Surgical Neurology International

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Case Report

Anterior clinoid mucocele presenting with orbital apex syndrome

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Received: 09 January 13 Accepted: 04 April 13 Published: 09 May 13

This article may be cited as:
Wang AC, Than KD, Ramnath S, Pandey AS. Anterior clinoid mucocele presenting with orbital apex syndrome. Surg Neurol Int 2013;4:63.
Available FREE in open access from: http://www.surgicalneurologyint.com/text.asp?2013/4/1/63/111583

Abstract

Background: Pneumatized anterior clinoid process is a common anatomic variant. Mucocele formation is a known complication of clinoid drilling during certain intracranial operations; however, mucoceles of pneumatized anterior clinoid processes have been found to spontaneously occur.

Case Description: A 44-year-old male presented with complaints of left-sided retro-orbital pain, double vision, and numbness over the upper face and scalp on the left side of 1-week duration. On examination, he was found to develop cranial nerve III, IV, and VI palsies with pupillary sparing, ophthalmic division cranial nerve V dysfunction, and eventually, the onset of vision loss.

Conclusions: We report a case of spontaneous anterior clinoid process mucocele presenting with orbital apex syndrome. This was treated successfully with anterior clinoidectomy for decompression.

Key Words: Anterior clinoid process, mucocele, orbital apex syndrome

Access this article online

Website: www.surgicalneurologyint.com

DOI: 10.4103/2152-7806.111583

Quick Response Code:

INTRODUCTION

Pneumatized anterior clinoid process (ACP) is a common incidental finding seen on cross-sectional imaging and intraoperatively. The sphenoid sinus varies in the extent of aeration along the sphenoid bone. Pneumatization begins in childhood and ends in adolescence. The sphenoid sinus drains into the superior meatus in the spheno-ethmoidal recess through variably sized, primarily membranous, ostia. Parasympathetic innervation of the mucus membranes lining the sphenoid sinus that controls secretion arises from the nearby pterygopalatine ganglion. Mucocele formation is a known complication of clinoid drilling during certain intracranial operations, and care must be taken to prevent this occurrence by sealing off any exposure to a pneumatized ACP when encountered. However, mucoceles of the paranasal sinuses are rarely found to spontaneously extend into the intracranial vault. As such, we report a case of anterior clinoid mucocele presenting with orbital apex syndrome (OAS).

CASE REPORT

A 44-year-old male with a history of diabetes and hypertension presented with complaints of left-sided retro-orbital pain, double vision, and numbness over the upper face and scalp on the left side of 1-week duration. He described being woken from sleep by stabbing left temporal headaches with associated diplopia, photophobia, nausea, and emesis. The following morning, he awoke with sensory numbness over his upper face and scalp on the left side. His headaches increased in intensity over the course of the week, which prompted him to seek medical attention.

On initial evaluation, the patient was a healthy-appearing male who was in obvious discomfort ascribed to his left retro-orbital pain. The left eye demonstrated resting esotropia and very slight infratropia. There was complete paralysis of left eye abduction, and incomplete paralysis of left eye adduction, while maintaining intact and full supraduction and infraduction. Right eye motility was normal. Pupillary constriction was brisk and symmetric bilaterally, visual field testing revealed no deficits, and dilated funduscopic examination revealed no papilledema. There was mild ptosis, and no noticeable chemosis or proptosis. Blood counts, serum metabolic testing, and cerebrospinal fluid analysis were all within normal limits.

Computed tomography (CT) and magnetic resonance imaging (MRI) were obtained [Figure 1], and the patient was referred for neurosurgical evaluation. Within 2 days, his neurologic examination progressed such that he demonstrated complete paralysis of abduction, adduction, and infraduction, with incomplete paralysis of supraduction and further ptosis of the left eye. He developed resting esotropia, infratropia, and anisocoria, and his left pupil became sluggish and less-reactive to light. He continued to demonstrate diminished pinprick, light touch, and vibratory sensation in the left V1 distribution. Finally, nonspecific superior and inferior visual field deficits were discovered on visual field testing.

