Spontaneous Unilateral Intrasphenoidal Meningocele

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ABSTRACT

The sphenoid sinus is an uncommon location for protrusion of a meningocele. When this does occur, it nearly always presents with leakage of cerebrospinal fluid through the nasal cavity. We present a case of a 38-year-old female found to have a meningocele protruding into the left sphenoid sinus, who presented with intractable headache but no CSF rhinorrhea. The lesion was discovered on computed tomography angiography, which was performed in order to rule out intracranial pathology as the etiology of her headache. Prior imaging, including pre- and post-contrast MRI, demonstrated the fluid within the sphenoid sinus, but did not reveal the communication through a defect in the base of the skull. Thus, it was assumed to be strictly related to sinus disease in the past. Our case represents a phenomenon whereby meningoceles protruding through the basilar skull into the sphenoid sinus or any other location are potentially misdiagnosed due to poor visualization of the osseous defect and lack of awareness of this entity.

CASE REPORT

CASE REPORT

Our patient is a 38-year-old female with a history of fibromyalgia and intermittent left hand tremor who was referred to neurology for left hand tremor, numbness and weakness. Patient underwent electromyography (EMG) which showed mild sensory peripheral neuropathy without radiculopathy or myelopathy. A pre- and post-contrast MRI of the brain was ordered for left-sided paresthesias and concern for demyelinating disease. Findings were described as "moderate fluid in the sphenoid sinus and accessory wing of the sphenoid" and diagnosed as sinus disease (Figure 1). No other findings, such as evidence of demyelinating disease, were identified.

After initial presentation, the patient also reported severe headache with blurred vision, for which she had two emergency room visits reporting "migraine-type" symptoms including severe headache, photophobia, phonophobia, nausea, and vomiting for three days. The patient's severe headache was relieved with a combination of Benadryl, ketorolac, and prochlorperazine at both encounters. She denied history of trauma or sinus surgery. Most recently, the patient had presented to an outpatient neurology clinic complaining of severe right-sided headache. A computed tomography angiography (CTA) of the head was ordered to exclude aneurysm, arterio-venous malformation (AVM), and vasculitis.

The CTA of the brain demonstrated an osseous dehiscence of the roof of the left lateral sphenoid sinus recess with protrusion of CSF into the sphenoid sinus (Figures 7 and 8). The clinoid segment of the left internal carotid artery abutted the sphenoid sinus without clear protrusion into the sphenoid sinus (Figure 9). Protrusion of the left temporal lobe parenchyma was questioned on the CTA (Figure 10), although

the possible encephalocele is best visualized retrospectively on the MRI (Figure 4).

In a retrospective analysis of our patient's initial imaging, CSF intensity is visualized on T1 imaging (Figures 1 and 2) within the sphenoid sinus, and the CSF is extending from the middle cranial fossa (Figures 1 and 3). Axial T2 and Sagittal T1 imaging also demonstrate questionable extension of the left temporal lobe parenchyma into the sphenoid sinus (Figures 1, 4, and 5).

Diagnosis of left intrasphenoidal meningocele with questionable encephalocele was made and findings were discussed with the ordering neurologist, who referred the patient to neurosurgery for evaluation.

DISCUSSION

Etiology & Demographics:

Meningocele refers to herniation of the meninges with accumulation of CSF through a bony defect in the skull or spinal canal. If any brain parenchyma is included in the herniation, it is referred to as a meningoencephalocele. Intracranial meningoceles may be classified as basal, frontal, occipital, or parietal, based on the location of the osseous defect through which herniation occurs. Basal meningoceles involving the sphenoid sinus have traditionally been labeled transsphenoidal, with subclassification as either intrasphenoidal i.e. contained completely within the sphenoid sinus, or true transsphenoidal i.e. protruding through the sphenoid sinus into the nasal cavity. [1]

Current understanding of basal meningocele suggests multiple etiologies including traumatic, iatrogenic, congenital, and spontaneous. [2] Regarding a congenital etiology, it has been theorized that basal bony defects are the result of failure of the sphenoid ossification centers to fully ossify and fuse together, forming the Sternberg Canal, also called the Lateral Craniopharyngeal Canal. [4, 5] Regarding spontaneous etiology, growing literature supports the notion that increased intracranial pressure (ICP) may be a common contributing factor. [1]

