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Case Report

Symptomatic cerebrospinal fluid leak secondary to sphenoidal Ecchordosis physaliphora: case report and literature review

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ABSTRACT

Ecchordosis physaliphora (EP) is a rare, benign, and hamartomatous tumor that is driven from ectopic notochordal remnants. It has a usual asymptomatic presentation and is often incidentally detected in about 2% of autopsies. A 50-year-old female presented to rhinology clinic with clear nasal discharge for 6 weeks associated with frontal headache. Unremarkable neurological and ENT examination. Brain computed tomography (CT) scan demonstrated a focal bony defect at the posterior wall of sphenoid sinus with partial opacification of the sphenoid sinus showing air fluid level suspicious for cerebrospinal fluid (CSF) leakage with a small cyst. Further assessment with MRI of the brain showed signs of rhinorrhoea secondary to ecchordosis physaliphora. Patient was managed successfully with endoscopic trans-nasal skull base reconstruction. Upon follow-up at two years post-operatively, no evidence of recurrence was detected. EP is indistinguishable from chordoma histopathologically hence imaging plays a crucial role in diagnosis. EP appears as hyperintense in T2-weighted images and hypointense in T1-weighted images. Symptomatic EP cases are extremely rare and most of these cases are managed by resection via craniotomy. In addition, endoscopic endonasal trans-sphenoidal surgery (ETSS) provides a good approach. Imaging is crucial for the diagnosis of these lesions. Surgical treatment is considered the gold standard for symptomatic ecchordosis physaliphora. Endoscopic endonasal transnasal skull base reconstruction is effective for management, it is a minimally invasive approach for complete resection of the lesions with reduced hospitalization time and less postoperative complications.

Keywords: Chordoma, Clivus, Trans-sphenoidal, Endoscopic, Case report

INTRODUCTION

Cerebrospinal fluid (CSF) leaks can present various symptoms, and their etiology may include traumatic or non-traumatic causes. Ecchordosis physaliphora (EP) is a congenital, benign, and hamartomatous lesion derived from notochordal remnants at the posterior clivus.

Unlike chordomas which are often symptomatic due to brainstem or cranial nerve compression, patients with EP are usually asymptomatic. They are found in ~2% of autopsies.[1] Herein, we report a case of a symptomatic CSF leak associated with EP in a 50-year-old female who presented to a tertiary hospital and treated successfully with endoscopic trans-nasal skull base reconstruction.

CASE REPORT

A 50-year-old female with a medical history of glucose-6-phosphate dehydrogenase (G6PD) deficiency presented to rhinology and skull base clinic with a history of intermittent clear nasal discharge primarily from the right nostril over the last six weeks associated with frontal headache. The nasal discharge occurred mainly while

bending the head forwards. The patient denied any visual or neurological complaints. There was no significant medical or past surgical history.

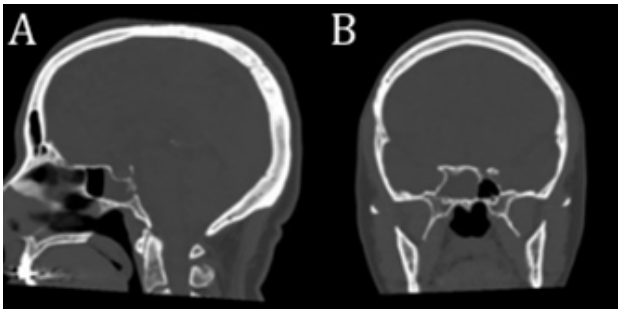

Figure 1: CT head (A) Sagittal view (B) Coronal view showing: Focal bony defect at the posterior wall of sphenoid sinus with partial opacification of the sinus, showing air-fluid level suspicious for CSF leakage with bony erosions and small cyst.

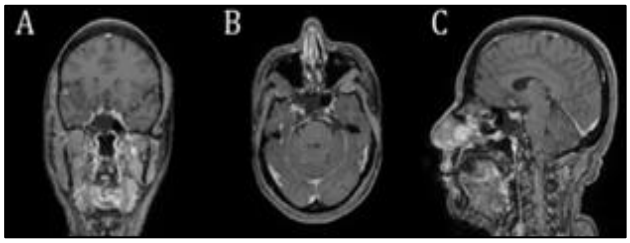

Figure 2: MRI of brain (A) Coronal view (B) Axial view (C) Sagittal view showing: Focal bony defect at the posterior wall of the right sphenoid sinus with erosion and small cyst-like structure at the cranial aspect of the wall and intra sinus calcification at the site of the defect.