The patient was thus administered high-dose dexamethasone therapy, and underwent a left pterional craniotomy with anterior clinoidectomy and resection of the mass lesion. An expanded ACP with thinned cortical bone was noted upon visualization of the mass. The mass lesion was soft, and thick green fluid was drained from the lesion. The lesion appeared to originate from within the sphenoid sinus. The superior orbital fissure and optic canal were confirmed to be well-decompressed, and the point of communication into the sphenoid sinus was sealed with a pericranial graft and fibrin glue [Figure 2]. Histopathologic evaluation confirmed a mucocele, showing the presence of ciliated epithelium and dense connective tissue with extensive lymphocytic infiltration.

Following resection of this ACP mucocele, the patient noted almost immediate improvement of vision in his left eye. Visual acuity returned to 20/20 in both eyes, though he had a few scattered spots of visual deficits in the superior hemi-field of the left eye, as demonstrated by Humphrey visual field testing. Within 8 weeks, his extraocular motility had returned to nearly full strength in every direction (he demonstrated approximately 90% abduction of both eyes), with no ptosis. While he had some relapse of ptosis of the left eye, his vision remained stable following decompression, and he has had no recurrence in over 2 years.

DISCUSSION

Gray's anatomy[13] describes the sphenoid sinus as occasionally extending into the greater and lesser wings of the sphenoid bone. At birth, the sphenoid sinus is merely an extension of the nasal cavity into the sphenoidal conchae. During early childhood, the sphenoid sinus begins to develop, reaching its full size in adolescence. A posterior ethmoid sinus can also invade the sphenoid, described as an Onodi cell. Internal expansion or resorption can lead to incompetence of the sinus wall, creating a direct connection between the sinus and the intracranial space.

Aerated ACP is uncommon, found in only 6-24% of cases on imaging studies, occurring bilaterally in over 50% of these cases.[1,3,5,18,25] One theory as to the formation of sphenoid sinus mucoceles involves the incorporation of aberrant mucinous tissue within the developing bone.[20] Such an explanation is unlikely, however, given that the sphenoid sinus does not form until approximately 4 years of age. ACP mucoceles have not been reported in the pediatric patient population, although children do demonstrate a propensity to suffer upper respiratory tract infections and sinonasal inflammatory conditions.

The more plausible hypothesis is one of repeated inflammation leading to obstruction of the ACP outlet. Another theory regarding the formation of mucoceles of the paranasal sinuses involves inflammation in, and the subsequent obstruction of, the drainage outlets – germane to this discussion, the outlet from an aerated ACP. The loculated, mucus-lined chamber then progressively enlarges with mucus production, causing bone erosion and possibly mass effect.[22] In our patient, the contralateral ACP was aerated, and it is likely that the ipsilateral ACP had been previously aerated prior to mucocele formation. We suspect this to be the mechanism by which our patient's ACP mucocele arose.

A relatively rare occurrence, sphenoid sinus mucoceles comprise an estimated 1% of all paranasal sinus mucoceles.[17,23] Sphenoid sinus mucoceles involving an aerated ACP have been sparsely reported in the past, with the majority presenting with vision loss as the primary complaint.[2,6,7,9,10,12,14,15,19,21,22,24] Ophthalmoparesis at presentation has been reported less frequently.[8,11,16] In this report, we describe a spontaneous ACP mucocele presenting as OAS, progressing to cause mass effect upon the ipsilateral optic nerve. Because of its nonspecific imaging characteristics and the uncommon pathology found on neurologic examination at presentation, we reviewed the existing literature to compare previously described cases of ACP mucocele [Table 1].

