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Small spheno-ethmoidal meningoencephalocele versus ethmoidal mucocele in spontaneous intracranial hypotension

Giulio Bonomo [a,b,c,1], Gennaro Bussone [d,1], Alessandro Gans [a,b], Guglielmo Iess [a,b], Roberta Bonomo [d,e,*], Francesco Restelli [a,b], Jacopo Falco [a,b], Elio Mazzapicchi [a,b], Mario Stanziano [f,g], Alessia Amato [h], Morgan Broggi [a], Francesco Acerbi [a,b], Paolo Ferroli [a], Marco Schiariti [a]

[a] Department of Neurosurgery, Fondazione IRCCS Istituto Neurologico C. Besta, Milan, Italy
[b] University of Milan, Milan, Italy
[c] Department of Neurological Surgery, Policlinico "G. Rodolico-S. Marco" University Hospital, Catania, Italy
[d] Department of Neurology, Fondazione IRCCS Istituto Neurologico C. Besta, Milan, Italy
[e] School of Medicine and Surgery, Kore University of Enna, Enna, Italy
[f] Neuroradiology Unit, Diagnostic and Technology Department, Fondazione IRCCS Istituto Neurologico C. Besta, Milan, Italy
[g] Neurosciences Department "Rita Levi Montalcini", University of Turin, Turin, Italy
[h] Department of Child Neuropsychiatry, Fondazione IRCCS Istituto Neurologico C. Besta, Milan, Italy

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ABSTRACT

Background: Meningoencephalocele is defined as an abnormal sac of brain tissue and meninges extending beyond natural skull margins, often leading to cerebrospinal fluid (CSF) leakage. When this condition arises in the spheno-ethmoidal region, the diagnosis becomes more challenging as it can be mistaken for other nasal pathologies, such as mucocele.
Research question: We show in this case report a non-congenital sphenoethmoidal meningoencephalocele causing rhinoliquoral fistula and spontaneous intracranial hypotension.
Results: this 65-year-old woman presented with sporadic rhinoliquorrhoea associated with orthostatic headache, nausea and dizziness. Brain MRI revealed a small lesion of an ethmoidal sinus, which was successfully treated with endoscopic endonasal surgery. Histology confirmed the presence of meningoencephalic tissue positive for S100 protein on immunohistochemistry.
Conclusions: When dealing with lesions of the paranasal sinuses in contact with the anterior skull base, rhinoliquorrhoea presence suggests meningoencephalocele. In dubious cases, a proper workup, including a thorough clinical history and neurological examination, specific imaging, and a direct search of CSF-like markers, is essential to support the differential diagnosis. In such cases, a transnasal endoscopic surgical approach is recommended to obtain a final histological diagnosis and to perform eventual dural plastic surgery.

1. Introduction

The herniation of cranial contents through skull bone defects is called an encephalocele. If the herniated content is limited to the meninges, it is described as a meningocele; if the brain parenchyma is involved as well, this is known as a meningoencephalocele (Ziade et al.,

Abbreviations: CSF, cerebrospinal fluid; CISS, Constructive Interference in Steady State; CT, computed tomography; HRCT, High-Resolution CT; ICHD-3, International Classification of Headache Disorders; MRI, magnetic resonance imaging; PSIR, Phase-Sensitive Inversion Recovery; SIH, spontaneous intracranial hypotension; MAV, Artero-Venous malformation.

* Corresponding author. Department of Neurology, Fondazione IRCCS Istituto Neurologico C. Besta, Via Giovanni Celoria 11, 20133, Milan, Italy.
E-mail addresses: dott.giuliobonomo@gmail.com (G. Bonomo), studiobussone@gmail.com (G. Bussone), gans.alessandro@gmail.com (A. Gans), guglielmoiess@gmail.com (G. Iess), roberta.bonomo@nhs.net (R. Bonomo), francesco.restelli91@gmail.com (F. Restelli), jacopofalco910@gmail.com (J. Falco), elio.mazzapicchi@unimi.it (E. Mazzapicchi), stanzianomario@gmail.com (M. Stanziano), dott.alessiaamato@gmail.com (A. Amato), morgan.broggi@istituto-besta.it (M. Broggi), francesco.acerbi@istituto-besta.it (F. Acerbi), paolo.ferroli@istituto-besta.it (P. Ferroli), marco.schiariti@istituto-besta.it (M. Schiariti).
[1] These authors contributed equally to this work.