Epidemiologic data is limited as this is an uncommon finding, however a study published by Zoli et al. may provide some useful information. [6] Among 23 patients diagnosed with either meningocele or meningoencephalocele through an osseous defect of the lateral wall of the sphenoid sinus (LWSS), there was a female/male incidence of 16/7, a left/right-sided incidence of 12/11, and an average age of 51 years old. Among the 12 of those patients with meningocele specifically, there was a female/male incidence of 9/3, a left/right-sided incidence of 6/6, and an average age of 53 years old. Although their patient population was rather small, the study demonstrates a clear predominance of females over males, and no preference for sidedness. Additional findings of interest which were reported in many of their LWSS defect patients, including both meningocele (MC) and meningoencephalocele (ME) included the following: the Sylvian fissure was wider on the ipsilateral side of the defect in 15 cases (65%, 5 MC, 10 ME), and the ipsilateral temporal horn of the lateral ventricle was enlarged in 5 cases (21%, 1 MC, 4 ME). In a review of the literature Radonjic et al. found that, out of 28 reported cases of spontaneous intrasphenoidal lesions, 17 involved the lateral recess. [3]

Clinical & Imaging Findings:

Our case presents a patient with spontaneous meningocele through a bony defect in the high lateral wall of the sphenoid sinus on the left side, discovered on CTA in a patient without history of trauma or sinus surgery. Our patient reported severe frontal headache but no evidence of either current or remote CSF rhinorrhea, which deviates from the usual presentation. [3, 4] Other common signs and symptoms, which are also absent in our patient, include recurrent meningitis, vertigo, seizures, and focal neurologic deficits. As increased ICP is a suspected cause, our patient's scans were retrospectively evaluated for indirect signs of increased ICP such as empty sella syndrome, as suggested by Radonjic et al., posterior globe flattening, sulcal effacement, ventricular effacement, and loss of gray-white differentiation, but none were apparent. [3] A significant challenge in making an accurate diagnosis of intrasphenoidal meningocele, as was encountered in our case, is to identify the osseous defect in the skull base, which is often very small and may not be apparent on thin-slice CT or contrast-enhanced MRI. If not identified, a meningocele can easily be misdiagnosed as a lesion confined to the sphenoid sinus. If suspicion for the presence of meningocele is high and no osseous deformity is evident, CT-cisternography is advised to evaluate for communication of intrathecal contrast. [8]

Treatment & Prognosis:

Surgical correction of the LWSS defect in patients with meningocele or meningoencephalocele is recommended even in cases presenting without CSF rhinorrhea, for the prevention of both meningitis and further progression of the herniation. [6] The Endonasal Endoscopic approach to correction of basal skull defects is the preferred surgical method over the Transcranial approach, especially in cases of LWSS defect as it can be difficult to reach via the latter approach. [4, 5, 9] Zoli et al. performed this operation on 23 patients, with antibiotics administered to those with signs of meningitis until resolution of CSF and serologic parameters. [6] At follow-up all 23 patients were in stable clinical condition with resolution of their symptoms, and no apparent neurological sequelae.

Differential Diagnoses:

The differential diagnosis for a fluid-density lesion located unilaterally within the sphenoid sinus primarily includes lesions arising from the sphenoid sinus itself, most commonly mucocele, mycetoma, and sinusitis. Mass effect by these lesions can cause remodeling and dehiscence of surrounding osseous structures and could present as headache, facial pain, nausea, and cranial nerve deficits. However, if the lesion is identified as a herniation of the meninges, consideration must also be given to the possibility of herniated brain parenchyma, which would distinguish it as a

meningoencephalocele rather than simply a meningocele. CT is very useful for the identification of a bony defect, and MRI is very useful for characterization of the lesion itself. Meningocele will follow all MRI signal characteristics of CSF: T1 low signal and T2 high signal. Encephalocele will appear as an extension of the brain parenchyma through an osseous defect into the sphenoid sinus. Meningocele will demonstrate no enhancement, whereas encephalocele will enhance the same as normal brain parenchyma.