Medical management using empiric antibiotics has been described.[9] We disagree with this method of treatment in cases involving visual symptoms for two reasons. First,

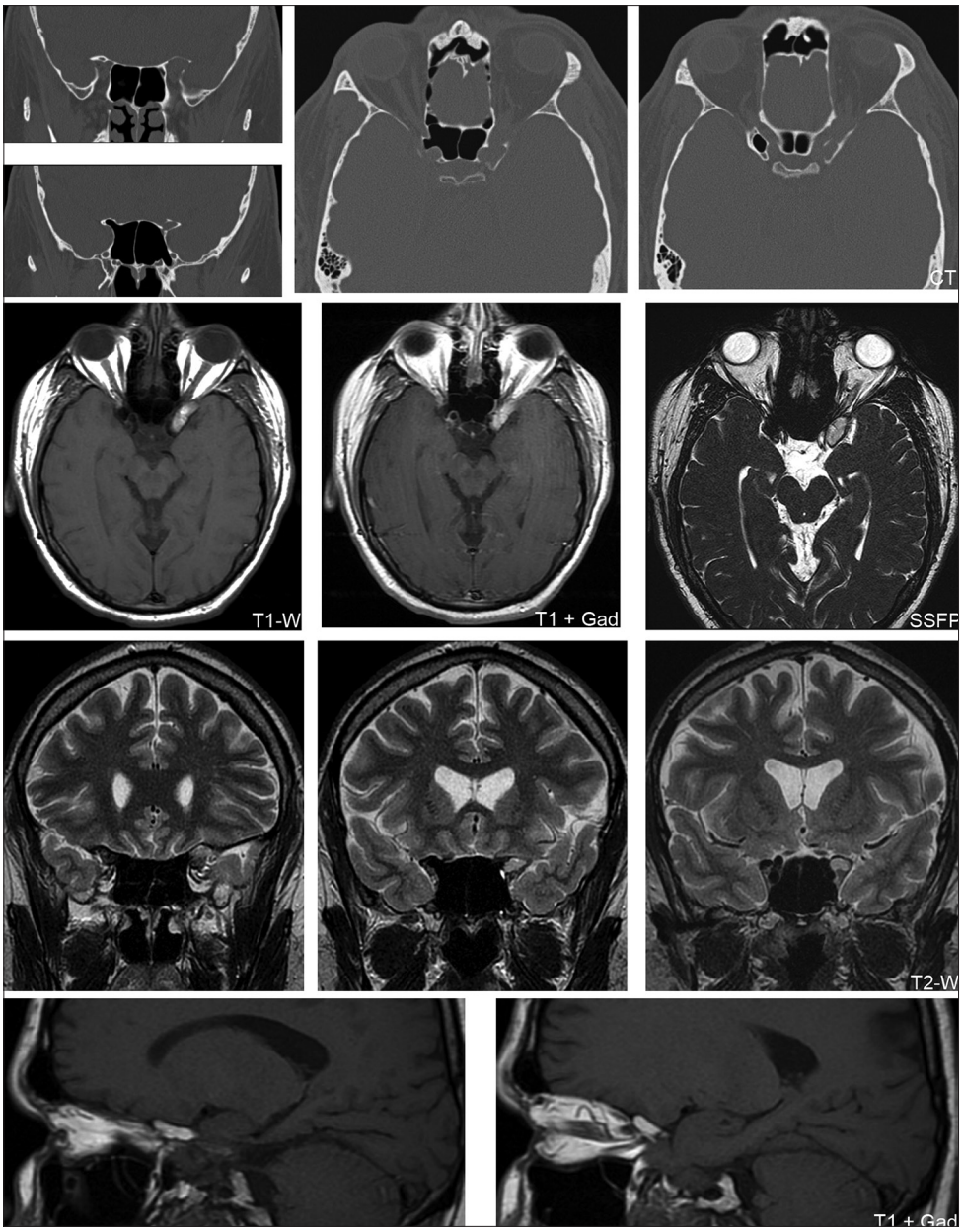

Figure 1: Top row: Coronal and axial CT of the head reveal an expansile lesion expanding the roof of the left optic canal and opacifying the left ACP. Of note, the right ACP is aerated. Row 2: Axial MRI of the brain demonstrates a lesion adjacent to the left ACP. T1-weighted fast spin echo sequences with and without gadolinium show no discernable contrast enhancement. Suppression of fat attenuation showed no changes in the appearance of the hyperintense lesion (not depicted). Rows 3 and 4: Coronal and sagittal T2-weighted and steady-state free precession sequences show mass effect upon the left optic nerve at the level of the orbital apex, and upon the anterolateral aspect of the left cavernous sinus

in the majority of reported cases, there was evidence of ipsilateral optic canal involvement even prior to the onset of visual symptoms, as seen in our case. The incidence of vision loss with symptomatic ACP mucocele is probably quite high, though the number of cases seen is too small to determine this with any certainty. Importantly, urgent decompression has met with significant recovery of some visual capacity.[6,19,26] Second, if the cumulative effect of several bouts of inflammation has led to the obstruction, then antibiosis is unlikely to have any decompressive effect, even if empiric coverage is adequate to cover the wide range of possible causative organisms. We do feel that a trial of antibiotics is warranted in patients with findings suggestive of acute bacterial infection and no neurologic deficits. However, given the apparent effectiveness of surgical decompression, such action should be performed with urgency when faced with a progressive neurologic deficit resulting from an ACP mucocele.

CONCLUSION

Here, we report a case of spontaneous ACP mucocele presenting with OAS. During the course of work-up,

Table 1: Summary of reported cases of mucocele of anterior clinoid process from the literature

Author	Patient age (years)	Sex	Clinical findings	Imaging findings	Treatment approach	Outcome
Arnavielle, 2010[2]	37	M	Pain, progressive VL, papilledema, APD	Pneumatized ACP with erosive nonenhancing mass on CT and MRI	Endoscopic endonasal	Full recovery
Bahadir, 2011[4]	72	–	Progressive VL	Pneumatized ACP with erosive nonenhancing mass on CT and MRI	Endoscopic endonasal; ceftriaxone	Slight improvement in vision