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2016). The earliest medical mention of encephalocele appeared during the 16th century, illustrating a stillbirth with encephalocele and severe deformities, later identified as Robert's syndrome (Abdel-Aziz et al., 2010).

Encephaloceles can be classified considering anatomical position and aethiology (Sasani et al., 2009; Leland Albright et al., 2015). Nasal encephaloceles are most commonly congenital and seen during childhood (Abdel-Aziz et al., 2010; Tirumandas et al., 2013; Vercler et al., 2014; Chatterjee et al., 2017; Gaab, 2016) arising with CSF rhinorrhea and/or nasal obstruction. Other etiologies occurring mostly during adulthood are infection, trauma, or tumors (Formica et al., 2002). If no known cause is described, encephaloceles are defined as spontaneous (McPheeters et al., 2015; Tomaszewska et al., 2015).

The most common primary clinical symptom of nasal encephaloceles is spontaneous cerebrospinal fluid (CSF) leakage, which manifests as clear nasal fluid and nasal obstruction and is occasionally ignored (Gaab, 2016; Zoli et al., 2016). Inappropriate handling can lead to seizures and meningitis (Tirumandas et al., 2013; Zoli et al., 2016; Fernández-Gajardo et al., 2018; Schlosser and Bolger, 2002; Lanza et al., 1996). CSF leakage may determine spontaneous intracranial hypotension (SIH) commonly due to cervicothoracic thecal sac defects or an unidentifiable cause in 44–55% of cases. Intracranial dural defects may lead to CSF leakage however, to date, there is no definite evidence of correlation with SIH (Schievink et al., 2012; Beckhardt et al., 1991; Rgen Beck et al., 2018). SIH predominant symptom is positional headache, worsening when standing and subsiding when lying down, possibly due to distortion of pain-sensitive intracranial structures by CSF hypovolemia. Other symptoms, such as pain or stiffness in the neck, nausea, emesis, tinnitus, and dizziness, are less frequently observed (Mokri, 2013; Higgins et al., 2020; Schievink, 2003; Portier et al., 2002).

Considering the condition's rarity, the literature regarding the adequate treatment is still limited and heterogeneous (Tomazic and Stammberger, 2009; Socher et al., 2008; Myssiorek and Cohen, 1987; d'Ávila et al., 2014; Castelnuovo et al., 2010; Blaivie et al., 2006). The introduction of functional endoscopic surgery for paranasal sinuses and the development of extended, endoscopic transsphenoidal surgery, which provides a broader operative view to resect parasellar tumors, has made feasible the repair of CSF leakage from anterior skull base defects (Castelnuovo et al., 2010; Ma et al., 2015; Cavallo et al., 2007; Wigand, 1981).

A literature review was conducted on PubMed and Cochrane database using the terms "sphenoethmoidal encephalocele", "sphenoethmoidal meningoencephalocele", "spheno-ethmoidal encephalocele", "spheno-ethmoidal meningoencephalocele", "traumatic basal meningoencephalocele", "traumatic basal encephalocele", "sphenoethmoidal rhinoliquorrhoea" without limitations. Of 70 articles found, no article mentioned acquired sphenoethmoidal meningoencephalocele. Basing on this literature review, to the best of our knowledge, we report the first case of acquired sphenoethmoidal meningoencephalocele causing inconstant and subclinical rhinoliquoral fistula responsible for SIH.

2. Case presentation

We present the case of a 65-year-old right-handed woman complaining of orthostatic headache, nausea, and dizziness for several years, exacerbated during the last year. Neurological examination revealed a headache aggravation with the Valsalva maneuver and slight gait instability. Occasional nose leakage with retropharyngeal discharge sensation and salty taste was referred. She also reported involvement in a road accident about ten years ago, probably caused by a seizure with loss of consciousness. Since then, she has been taking levetiracetam therapy with no further crises. No other relevant comorbidities have been reported, besides arterial hypertension and osteoporosis.