Mucocele

A mucocele is a benign expansile lesion, which occurs due to the accumulation of normal physiologic secretions by the sinus mucosa. One may occur as the result of obstruction by a polyp or tumor, inflammation by infection or allergy, or injury by trauma or surgery. Among the paranasal sinuses, the sphenoid sinus is the least common location for a mucocele (1-7%), with the frontal sinus being the most common (65%), followed by the maxillary sinuses. [10, 11] As the simple fluid of a mucocele is of such similar density to that of CSF, mucocele is likely the most common misdiagnosis in the case of a sphenoid sinus meningocele. CT typically demonstrates a homogeneous lesion of fluid density with minimal peripheral enhancement and with no central enhancement. Bone window often shows expansion of the sinus walls, though without osseous destruction. MRI may demonstrate variable T1 signal based on protein content, and T2 signal which is high if there is significant water content, or low if there is inspissated mucous. T1 C+ shows minimal peripheral enhancement and no central enhancement.

Mycetoma

A mycetoma is an infectious lesion caused by infiltration of the skin or mucosa resulting in mass-like inflammation of the tissues. Etiologic organisms may be bacterial (actinomycetoma) or fungal (eumycetoma), and have a predilection for individuals with immunocompromise. [12] Though typically affecting the soft tissues of the extremities, a mycetoma may also be found in the paranasal sinuses, most commonly the maxillary sinus, followed by the sphenoid sinus. General radiologic features suggestive of mycetoma include heterogeneous soft tissue density, scattered calcific deposits, and inflammatory changes in the affected mucosa. The infectious and inflammatory nature of this lesion also increases the likelihood that the sinus outlet will be obstructed, leading to complete opacification of the affected sinus. CT may demonstrate thickened mucosa at the periphery of the sinus, with or without enhancement. Bone window shows a focal mass in the sinus lumen with central high density and occasional calcification, usually without expansion of the bony walls. MRI may demonstrate T1 hypointense signal within a solid mass, and T2 hypointense signal due to large amounts of protein and calcium. T1 C+ may show inflamed peripheral mucosal enhancement.

Sphenoethmoidal Sinusitis

Sinusitis refers to inflammation of the mucosa lining the paranasal sinuses, and may be caused by viral, bacterial, or fungal infection. Similarly to mycetoma, sinusitis may easily cause obstruction resulting in complete opacification of the affected sinus. Sinusitis is a common finding; however, it is extremely rare for sphenoid sinusitis to be found in isolation without concurrent infection of the other paranasal sinuses. [13] Mucoid secretions may also form mass-like accumulations of fluid density. CT may demonstrate air-fluid levels with "bubbly" secretions, and moderate mucosal thickening, usually >1cm. The mucosa may enhance, but any accumulated secretions should not. Bone window typically shows no destruction of the bony walls. MRI may demonstrate air-fluid levels on T1 and thickened mucosa on T2, isointense to soft tissues. Secretions may have low T2 signal due to proteinaceous content. The mucosal lining may enhance on T1 C+.

TEACHING POINT

A meningocele protruding through the basilar skull into the sphenoid sinus may be misdiagnosed due to poor visualization of the osseous defect and lack of awareness of this entity. If suspicion for the presence of meningocele is high and no osseous deformity is evident, CT-cisternography is advised to evaluate for communication of intrathecal contrast.

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FIGURES

Figure 1: 38-year-old female with intrasphenoidal meningocele.
FINDINGS: T2-weighted axial MRI shows high T2 signal in the left lateral recess of the sphenoid sinus with extension from the middle cranial fossa.
TECHNIQUE: MRI without contrast, performed on a 1.2T Open MRI.