Chagla, 2010[6]	38	F	HA, progressive VL	Pneumatized ACP with expansile nonenhancing mass on CT and MRI	Supraorbital craniotomy	Significant improvement in vision
Chou, 1999[7]	68	M	HA, progressive VL, CN V1, CN VI	Pneumatized ACP with eroded optic canal on CT from a minimally enhancing mass on MRI	Supraorbital craniotomy	Slight improvement in vision, progressive optic nerve atrophy
Chung, 1999[8]	48	F	HA, diplopia	Pneumatized ACP with optic canal erosion from a nonenhancing mass on CT and MRI	Pterional craniotomy	Full recovery
Deshmukh, 2007[9]	20	F	Sinusitis, stable VL	Pneumatized ACP with nonenhancing soft tissue mass without optic canal bone destruction on CT and MRI	Antibiotics	Full recovery
Dunya, 1996[10]	32	M	Vertical diplopia, progressive VL, cecocentral scotoma, color vision loss, APD	Nonenhancing mass involving inferior aspect of optic canal on MRI	Endoscopic endonasal	Full recovery
Forer, 2010[11]	50	M	Pain, conjunctival injection, ophthalmoparesis, APD	Pneumatized ACP with erosive, expansile nonenhancing mass on CT and MRI	Endoscopic endonasal	Full recovery
Garaventa, 1997[12]	29	F	HA, stable VL	Erosive, expansile nonenhancing mass on CT and MRI	Endoscopic endonasal	Full recovery
Johnson, 1986[14]	56	M	Pain, progressive VL, APD	Erosive, expansile mass involving optic canal and superior orbital fissure on CT	Pterional craniotomy	Slight improvement in vision
Johnson, 1986[14]	59	M	Episodic slowly progressive VL, APD	Pneumatized ACP, expansile soft tissue mass on CT	None	No changes
Kwon, 2009[15]	52	M	Sudden onset VL	Pneumatized ACP, expansile nonenhancing mass on CT and MRI	Endoscopic endonasal	Full recovery with transient mild VL treated with steroids and antibiotics
Lim, 1999[16]	61	M	Diplopia, CN IV, CN V1	Pneumatized ACP, erosive mass involving optic canal and superior orbital fissure on CT, with minimal peripheral enhancement on MRI	OZ craniotomy	Full recovery
Nundkumar, 2012[19]	32	M	VL	Pneumatized ACP, expansile nonenhancing mass compressing optic canal	Endoscopic endonasal	Full recovery
Righini, 2006[21]	18	F	Episodic VL	Fibrous dysplasia, nonenhancing mass on CT and MRI	Endoscopic endonasal	Full recovery
Schwaighofer, 1989[22]	34	F	Long-standing pain, sudden VL	Pneumatized ACP, expansile nonenhancing mass on CT and MRI	Frontal craniotomy	Full recovery
Thurtell, 2007[24]	50	F	Pain, progressive VL, APD	Pneumatized ACP, expansile mass on CT, with slight peripheral enhancement on MRI	Pterional craniotomy	No recovery; optic nerve atrophy
Vaphiades, 2007[26]	36	M	Progressive VL, central scotoma, APD	Pneumatized ACP, expansile nonenhancing mass on CT and MRI	Endoscopic endonasal	Significant improvement in visual acuity and color vision