An electroencephalogram did not detect any abnormal activity. Brain 1.5T Magnetic Resonance Imaging (MRI) showed very subtle marks of intracranial hypotension such as prominence of inferior intercavernous sinuses with redundant pituitary contrast enhancement (elsewhere mistaken for a pituitary lesion), slight reduction in ponto-mesencephalic angle and faintly visible brainstem sagging (Fig. 1 A). Considering the MRI-based SIH score (bSIH) applied to our case we found a high probability of CSF loss (total score 8; 2.16mm suprasellar cistern, 2.9mm prepontine cistern, 3.68mm mamillopontine distance; Fig. 2) (Dobrocky et al., 2022). In addition, a small cystic lesion (about 5 mm in size) was found close to the medial wall of the left orbit claiming for the differential diagnosis between meningoencephalocele of the left spheno-ethmoidal planum and mucocele with bone remodelling (Fig. 1 B). In particular, in the MRI T2-weighted images and computed tomography (CT) scan with bone reconstruction algorithm it was possible to notice, mainly for asymmetry, a focal bone thinning with possible interruption just above the small cystic lesion (Fig. 1 C–F). A diagnostic integration with Phase-Sensitive Inversion Recovery (PSIR) and post-contrast T1-weighted sequences on 3T MRI was performed, with the latter confirming the hypothesis of meningoencephalocele as more likely (Fig. 1 G, H). Finally, she underwent a spinal MRI which demonstrated no findings consistent with a CSF leak. Lumbar puncture showed a low opening pressure (3 cm H2O). In October 2021, the case was discussed by a multidisciplinary committee which set the indication for trans-nasoethmoidal endoscopic exploration to confirm the meningoencephalocele and eventual repair of the dural fistula. The patient gave written informed consent for the procedure. On the day of surgery, the patient was positioned supine and, after intrathecal fluorescein administration by lumbar injection (50 mg, 1 ml, 5% solution), an endoscopic exploration of the left ethmoidal sinuses was performed and, on the roof, a small fluorescent lesion was confirmed which was excised revealing a small osteodural cleft (Fig. 1 I). Then, we proceeded by drilling the sphenoethmoidal planum, and the dural defect was sealed with a combination of fat tissue harvested from an abdominal incision, collagen-based dural grafts, and fibrinogen- and thrombin-based sealant matrices (Fig. 1 J). Histology confirmed the presence of meningoencephalic tissue positive for S100 protein on immunohistochemistry. The postoperative course was uneventful and characterized by early autonomous mobilization. The patient was discharged from our Institute two days after the surgical procedure. She exhibited no signs or symptoms of intracranial hypotension at three months of follow-up. In addition, she never reported any signs of rhinoliquorrhoea post-operatively.

3. Discussion

Encephalocele is mainly a congenital type of neural tube defect, where a sack containing brain-meninges-CSF forms outside the skull through a bony defect. When the sac is composed of the protruding meninges and CSF, it is most appropriately termed a meningocele, whereas it is known as an encephalocele if containing brain tissue (Ziade et al., 2016).

The underlying mechanism of congenital encephalocele formation has not been fully established yet. Nevertheless, several theories have been proposed, considering multifactorial pathogenesis involving genetics and environmental factors (Cinalli et al., 2019). Some studies have suggested a role of vitamin deficiency during childhood, viral infections, hypoxia, irradiation, hyperthermia, folic acid therapy during pregnancy, insufficient apoptosis between neural and surface ectoderm, impaired mesoderm, amniotic band syndrome, and anomalies in Sternberg's canal (Tirumandas et al., 2013; Tomaszewska et al., 2015; Zoli et al., 2016; Morina et al., 2011; Marin-Padilla, 1980; Chen and Gonzalez, 1987). Other theories consider an early neural tube outgrowth through an arrested frontal floor bone closure in the development of nasal encephaloceles (Tirumandas et al., 2013). Posteriorly to the cribriform plate, the herniation of meninges and brain parenchyma arises basal meningoencephaloceles, such as transethmoidal and sphenoethmoidal meningoencephaloceles, the latter developing through the posterior nasal cavity in the sphenoethmoidal junction (Tirumandas