Associated air-fluid level is noted at the related sinus with no restricted diffusion or abnormal post IV contrast enhancement. The appearance likely represents CSF rhinorrhoea secondary to EP.

ENT and neurological examination revealed no abnormalities. Imaging studies, including computed tomography (CT) and magnetic resonance imaging (MRI) were requested and showed a focal bony defect at the posterior wall of the right sphenoid sinus (Figure 1 & 2). The defect is associated with a cyst-like structure and intra-sinus calcification, that was consistent with rhinorrhoea and cerebrospinal fluid (CSF) leak secondary to EP.

The patient subsequently underwent endoscopic trans-nasal skull base reconstruction by a rhinologist and a neurosurgeon with a biopsy from the clival lesion (Figure 3).

An endoscopic endonasal transsphenoidal approach was employed, involving a bilateral sphenoidotomy. A cystic lesion was identified just behind the posterior wall of the sphenoid, and a biopsy was obtained from the lesion.

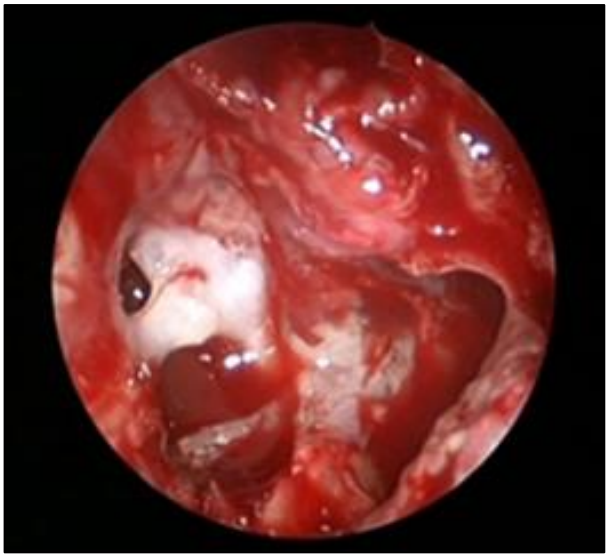

Figure 3: Endoscopic view of posterior wall of the sphenoid sinus after grafting and reconstruction.

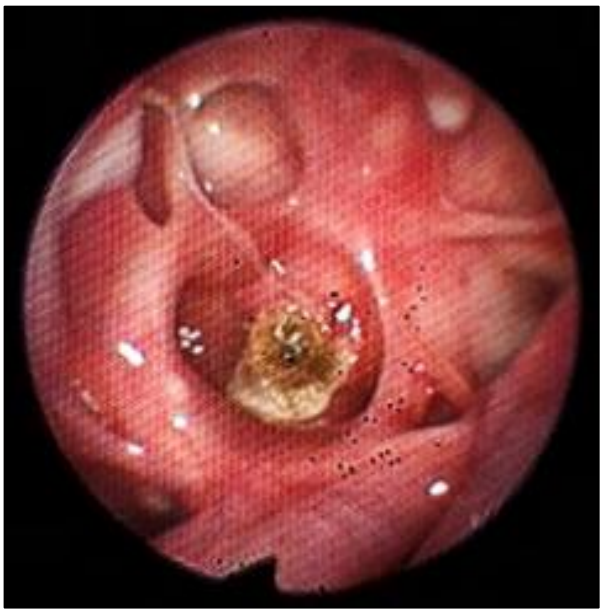

Figure 4: Endoscopic view of the surgical site 6 months post-operatively.

The defect was then closed using a multilayer technique that utilized septal bone followed by a naso-septal flap. Histopathology results from the lesion were inconclusive. However, based on imaging findings suggestive of EP, the decision was made for close follow-up with clinical and imaging monitoring.

Over a 2-year follow-up period with MRI and nasal endoscopic examination, there was no evidence of leak recurrence, the flap was intact in place, and the patient was free of symptoms (Figure 4).