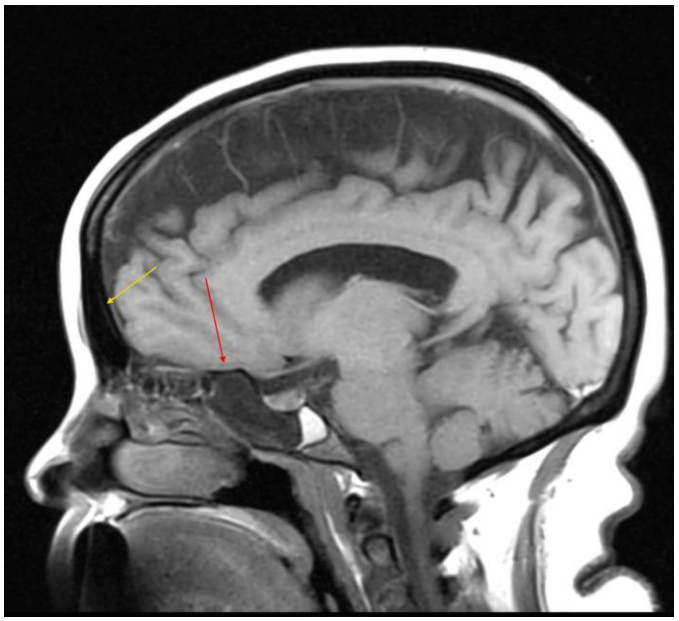


Figure 2: 38-year-old female with intrasphenoidal meningocele.
FINDINGS: Sagittal T1-weighted MRI shows CSF signal intensity in the sphenoid sinus (red arrow) when compared to the intensity of air in the frontal sinus (yellow arrow).
TECHNIQUE: MRI without contrast, performed on a 1.2T Open MRI.

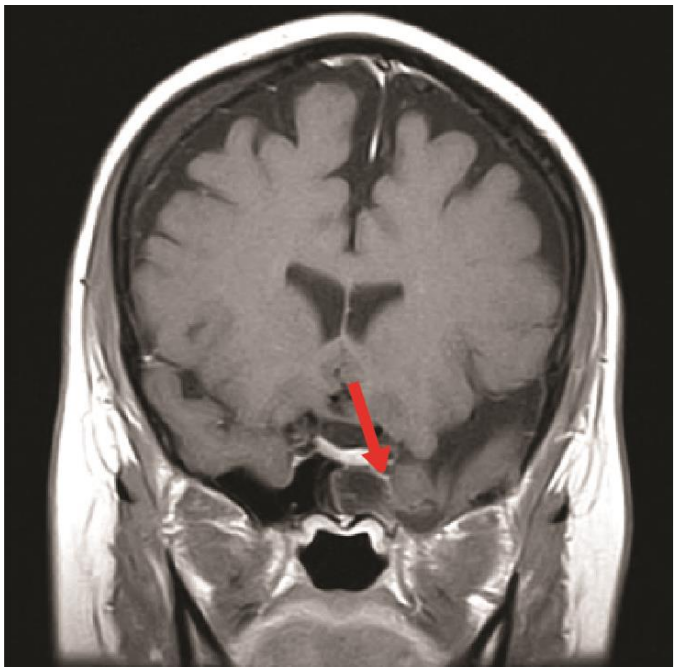


Figure 3: 38-year-old female with intrasphenoidal meningocele.
FINDINGS: Coronal T1 Post-contrast MRI imaging shows extension of CSF into the sphenoid sinus.
TECHNIQUE: MRI with contrast, performed on a 1.2T Open MRI, (20mL of Multihance intravenous contrast).

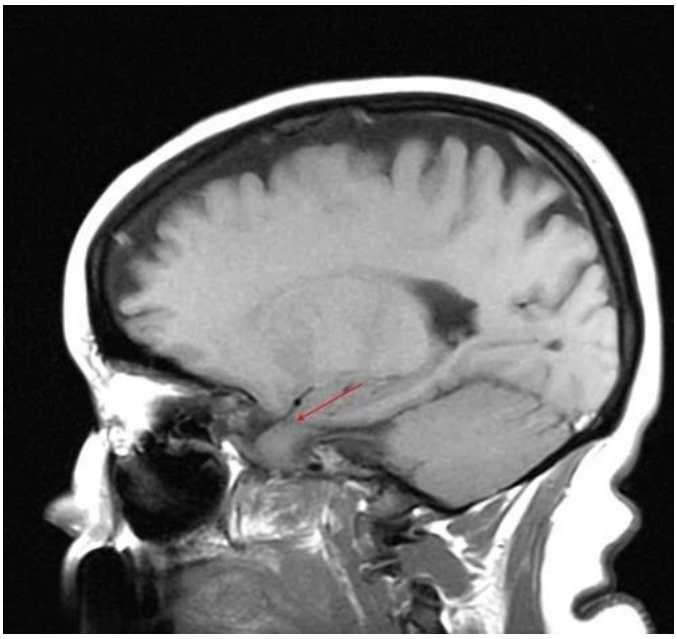


Figure 4: 38-year-old female with intrasphenoidal meningocele.
FINDINGS: Sagittal T1-weighted MRI shows extension of the meninges into the sphenoid sinus.
TECHNIQUE: MRI without contrast, performed on a 1.2T Open MRI.