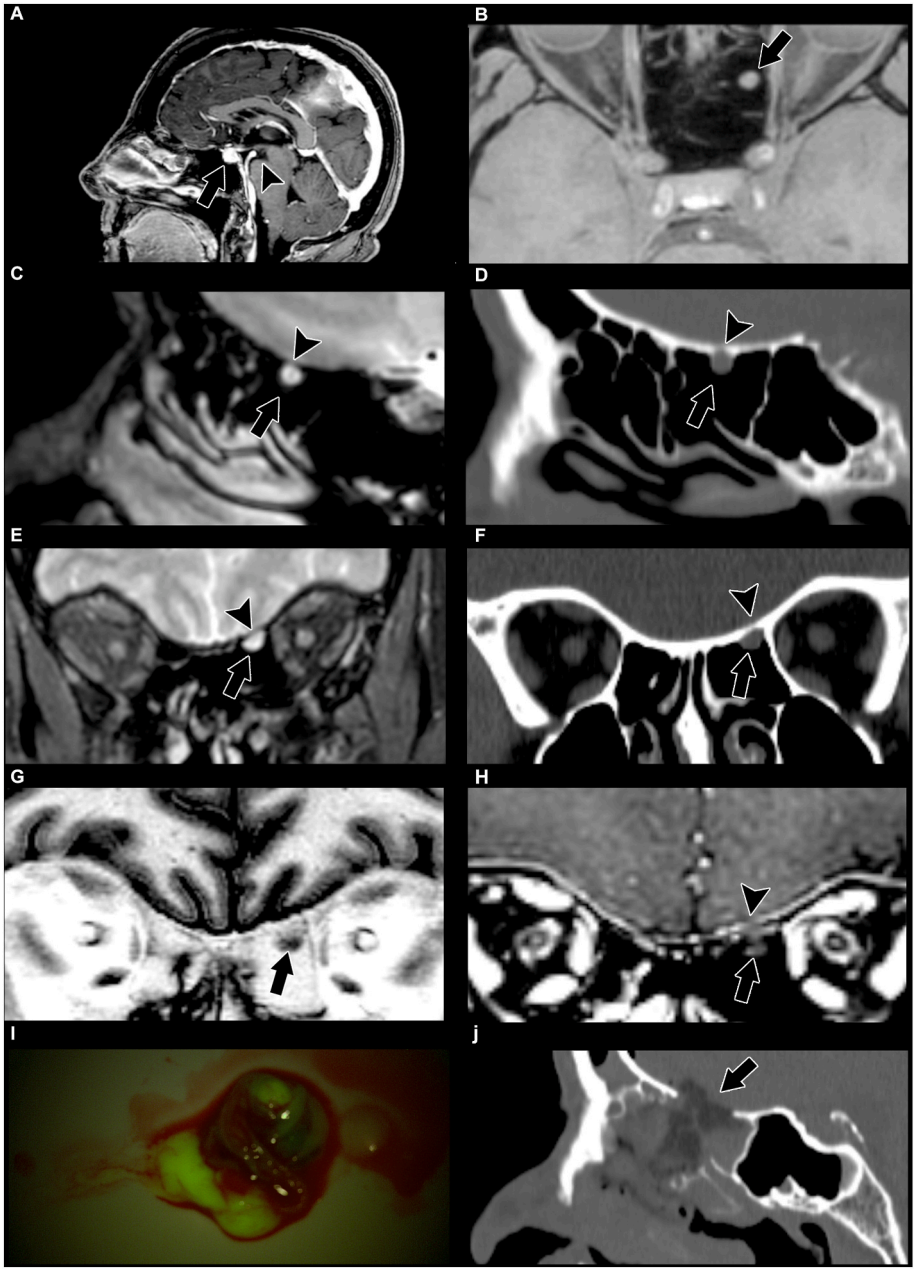

Fig. 1. A) Volumetric T1-weighted post-contrast (gadolinium) brain 1.5T MRI showing very subtle marks of intracranial hypotension in the sagittal plane, such as redundant pituitary enhancement (arrow) and slight reduction in ponto-mesencephalic angle (arrowhead). **B)** Volumetric T1-weighted 1.5T MRI with fat saturation without contrast showing on the axial plane a small cystic lesion close to the medial wall of the left orbit (arrow). **C), D), E)** Volumetric T2-weigthed 1.5T MRI images and **F)** volumetric computed tomography (CT) scan with bone reconstruction algorithm showing both on the sagittal and coronal plane a focal bone thinning with possible interruption (arrowhead) just above the small lesion (arrow). **G), H)** Phase-Sensitive Inversion Recovery (PSIR) and post-contrast T1-weighted 3T MRI images on the coronal plane showing that the cystic lesion (arrow) was associated with focal dural thinning or possibly interruption along the spheno-ethmoidal planum (arrowhead). **I)** Microscope view (Pentero Microscope with YELLOW 560 filter) demonstrating the fluorescence of the freshly excised lesion on the roof of an ethmoidal sinus after intrathecal fluorescein administration. **J)** Postoperative volumetric CT scan with bone reconstruction algorithm showing on the sagittal plane the surgical repair of the ostodural cleft. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