Table 1: Summarized findings of case reports on CSF leak secondary to sphenoidal ecchordosis physaliphora.

Average age	**53.25**
Gender distribution	F 76% (19/25) M 24% (6/25)
CSF rhinorrhoea side	Right (5/25) Left (5/25) Unreported (15/25)
Associated symptoms	Headache (8/25)
Average duration of CSF leak	10.5 months
Meningitis	52% (13/25)
Location	Prepontine 36% (9/25) Clivus 64% (16/25)
Surgical approach	Microscopic transsphenoidal 8% (2/25) endoscopic endonasal 92% (23/25)
Average follow-up duration	19.7 months

Table 2: Case reports on CSF leak secondary to sphenoidal ecchordosis physaliphora.

Authors	Age	Gender	Side (clinical or imaging)	Symptoms	Duration of CSF leak rhinorrhoea	Meningitis	Investigations	Location	Management	Follow-up
Macdonald et al[8]	66	F	Right	Rhinorrhoea, Anosmia (since childhood)	24 months	No	Glucose levels and chloride, CT, MRI, histopathology	Prepontine	Microscopic Sublabial midline rhinoseptal mass resection and repair with autogenous fascia lata.	4 months
Alli et al[9]	52	F	Left	Clear rhinorrhoea	2 months	Yes	Glucose levels and positive β-2-transferrin, CT, MRI	Clivus	Microscopic Transsphenoidal repair with fascia lata graft, DuraSeal tissue sealant, fat graft, and spongistan.	24 months
Dias et al[10]	54	F	Right	Fever, headaches, meningism, clear rhinorrhoea	6 months	Yes	LP, CT, MRI, histopathology	Clivus	Endoscopic endonasal Transsphenoidal mass resection and repair with hydroxyapatite bone cement, nasoseptal flap, and a layer of polyethylene glycol hydrogel sealant.	24 months
Bolzoni-Villaret et al[11]	51	F	Right	Recurrent rhinorrhoea	36 months	No	CT, MRI	Clivus	Endoscopic endonasal Transsphenoidal partial mass resection and repair with nasoseptal flap, fat tissue or the iliotibial tract graft.	12 months

Continued.

Authors	Age	Gender	Side (clinical or imaging)	Symptoms	Duration of CSF leak rhinorrhoea	Meningitis	Investigations	Location	Management	Follow-up
Ferguson et al[12]	Mid-age	F	Left	Long standing recurrent rhinorrhoea, and new onset headache and confusion	Years	Yes	LP, CT, MRI, histopathology	Prepontine	Endoscopic endonasal Transsphenoidal mass resection and repair with fat graft.	-
Galloway et al[13]	40	F	Left	Rhinorrhoea and meningism	3 weeks	Yes	CT, MRI, histopathology	Clivus	Endoscopic endonasal Transsphenoidal mass resection and repair with nasoseptal flap and DuragenTM inlay.	36 months
Derakhshani et al[14]	68	F	-	Imbalance, worst headache, occasional unprovoked clear rhinorrhoea	5 months	No	CT, MRI, histopathology	Clivus	Endoscopic endonasal Transsphenoidal mass resection and repair with nasoseptal flap and AlloDerm graft.	2 months
Ghimire et al[15]	65	F	-	Excessive rhinorrhoea, and 3 days progressive left-sided weakness preceded by a week of general malaise, anorexia, and lethargy	1 month	No	CT, MRI, histopathology	Clivus	Endoscopic endonasal Transsphenoidal mass biopsy and repair with nasoseptal flap and fluorescein.	3 months
Georgalas et al[6]	81	M	-	Recurrent meningitis, watery rhinorrhoea	5 days	Yes	CT, MRI, histopathology	Prepontine	Endoscopic endonasal Transsphenoidal mass resection and repair with triple-layer repair (fat, fascia, nasoseptal flap).	28 months
Georgalas et al[6]	60	M	-	Watery rhinorrhoea	15-day	Yes	CT, MRI	Prepontine	Endoscopic endonasal Transsphenoidal mass resection and repair with triple-layer repair (fat, fascia, nasoseptal flap).	20 months
Georgalas et al[6]	64	F	-	Intermittent watery rhinorrhoea	Several years	Yes	CT, MRI	Prepontine	Endoscopic endonasal Transsphenoidal mass resection and repair with triple-layer repair (fat, fascia, nasoseptal flap).	7 months

Continued.

