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Abstract:	Abstract Anterior skull base defects with encephalocele in adults are quite rare and can be a cause of spontaneous rhinoliquorrea; however, cerebrospinal fluid (CSF) fistula can be not rarely misdiagnosed for several months or years. Five adult patients affected by ethmoidal encephalocele with CSF fistula has been treated in our Institute from 2006 through to 2011. Onset of clinical history was represented by rhinoliquorrea which was precociously recognized in only one patient; in the other four, it was misdiagnosed for a period ranging from 11 months to 5 years. After clinical diagnosis of CSF fistula and after brain MRI, ethmoidal encephalocele was evident in all patients; preoperative study was completed by spiral CT scan, to clearly identify the skull base bone defect. All patients were operated on by transsphenoidal endonasal endoscope-assisted microsurgical approach through one nostril. The herniated brain was coagulated and removed, and reconstruction of cranial base was performed. Postoperative rhinoliquorrea or other complications didn't occur in any patient at short and late follow-up. All patients were discharged after few days. Endonasal endoscope-assisted microsurgical approach resulted effective to expose and repair the ethmoidal bone defect; tridimensional vision and wide lateral and superior exposition of the operative field were possible in each patient thanks to the use of microscope and angulated endoscope.

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*Cover Letter

Rome, 8th August, 2013

Dear Editor, we would like to submit to your attention the enclosed manuscript regarding the mininvasive transnasal treatment of the ethmoidal encephalocele associated with CSF fistula

Kind regards,

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Ethmoidal encephalocele associated with cerebrospinal fluid fistula. Indications and results of mininvasive transnasal approach.

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Abstract

Anterior skull base defects with encephalocele in adults are quite rare and can be a cause of spontaneous rhinoliquorrea; however, cerebrospinal fluid (CSF) fistula can be not rarely misdiagnosed for several months or years.

Five adult patients affected by ethmoidal encephalocele with CSF fistula has been treated in our Institute from 2006 through to 2011. Onset of clinical history was represented by rhinoliquorrea which was precociously recognized in only one patient; in the other four, it was misdiagnosed for a period ranging from 11 months to 5 years. After clinical diagnosis of CSF fistula and after brain MRI, ethmoidal encephalocele was evident in all patients; preoperative study was completed by spiral CT scan, to clearly identify the skull base bone defect. All patients were operated on by transsphenoidal endonasal endoscope-assisted microsurgical approach through one nostril. The herniated brain was coagulated and removed, and reconstruction of cranial base was performed.

Postoperative rhinoliquorrea or other complications didn't occur in any patient at short and late follow-up. All patients were discharged after few days.

Endonasal endoscope-assisted microsurgical approach resulted effective to expose and repair the ethmoidal bone defect; tridimensional vision and wide lateral and superior exposition of the operative field were possible in each patient thanks to the use of microscope and angulated endoscope.

Key-words: ethmoidal encephalocele, rhinorrhea, microsurgery, endoscopy

Introduction

Anterior skull base defects with encephalocele in adults are quite rare (1) and can be a cause of spontaneous rhinoliquorrhea; however, cerebrospinal fluid (CSF) fistula can be misdiagnosed for several months or years (2,3), because liquor leak can be irregular and intermittent (4). We present five adult patients affected by ethmoidal encephalocele with CSF fistula, treated by transnasal microsurgical endoscope assisted approach. Indications, surgical technique and results of this mininvasive approach are discussed.

Materials and Methods

Patients and clinical history

Three female and two male patients were treated by transnasal approach from 2006 through to 2011 by the same operator (MF Fraioli). Average age was 58 years (range 47 – 68 years). Onset of clinical history was represented in all patients by spontaneous rhinoliquorrea which was immediately recognized in only one patient; in the other four, it was not diagnosed for a period ranging from 11 months to 5 years.

One patient was admitted to emergency unit of our hospital for a progressive hemiparesis due to unilateral chronic subdural hematoma which was evacuated through a craniectomy; after clinical anamnesis, it was clear that an intermittent rhinoliquorrea was present since 3,5 years in absence of other related symptoms. In another patient, two recurrent meningitis had occurred during 3 years, and intermittent rhinoliquorrea was present since 4 years; liquor leak had presented temporary discontinuance after each episode of meningitis. Another patient was admitted to emergency unit of our hospital for persistent cephalalgia not responding to nonsteroidal antinflammatory drugs, and cranial CT scan showed a frontal pneumoencephalus without mass effect; clinical history revealed the presence of rhinoliquorrhea since 2,7 years. The last patient, presenting a clear rhinoliquorrea since 11 months on the basis of anamnestic

features, was addressed to our attention after an encephalic MRI performed for cephalalgia, which revealed an ethmoidal encephalocele.

Preoperative diagnostic studies

After clinical diagnosis of CSF fistula, brain MRI revealed in all patients a clear ethmoidal encephalocele; preoperative study was completed by a spiral CT scan with coronal and sagittal reconstructions, with clear identification of the skull base bone defect. In three patients, the bone defect was located in the midline of the ethmoidal plane and was not larger than 2,1 cm of maximum diameter; in the two other patients, the bone defect presented extension on the left side and it was in both cases not larger than 1,3 cm of maximum diameter. In all patients, presence of brain tissue herniated out from the ethmoidal bone defect was clear at the MRI and CT scan (fig 1, fig. 2).