et al., 2013; Wacharasindhu et al., 2005). Some authors suggest that spontaneous encephaloceles may be post-traumatic, with the traumatic event not being recalled or considered irrelevant by the patients (McPheeters et al., 2015). Similarly, some patients may present occult skull defects being enlarged by trauma, thus leading to herniation and encephalocele development (Xue et al., 2020). Non-congenital encephaloceles usually develop in adulthood and occur either as a result of known triggering factors such as trauma, tumors, iatrogenic conditions or secondary to undetermined causes (Formica et al., 2002). In our case, as demonstrated by the histological findings, it was a small meningoencephalocele possibly of traumatic nature given the small size, the age of manifestation, and the temporal correlation of SIH symptoms with the car accident ten years earlier. Moreover, the hypothesis that seizure was triggered by dysplastic nerve tissue in the meningoencephalocele, as suggested in the literature, cannot be excluded (Gaab, 2016).

A basal meningoencephalocele can be responsible for a CSF fistula, commonly presenting as a unilateral watery positional rhinorrhea, nasal

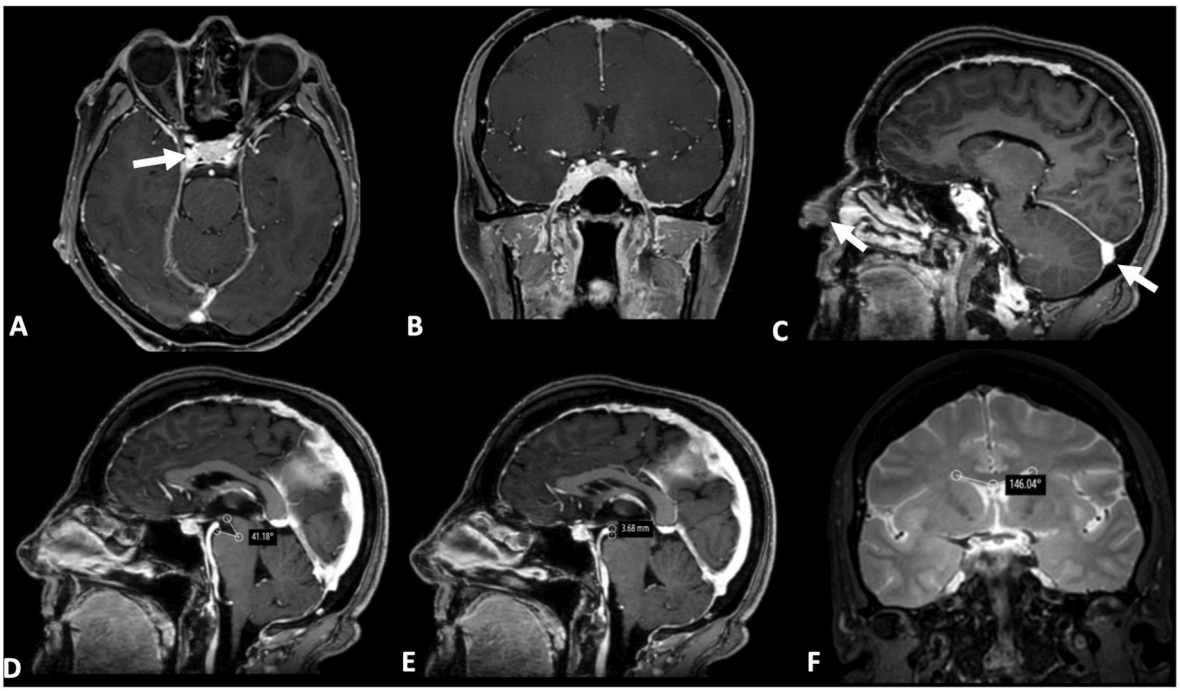

Fig. 2. Volumetric T1-weighted post-contrast (gadolinium) brain 1.5T MRI showing signs of intracranial hypotension: pituitary hyperemia (arrow) (**A**), pachymeningeal enhancement (**A, B, C**), venous distention sign with convexity of inferior contour of dominant transverse sinus (arrow) (**C**), abnormal pontomesencephalic (**D**) and lateral ventricular (T2-weighted) (**F**) angles, mamillopontine distance (**E**).

