less invasive approach, with a reduction of hospitalization, and a more rapid functional recovery. An optimal visualization of the field of action shows the exact location and size of the dural defect that is strictly necessary to a proper repair, with a graft matching precisely the size of the lack. Once the dural defect has been exposed, the area is prepared by dissection of the surrounding mucosa. The endoscopic approach is different depending on the location of the CSF fistula and the anatomical features of the surrounding areas, with a considerable variability from person to person[10,11].

A spontaneous leak located in the olfactory groove, ethmoid and sphenoid can be treated with an endoscopic approach. A lower probability of success has been demonstrated for defects of the frontal sinus[10,11], needing an open treatment.

For the bone defects in the temporal bone there are different options of closure [12-4]. Little defects are reached via a transmastoid approach. A mastoidectomy with opening of the antrum exposes the posterior wall of the external auditory canal anteriorly, the sigmoid sinus posteriorly and the floor of the middle cranial fossa superiorly. Tegmen antri and tympani are exposed and if bone dehiscence is evident, a single bone defect with clear margins has to be obtained by drilling. Dura, though disrupted, must be exposed in all the area and has to be further detached all along the bony margins of the defect. The graft has to be bigger than the bony defect as it must be placed safely on the bony margins of the middle cranial fossa, under the dura of the temporal lobe, with an underlay technique. The dural defect can be reconstructed with a layer of fascia lata or different synthetic material. Another sheet of fascia lata or other material is then placed overlay from the mastoid, followed by bone paté and fibrin glue to fix the plasty.[14-16]

Fascia lata is considered the best graft material: it is resistant, easy to harvest and seems to promote a fast replacement through cellular outspread from the margins of the dura tear. The cavity of mastoidectomy can be obliterated with fibro-periosteal or temporalis muscle flap. It can be rotated antero-medially to fill the antrum and reinforce the plasty along the tegmen tympani furtherly. When a defect involves the anterior epitympanum the sacrifice of the incus and manubrium is necessary, involving a conductive hearing loss [14-17].

The transmastoid technique is easy to perform but sometimes it does not offer adequate exposure of the defect and safe reconstruction may be affected. This approach is suitable for small-medium size and laterally-placed defects in the tegmen tympani.

For large bone defects or for medially located dehiscences of the floor of the temporal bone a subtemporal approach is more indicated unless there are contraindications. The middle cranial fossa approach is

suitable for small defects as well, but the slight retraction of the temporal lobe required by the approach itself relates to a risk of potential morbidity, especially in aged patients and when co-morbidities are associated. Defects in microcirculation and anti-trombotic drugs assumption may relate to the surgical risk of temporal lobe hematoma and oedema. A small temporal craniotomy with slight retraction of the temporal lobe is required, to detach the dura and expose the defect from above[18]. The size of the defect does not interfere with the efficacy of the procedure, as the exposure of the floor of the middle cranial fossa is excellent from the subtemporal approach and is not influenced by any of the conditions of the middle ear, like flogosis or infection.

The ossicular chain is preserved. After detaching the dura from the floor of the middle cranial fossa a large layer of fascia lata is placed over the bony margins of the defect, widely covering them. Bone paté and fibrin glue fix the plasty from above. The temporal lobe is replaced over the bony floor of the middle cranial fossa and contributes to reinforce the plasty. This approach has excellent results in terms of efficacy of the closure of tegmen tympani, is the advised procedure in large defects or when the mastoid is not free from pathology. When co-morbidities, age and drugs-related risk of bleeding are not in favor of it and the transmastoid approach is not safe, lateral petrosectomy is another option to close bony defects in the floor of the middle cranial fossa. The technique involves the traditional transmastoid approach, exposure of the bone defect and dura tears, plasty with fascia lata from below with the underlay technique, reinforcement with bone pate and fibrin glue. The procedure is safe and offers durable results, but the whole middle ear is excluded from communication with the nose. The tympanic cavity is exenterated, the Eustachian tube is closed, the mastoid and all the cavity is filled with fat. It is not a first choice procedure as it involves conductive hearing loss and the aestethic defect of the cul-de-sac closure of the external ear. It can be an option to repair temporal bone CFS leak when concomitant cholesteatoma or inflammatory disease of the middle ear/mastoid cavity are evident and when middle cranial fossa approach is not advisable for its potential surgical morbidity.

Our case suggests that when a recurrent spontaneous CSF leak occurs a second skull base leak site must be investigated since temporal bone disruption may be present and be the hidden cause of leak. The surgical options to repair skull base defects have been exposed both in the anterior and lateral skull base defects. The principles driving the choice of the most appropriate technique relates to the procedure which proves to have the minimal surgical morbidity and the highest likeliness to be efficacious and long-lasting. It may happen, as in our case, to adapt the diagnostic assessment and the therapeutic planning to the clinical conditions of the patient.

Conclusions

Though rare [3], multiple spontaneous skull base defects are to be looked for and the middle cranial fossa floor has always to be investigated together with the anterior skull base. High resolution CT scan of the skull base is indicated as first choice diagnostic step, focusing on bone defects at the tegmen tympani, ethmoid bone and floor of the sphenoidal sinus. MRI is added to complete the diagnosis and rule out the possibility of cerebral herniation and has to investigate both the anterior and middle cranial fossa. The radiological imaging suggesting the site of leak is confirmed with intraoperative fluorescein test, which plays a adjunctive role in showing the site of the CFS leak. It is routinely used even if there is no general consensus about absence of side effects, some authors [4] suggesting to reserve it only to those cases without a preoperative diagnosis. Lumbar drain is not routinely used but it may be an option in presence of high-pressure CFS leak.

When small bone defects are accidentally detected by imaging without any clinical sign of rhinorrhea neither meningocele, the need of surgically repair them is still a matter of discussion.

Though aetiology of spontaneous multiple skull base CFS leaks is not yet well understood, our contribution is to support the need of a good radiological preoperative diagnosis, not to miss any site of origin, discuss the best surgical option and recommend regular and long lasting follow-up. Our misleading conduct was influenced by the disability of the patient, as we renounce to perform proper preoperative imaging in favour of an immediate surgical treatment. Although surgery was successful and, in the case of the patient, the problem was solved with a

single anesthesia and procedure, our management should not be the routine practice. Unlike what we did in this particular case, we strongly recommend to always investigate the whole skull base with CT scan and MRI of the anterior and lateral skull base, rule out the presence of double site defects and avoid inefficacious and non-definitive surgery.

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