Surgical technique

All patients were operated on through one nostril with submucosal microsurgical endoscope assisted transnasal approach; sphenoidal sinus was not opened because the skull defect was located in all cases in the ethmoidal plane. At the beginning, the approach was conducted by microscope and exposition of ethmoidal plane was achieved; once the skull defect with herniation of brain and meningeal tissue was detected, with evidence of liquor leak, endoscope was introduced to better visualize the lateral and superior limits of the bone defect. The herniated brain was coagulated and removed, and reconstruction of the cranial base was performed: at the beginning, a single layer of absorbable hemostat was placed, then a layer of synthetic absorbable dura mater substitute was placed and covered by synthetic glue (2 cc); then, another layer of synthetic absorbable dura mater covering all the operative field was placed. Finally, a thin layer of wax for bone was applied. After the closure of the bone defect, a Valsalva maneuver was performed by the anaesthesist: in four patients no liquor leak was observed, while in the last one a very little leak was evident, so that another layer of synthetic

glue was placed. Postoperative lumbar drainage was placed only in this last patient and maintained for 4 days, draining 80-100 ml per day. Nasal packing was removed in this last patient in the fifth postoperative day, while in the others it was removed in the first postoperative day. All the absorbable materials (dura mater substitute, absorbable hemostat, synthetic glue) applied to close the bone defect, presented a time of absorption ranging from 2 to 4 months.

Results

No surgical nor perioperative complications were registered. The patient with postoperative lumbar drainage was put out of bed in the fifth postoperative day, and he was discharged after ten days from surgery. The other four patients were put out of bed in second postoperative day and they were discharged after four and five days from surgery in three and one case respectively. No patient presented liquor leak in the immediate postoperative period or at late follow-up, during an average period of 3,8 years. One patient reported temporary cacosmia which spontaneously regressed after 2 months.

Discussion

Diagnosis

First of all, a precocious diagnosis of CSF fistula is mandatory to avoid the possible related complications, from cephalalgia to subdural hematoma, from pneumoencephalus to meningitis. Cerebrospinal fluid fistula cannot be diagnosed for long periods of time (2,3), as occurred in our patients, because in some cases CSF leak can be intermittent (1,4). This intermittence can be explained by the fact that recurrent inflammation/infections can occur when a CSF fistula is present: the usual fibrosis following this infective/inflammatory events can temporary produce a fibrotic plane which occludes the bone defect, but during several weeks or months this post-inflammatory fibrotic layer is reabsorbed and CSF fistula starts

again. In other cases, CSF leak can be misdiagnosed with an allergic status or a common cold with rhinorrhea.

Endoscope / Microscope

In our opinion and experience, for this approach it is very important the use of microscope and endoscope during surgery, even if very good results have been reported using microscope (5) or endoscope (6) alone. The use of both devices, as other Authors reported for similar pathologies (7), allows to perform a better surgical approach with 3D visualization thanks to the microscope and wide lateral and superior vision thanks to the angulated endoscope. So that in our opinion, microscope and endoscope should be considered complementary and not rival devices, as reported by several Authors (8,9,10) also for other transsphenoidal pathologies.

Indications and objective of endonasal approach

Transnasal approach can be indicated when the skull base defect is not wide (4) (within 2,5 cm of maximum diameter in our experience), and when it is located in the midline or however lateral extension is limited. When the bone defect presents important lateral extension involving also the temporal bone (temporo-sphenoidal encephalocele), or large defect is present, transcranial approach is more effective than transnasal (11,12,13) one. Moreover, it is important to kept in mind the objective of surgery: operation should be conservative as much as possible, avoiding iatrogenic increase of the bone opening during surgery, but the approach should be limited to remove the herniated brain and to perform an effective reconstruction of the skull base defect.

Skull base reconstruction

During the development of microsurgical and endoscopic transnasal approaches in the past years and decades, many techniques for repairing the iatrogenic and congenital skull base defects have been reported, using several materials, from fascia lata to autologous fat graft,

from muscle to fibrin glue, to vascularized pedicled nasoseptal flap (14,15,16). In our experience, the materials used to close the skull bone defect were represented by absorbable synthetic tissues and wax for bone. This choice is due to the fact that in our past experience in transsphenoidal surgery, the employment of non absorbable materials, such as non absorbable dura mater substitute (neuropatch), could be the cause of postoperative cacosmia in some patients. This disturbance regressed only after surgical removal of the non absorbable materials and however, after this maneuver, we never observed postoperative CSF leak, so that we deduced that the implanted materials play a role in avoiding postoperative CSF leak only during the first weeks before the natural fibrosis is produced. This assumption can be confirmed by the presented study: after application of absorbable materials to close the skull base defect, no patient presented late postoperative rhinoliquorrea. Moreover, the choice of using synthetic materials (synthetic dura mater substitute, synthetic sealing) than biologic ones, was made to mostly reduce the rate of postoperative infections which can more frequently occur with application of biologic materials, graft in particular (17).