congestion, and sometimes salty taste in the mouth (Gaab, 2016; Zoli et al., 2016). However, other rhinological conditions, including seasonal allergic rhinitis, perennial non-allergic rhinitis, and vasomotor rhinitis, are common and may mimic some of the signs and symptoms of CSF rhinorrhea or may even co-occur with CSF leakage. One rare cause of rhinoliquorrhoea might also be the presence of vascular malformations such as artero-venous malformations (MAVs) (Ramírez-Ferrer et al., 2023).

The presence of a halo sign (a clear annulus encircling a central bloodstain) on gauze, tissue, or linen has been traditionally used as a marker to predict CSF loss; however, this test should only be used to raise initial suspicion (Kerr JT Chu et al., 2005; Kirsch, 1967; Katz and Kaplan, 1985). The measurement of rhinorrhea components may be useful, though sensitivity and specificity are unacceptably low. B2-Transferrin has become established as a highly sensitive and specific way of identifying CSF and is now the preferred method for confirming a fluid as CSF. Beta-trace protein is another marker that has been used to identify CSF. Upon identification of a traumatic CSF leak, nasal endoscopy should be performed. Albeit direct visualization plays an important role, skull base imaging is crucial to localize CSF leaks, especially those that are traumatic. Imaging includes High-Resolution CT scan (HRCT), intrathecal fluorescein, CT Cisternograms, Radionuclide Cisternograms, MRI and MR Cisternograms (Prosser et al., 2011; Eljazzar et al., 2019). HRTC is recommended early in the diagnostic workup to evaluate any skull base bone lesion (including the petrous bone); thereafter, MRI can be used to reveal meningocele, meningoencephalocele, inflammatory reaction of the nasal mucosa or middle ear, and any meningeal or intracranial involvement (e.g., meningitis or meningoencephalitis, etc.). MRI is also relevant in cases of spontaneous CSF leakage to suggest concomitant SIH since more than 80% of suspected patients show the typical signs: subdural fluid collections, pachymeningeal enhancement, venous structures' engorgement, pituitary hyperemia and sagging of the brain (Schievink, 2003).

According to previous literature, CSF fistulas at the skull base are not typically associated with the development of intracranial hypotension, and in the case of SIH, it is suggested to look for CSF leakage at the spinal level (Schievink et al., 2012; Rgen Beck et al., 2018; Ducros and Biousse, 2015). Rhinoliquorrhoea is often intermittent, positional, of low volume, secondary to low subarachnoid cistern pressure, and may not be expected to result in a CSF leak sufficient to cause SIH. In our case rhinoliquorrhoea was reported as minimal and sporadic by the patient and was not present at the time of the examination. However, typical radiological signs and symptoms of SIH were present, meeting all ICHD-3 criteria (Olesen, 2018). Based on this suspicion and the evidence brought by the MRI (small lesion of an ethmoidal sinus compatible with a meningoencephalocele), we performed a transnasal exploration to repair a possible osteodural defect responsible for the CSF leak and consequent SIH. To the best of our knowledge, the reported case may be the first to link a sphenoethmoidal meningoencephalocele with the development of intracranial hypotension.

The main pathology in the differential diagnosis of basal meningoencephalocele is mucocele. Sinusal mucocele is a cyst, enclosed by respiratory epithelium and mucus-filled, which arises from the paranasal sinus and is likely to expand (Natvig and Larsen, 1978). The condition is relatively rare, with prevalent onset in adulthood. In 90% of all cases, the frontal sinus and frontoethmoidal region are involved (Scangas et al., 2013). Symptoms are mild or absent, with the typical presentation of axial or non-axial exophthalmos sometimes associated with lateral diplopia. Rarely it may also include a frontal headache or supraorbital skin swelling. With imaging techniques, diagnosis is generally made before the onset of any complications. Most mucoceles, particularly those occurring in the frontal sinus, may be diagnosed using CT or plain radiography. MRI is only recommended when uncertainty persists after using these techniques. The usual signaling characteristics are T1 hypointensity and T2 hyperintensity. As a rule, the older the mucocele, the shorter the T1 relaxation time. Nevertheless, any signal combination might be observed due to the presence of blood products or degree of hydration. In more uncertain cases, gadolinium-enhanced MRI is a more reliable tool for diagnosis. The lack of contrast enhancement confirms the presence of fluid content (Lloyd et al., 2000). Sinus mucocele pathophysiology is complex and may include obstruction of a sinus ostium and mucosal inflammation (Lund et al., 1993). The cultures