Authors	Age	Gender	Side (clinical or imaging)	Symptoms	Duration of CSF leak rhinorrhoea	Meningitis	Investigations	Location	Management	Follow-up
Veiceschi et al[16]	59	M	-	Rhinorrhoea		Yes	CT, MRI, histopathology	Prepontine	Endoscopic endonasal Transsphenoidal partial mass resection and repair with autologous layer of fascia lata, adipose tissue, vascularized nasoseptal fap	36 months
Veiceschi et al[16]	64	F	-	Spontaneous CSF leak		No	CT, MRI, histopathology	Prepontine	Endoscopic endonasal Transsphenoidal partial mass resection and repair with autologous layer of fascia lata, adipose tissue, vascularized nasoseptal fap	24 months
Veiceschi et al[16]	41	M	-	CSF rhinorrhoea was referred appearing after minor head trauma		No	CT, MRI, histopathology	Prepontine	Endoscopic endonasal Transsphenoidal partial mass resection and repair with autologous layer of fascia lata, adipose tissue, vascularized nasoseptal fap	72 months
Veiceschi et al[16]	57	M	-	CSF rhinorrhoea was referred appearing after minor head trauma		No	CT, MRI, histopathology	Prepontine	Endoscopic endonasal Transsphenoidal partial mass resection and repair with autologous layer of fascia lata, adipose tissue, vascularized nasoseptal fap	18 months
Sooltangos et al[17]	39	F	Left	excessive rhinorrhoea, in addition to headache	6 months	Yes (postoperatively)	CT, MRI, histopathology, beta-2 transferrin, fluorescin	Clivus	Endoscopic endonasal Transsphenoidal mass biopsy and repair with abdominal fat, dural substitute, and tissue fibrin glue.	8 months
Sooltangos et al[17]	43	F	Right	Excessive rhinorrhoea, symptoms of meningism.	1 week	Yes	CT, MRI, histopathology	Clivus	Endoscopic endonasal Transsphenoidal mass resection and repair with with abdominal fat graft, dural substitute, and fibrin glue	-
Sooltangos et al[17]	39	F		Excessive rhinorrhoea and long- standing headache, postnasal	4-5 months	No	CT, MRI, histopathology	Clivus	Endoscopic endonasal Transsphenoidal mass resection and repair with triple-layer repair (inlay	21 months

Continued.

Authors	Age	Gender	Side (clinical or imaging)	Symptoms	Duration of CSF leak rhinorrhoea	Meningitis	Investigations	Location	Management	Follow-up
				drip, altered sense of smell.					fascia lata, onlay dural substitute, nasoseptal flap, and fibrin glue).	
Sooltangos et al[17]	45	F		Vasovagal collapsed at home sustaining a head injury, 1 year history of unilateral rhinorrhoea.	1 year	No	CT, MRI, histopathology	Clivus	Endoscopic endonasal Transsphenoidal mass biopsy and repair.	-
Hasegawa et al[18]	39	F		Intermittent rhinorrhoea with progressive headache	1-month	Yes	CT, MRI	Clivus	Endoscopic endonasal Transsphenoidal repair with fat, fascia lata, and tissue fibrin glue.	10 years
Hasegawa et al[18]	62	M		Rhinorrhoea postoperative of transsphenoidal resection of pituitary macroadenoma	5 days	No	CT, MRI	Clivus	Endoscopic endonasal Transsphenoidal mass resection and repair with duragen, fat, nasoseptal flap	24 months
Hasegawa et al[18]	41	F		Rhinorrhoea longstanding history of headache, obesity	1 month	No	CT, MRI	Clivus	Endoscopic endonasal Transsphenoidal partial mass resection and repair with duragen, fat, nasoseptal flap	-
Ruiz et al[5]	46	F		Spontaneous rhinorrhoea		Yes	CT, MRI, Biopsy (histopathology)	Clival	Endoscopic endonasal Transsphenoidal mass resection and repair	-
Aljawi et al[19]	52	F	Left	clear rhinorrhoea, associated with on-and-off headaches	7 years	Yes	β-2-transferrin, CT, MRI	Clival	Endoscopic endonasal Transsphenoidal repair with bovine pericardium graft and fibrin sealant were placed, followed by a pedicled septal flap.	9 months
Present case	50	F	Right	Rhinorrhoea	6 weeks	No	CT, MRI	Clival	Endoscopic endonasal Transsphenoidal repair with septal bone, nasoseptal flap, gel foam and glue.	12 months