In conclusion, anterior skull base defects with encephalocele in adults can be a cause of spontaneous rhinoliquorrea. Clinical and radiologic features are mandatory for diagnosis and for the choice of the most appropriate surgical approach. Endonasal endoscope-assisted microsurgical approach resulted very effective in our experience to repair the ethmoidal bone defect, in absence of complications at short and late follow-up. Transnasal approach can be employed, in our opinion, when the bone defect is located in the midline or when lateral extension is very limited, and however in the cases in which the width of the bone defect is quite small (no larger than 2,5 - 3 cm of maximum diameter in our experience). Microscope and endoscope resulted complementary instruments; finally, the prevalent use of absorbable and synthetic materials assured excellent results in absence of complications.

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Figures legend

Fig. 1

Patient 2. T2 weighted MRI in sagittal (A) and coronal (B) planes showing the presence of a left ethmoidal encephalocele.

a: long white arrow indicates a normal right ethmoidal plane; short white arrow shows brain tissue herniated out from the left ethmoidal plane.

b: white circle shows the brain tissue herniated out from the ethmoidal plane; short white arrow shows the occasional presence of an empty sella not involved in the pathogenesis of the CSF fistula.

Fig. 2

Patient 5. Millimetric thickness CT scan in coronal (A) and sagittal (B) planes showing a left ethmoidal small encephalocele.

a: long white arrow shows a normal right ethmoidal plane; short white arrow indicates a small left encephalocele.

b: short white arrow indicates the encephalocele; short black arrow indicates a normal sella turcica.

Figure
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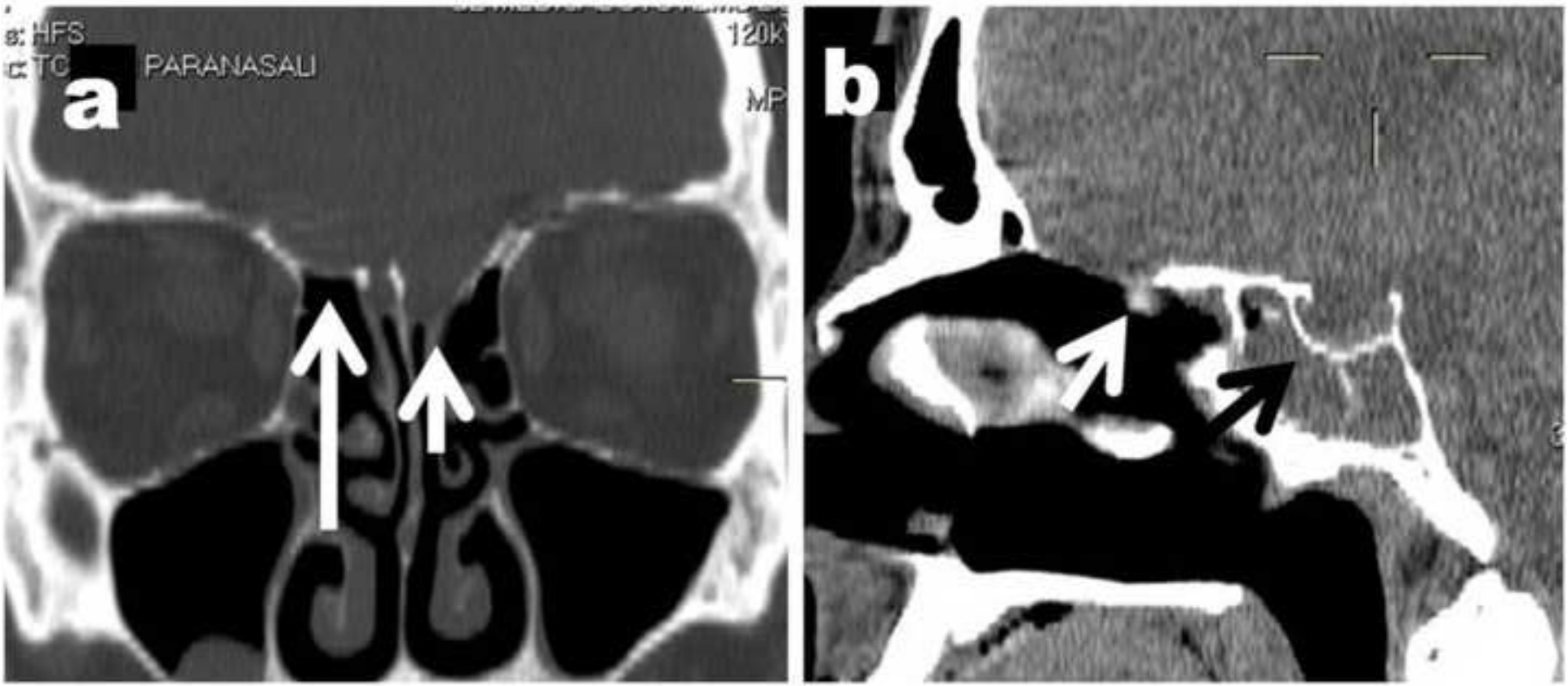

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