of mucocele contents are not always sterile and often show aerobic and rarely anaerobic pathogens (Brook and Frazier, 2001). Predisposing factors include trauma, such as previous surgery (endonasal, orbital, skull base) or frontonasal injury. The time interval between trauma and diagnosis is long, often several years. Occasionally the predisposing factor may be inflammation or infection (chronic rhinosinusitis, rhinosinus polyposis, fungus ball), bone disease (osteoma, Paget's disease), or radiotherapy, but one-third of cases have no predisposing factors (Raynal et al., 1999; Lund, 1987; Moriyama et al., 1992; Brunori et al., 1995).

Our patient's MRI also showed a hyperintense lesion in T2-weighted sequences in the roof of an ethmoidal sinus with bone thinning, potentially compatible with mucocele. The drive for the encephalocele hypothesis was given in this case by the clinic and imaging suggestive of CSF leak and SIH, since the traumatic events on the road may have been the cause of both entities.

Encephalocele treatment is surgical. The aim is to mend the bone defect with a watertight dural seal and resect non-functional brain tissue. There are two main approaches for anterior basal encephaloceles: intracranial and extracranial (endoscopic) surgery. Whether to choose between these two surgical strategies depends on the dimensions and site of the leakage; relapses; the patient's general status; surgeon's preferences (Galli et al., 2021). Open intracranial surgery has traditionally been employed for repairing CSF leaks. However, lately, this technique has been replaced largely by endoscopic endonasal procedures due to their high efficiency, lower rates of morbidity and complications, lower invasiveness, and shortened hospital stay (Dai et al., 2020). Prognosis is determined by many variables such as position, size, amount of brain inside the sac, presence of dural sinuses in the sac, and occurrence of hydrocephalus. There is a better prognosis for patients with fronto-ethmoidal encephalocele rather than those with occipital or parietal encephalocele. Frequent complications are CSF leakage and meningitis, followed by hydrocephalus, recurrence, and seizures (Cruz and Jesus, 2021). In our case, given the diagnostic doubt, we opted first for an exploratory transethmoidal endoscopic approach. Given the small size of the lesion and the suspected osteodural defect on imaging, this strategy was also more appropriate and less invasive for removing the meningoencephalocele and dural plastic surgery.

4. Conclusions

The diagnostic workup of lesions of the paranasal sinuses in contact with the anterior skull base demands an accurate radiological and clinical study to search for elements of differential diagnosis between mucocele and meningoencephalocele. The suspicion of rhinoliquorrhoea, especially when associated with clinical and radiological signs of SIH, strongly suggests the latter. In these cases, a transnasal endoscopic surgical approach would be recommended to excise the lesion for a definitive histological diagnosis and, in cases of CSF leak, perform dural plastic surgery.

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Conflicts of interest

The authors have no conflicts of interest to declare that are relevant to the content of this article.

Ethical compliance statement

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

Consent to participate

Informed consent was obtained from all individual participants included in the study.

Consent to publish

Patient signed informed consent regarding publishing her data and photographs.

Data availability statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Authors' contribution

G.B., M.S., F.R. and P.F. performed the clinical assessment. M.S. performed the radiological assessment. G. Bo., G. Bu., A.G., G.I. and R.B. critically reviewed the literature and drafted the manuscript. All authors were responsible for important intellectual content. All authors read and approved the final version of the manuscript.

Structured review

A literature review was conducted on PubMed and Cochrane database using the terms "sphenoethmoidal encephalocele", "sphenoethmoidal meningoencephalocele", "spheno-ethmoidal encephalocele", "spheno-ethmoidal meningoencephalocele", "traumatic basal meningoencephalocele", "traumatic basal encephalocele", "sphenoethmoidal rhinoliquorrhoea" without limitations. Of 70 articles found, no article mentioned acquired sphenoethmoidal meningoencephalocele. Basing on this literature review, to the best of our knowledge, we report the first case of acquired sphenoethmoidal meningoencephalocele causing inconstant and subclinical rhinoliquoral fistula responsible for SIH.

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