DISCUSSION

Ribbert defined EP in 1894 as a remnant of notochordal tissue that did not develop malignantly into a chordoma. EP is still categorized as a benign congenital disorder distinguished from chordoma by the absence of aggressive characteristics, despite the fact that they seem almost identical histologically. Although it is classified as a notochordal residual lesion, its unusual location and asymptomatic nature distinguishes it from other notochord-derived malignancies.[2] It is commonly found in the clivus and sacrum. Most symptomatic EP lesions (77%) are located in the prepontine region, while a significant minority (17%) are observed in the posterior wall of the sphenoid sinus. Notably, the majority of cases situated in the posterior sphenoid sinus wall were females, accounting for 90% of these occurrences.[3]

Histologically, EP resembles chordoma and is composed of notochordal remains with physaliferous (bubbly) cells inside a myxoid stroma. It usually lacks mitotic activity or cellular atypia and shows no malignancy markers that are associated with chordoma. Both EP and chordoma arise from notochordal tissue; however, EP does not exhibit invasive growth.[4] EP can present with various clinical symptoms, including headache, facial pain, cerebrospinal fluid (CSF) leak, and meningitis. The two largest systematic reviews included 47 and 60 cases of symptomatic EP, with headache and rhinorrhoea being the most common initial presenting symptoms,3,5 with spontaneous rhinorrhoea accounting for 35% of symptomatic EP cases.[5] Meningitis due to CSF leakage has also been reported.[6]

Other less common symptoms include diplopia, abducens nerve palsy, and intracranial hypertension.[7] In the literature there are only 24 reported cases of EP presenting as spontaneous CSF rhinorrhoea (Table 1).[5,6,8-19] The majority of those patients were female (79%) with an average of 53 years. The most common location was the clivus, and almost half of them reported a history of meningitis (Table 1). It is important to note that EP can be asymptomatic and incidentally detected during imaging or autopsy.[20]

Accurate diagnosis and differentiation from other lesions, particularly chordomas, are crucial for appropriate management. Histopathological analysis might not distinguish between EP and other malignant lesions like clivus chordoma, given their shared embryonic origins, leading to nearly identical histopathological characteristics. Both conditions exhibit similar features, including the presence of physaliphorous cells when examined under electron microscopy, along with comparable immunohistochemical patterns.[21] Neuroradiological studies play a crucial role in the diagnosis and differentiation of EP. On MRI, EP typically appears as a well-defined retroclival cystic lesion with hypointense signal on T1-weighted images and hyperintense signal on T2-weighted images. The lesion shows cerebrospinal fluid (CSF) signal on T2-FLAIR images and does not demonstrate abnormal enhancement. Importantly, EP can be distinguished from chordoma, its pathologic counterpart, based on imaging characteristics such as the absence of contrast enhancement or osseous destruction.[22]

Surgical resection is the preferred treatment for symptomatic EP. Various surgical approaches have been employed, including endoscopic endonasal transsphenoidal trans clival approach which has been reported as a safe and effective technique for resection and addressing CSF leaks.[6,21] However, for asymptomatic lesions, a reasonable management approach involves periodic radiological monitoring, and operative intervention should be considered once symptoms manifest in the patient.[21]

CONCLUSION

This case describes a 50-year-old female with a spontaneous CSF rhinorrhoea as the initial presenting symptoms of EP. Imaging plays an important role in the diagnosis of these lesion. Moreover, endoscopic trans-nasal skull base reconstruction is an effective management of the CSF leak. However, close follow-up is necessary to monitor the patient's progress and determine the exact nature of the clivus bone lesion when histopathology is not diagnostic.

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