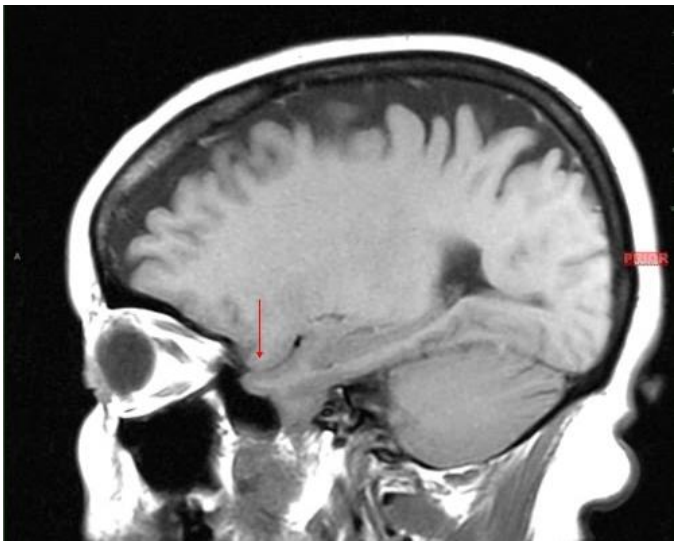


Figure 5: 38-year-old female with intrasphenoidal meningocele.
FINDINGS: Sagittal T1-weighted MRI of the normal right temporal lobe of the same patient for comparison.
TECHNIQUE: MRI without contrast, performed on a 1.2T Open MRI.

Figure 8 (right): 38-year-old female with intrasphenoidal meningocele.
FINDINGS: Coronal Computed Tomography Angiogram of the Brain in Soft Tissue Window shows protrusion of CSF through the osseous dehiscence.
TECHNIQUE: CT Angiogram of the Brain, kVP 120, 5.0mm thick slices, 75mL of Isovue 370 intravenous contrast.

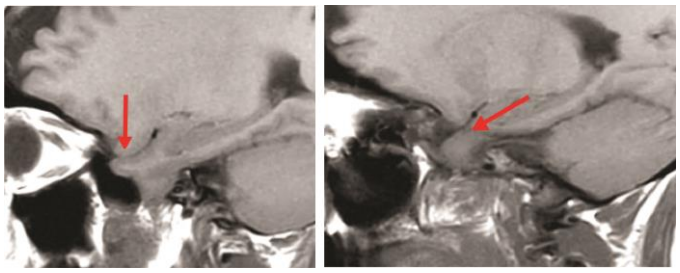


Figure 6: 38-year-old female with intrasphenoidal meningocele.
FINDINGS: Magnification and side-by-side for comparison of Figures 4 and 5.

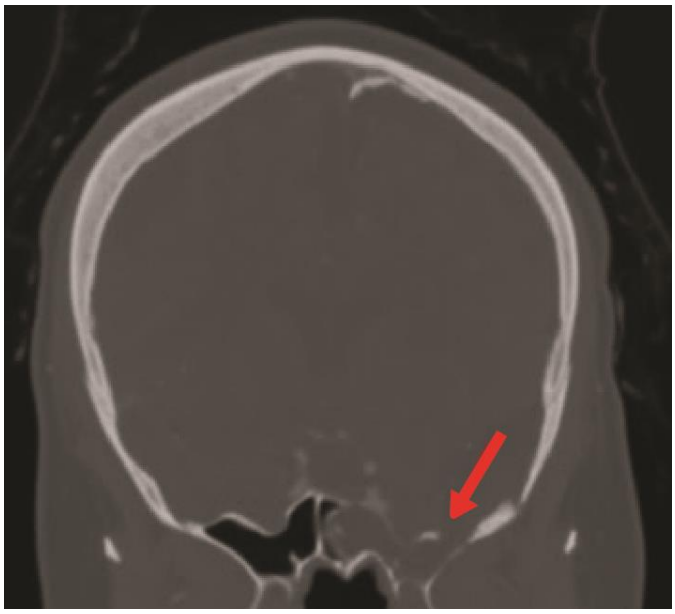
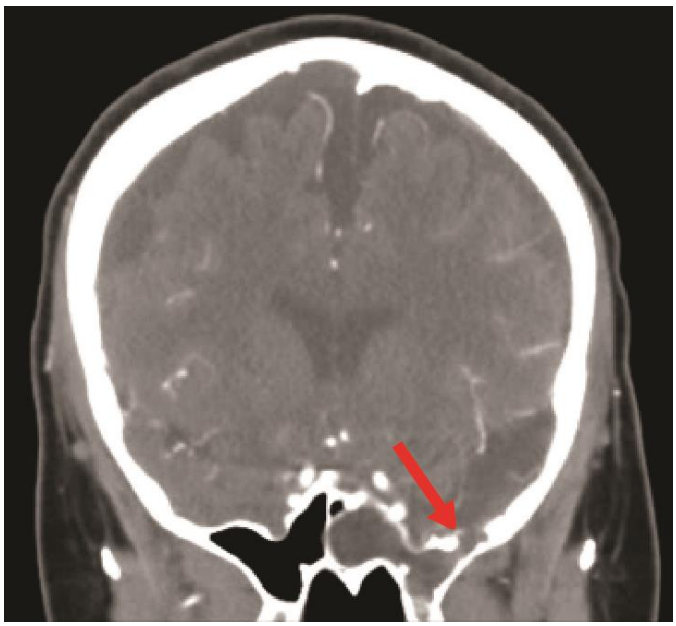


Figure 7: 38-year-old female with intrasphenoidal meningocele.
FINDINGS: Coronal Computed Tomography Angiogram of the Brain in Bone Window shows osseous dehiscence of the roof of the left lateral sphenoid sinus.
TECHNIQUE: CT Angiogram of the Brain, kVP 120, 5.0mm thick slices, 75mL of Isovue 370 intravenous contrast.



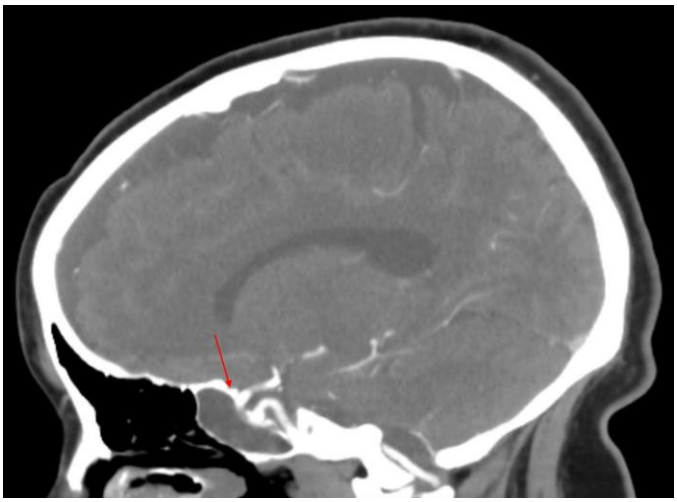


Figure 9: 38-year-old female with intrasphenoidal meningocele.
FINDINGS: Sagittal Computed Tomography Angiogram of the Brain in Soft Tissue Window shows the clinoid segment of the left internal carotid artery abutting the sphenoid sinus without protrusion.
TECHNIQUE: CT Angiogram of the Brain, kVP 120, 5.0mm thick slices, 75mL of Isovue 370 intravenous contrast.

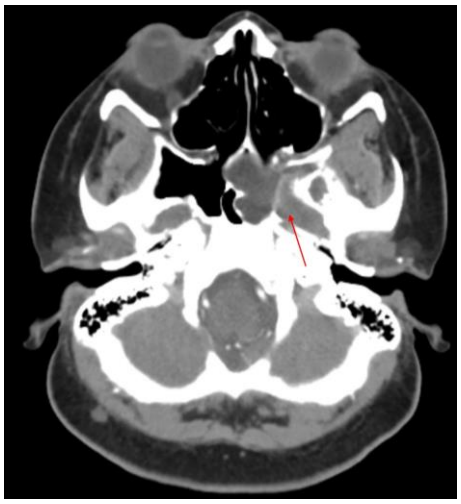


Figure 10: 38-year-old female with intrasphenoidal meningocele.
FINDINGS: Axial Computed Tomography Angiogram of the Brain in Soft Tissue Window shows slightly hyperdense material protruding into the sphenoid sinus compared to CSF, possibly representing left temporal lobe parenchyma.
TECHNIQUE: CT Angiogram of the Brain, kVP 120, 5.0mm thick slices, 75mL of Isovue 370 intravenous contrast.

Definition	Herniation of the meninges with accumulation of CSF, through a bony defect in the skull base and protruding into the sphenoid sinus
Etiology	The herniation may be present congenitally or may occur later in life due to the presence of a skull base deformity, which was present congenitally or which has arisen due to traumatic or iatrogenic injury
Incidence	Unknown
Gender Ratio	Clear female predominance, with the female:male ratios as follows -16:7 if considering both meningocele and meningoencephalocele -9:3 if considering meningocele only
Age Predilection	-Average age 51 if considering both meningocele and meningoencephalocele -Average age 53 if considering meningocele only
Risk Factors	Trauma and surgery near the sinuses represent clear risk factors. Regarding spontaneous etiology, increased intracranial pressure may represent a risk factor. Regarding congenital etiology, the presence of the Lateral Craniopharyngeal Canal may be a risk factor
Treatment	Surgical correction of the herniation with closure of the bony defect using the endonasal endoscopic approach is recommended for the prevention of both meningitis and further progression of the herniation
Prognosis	After performing surgical correction, Zoli et al. achieved symptomatic resolution without neurologic sequelae in all 23 of their patients
Findings on Imaging	-CT findings: CT is essential to evaluate for bony defect where the meninges/brain parenchyma protrude into the sphenoid sinus -MRI findings: Meningocele and encephalocele will protrude through the osseous defect into the sinus. Meningocele will follow all signal characteristics of CSF: T1 low signal and T2 high signal. Encephalocele will appear as extension of the brain parenchyma through the osseous defect into the sphenoid sinus. Meningocele will demonstrate no enhancement. Encephalocele will enhance the same as normal brain parenchyma -X-ray findings: Non-specific opacification of the sphenoid sinus

Table 1: Summary table for intrasphenoidal meningocele.

Differential Diagnosis	CT findings	MRI findings	X-ray findings
Intrasphenoidal Meningocele / Encephalocele	CT is essential to evaluate for bony defect where the meninges/brain parenchyma protrude into the sphenoid sinus	Meningocele and encephalocele will protrude through the osseous defect into the sinus. Meningocele will follow all signal characteristics of CSF: T1 low signal and T2 high signal. Encephalocele will appear as extension of the brain parenchyma through the osseous defect into the sphenoid sinus. Meningocele will demonstrate no enhancement. Encephalocele will enhance the same as normal brain parenchyma	Non-specific opacification of the sphenoid sinus
Sphenoid Sinus Mucocele	No central enhancement. Minimal peripheral enhancement may be present. Bone windows show expansion of the sinus walls. No osseous destruction. No air fluid levels	Variable T1 signal based on protein content. T2 signal may be high if there is high water content, or low if there is inspissated mucous. No central enhancement on T1 C+. Minimal peripheral enhancement may be present	Suggested from opacification of the sinus with loss of normal mucoperiosteal line of the sphenoid sinus wall
Mycetoma	Thickened mucosa at the periphery of the sinus may enhance. Bone CT shows a focal mass in the sinus lumen with central high density and occasional calcification. The sinus is not usually expanded	T1 hypointense signal in solid mycetoma mass. T2 hypointense signal due to protein and calcium in the mass. T1 C+ may show inflamed peripheral mucosal enhancement	Non-specific opacification of the sinus
Sphenoethmoidal Sinusitis	Air-fluid level with "bubbly" secretions. Moderate mucosal thickening, usually >1cm. Sinus mucosa may enhance, but secretions should not. Bone CT shows no destruction typically	T1 air-fluid level. Mucosal thickening isointense to soft tissue. T2 thickened mucosa. Secretions may have low T2 signal due to proteinaceous content. T1 C+ enhancing mucosal lining of the sinus cavity. Secretions do not enhance	Sinus opacification with air-fluid level. Should be supplemented with CT

Table 2: Differential diagnosis table for intrasphenoidal meningocele.

ABBREVIATIONS

AVM = Arteriovenous Malformation
CSF = Cerebrospinal Fluid
CT = Computed tomography
CTA = Computed tomography angiography
EMG = Electromyography
ICP = Intracranial Pressure
LWSS = Lateral Wall of Sphenoid Sinus
MC = Meningocele
ME = Meningoencephalocele
MRI = Magnetic resonance imaging

KEYWORDS

Intrasphenoidal; Meningocele; Meningoencephalocele; Sphenoid Sinus; Computed Tomography Angiography; Sternberg Canal; Endoscopic Endonasal Repair

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