ACP: Anterior clinoid process; APD: Afferent pupillary defect; HA: Headache; OZ: Fronto-temporo-orbito-zygomatic; VL: Vision loss

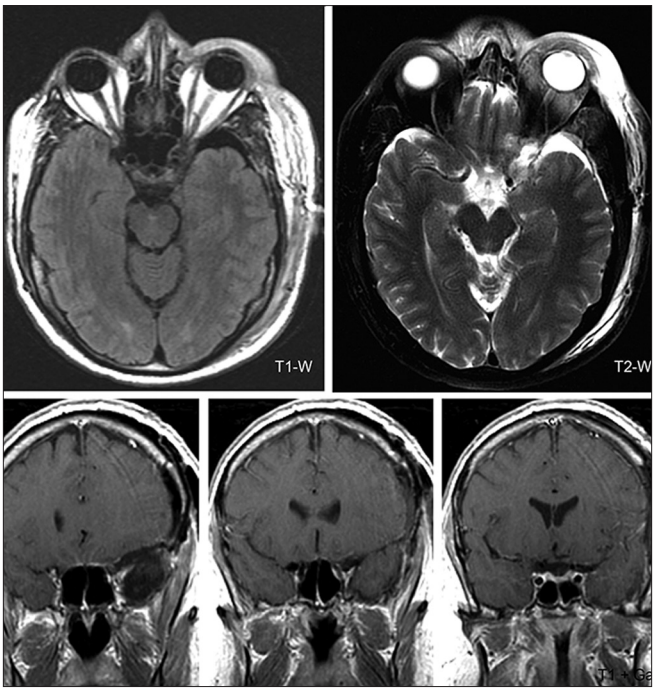

Figure 2: Top row: Axial T1- and T2-weighted MRIs of brain demonstrate removal of the lesion adjacent to left ACP. The left optic nerve is visible and well-decompressed. Bottom row: Coronal postgadolinium T1-weighted MRIs show resolution of mass effect upon left optic nerve at the orbital apex and cavernous sinus

the patient's symptoms quickly progressed to include vision loss as well as worsened cranial neuropathies. An intracranial approach was taken for decompression of the nerves, resection of the lesion, and pathologic diagnosis. A review of the literature revealed few reports of ACP mucoceles. Incidence, natural history, and treatment outcomes are as yet unknown. However, it appears that visual symptoms are common with ACP mucocele, while oculomotor symptoms are possibly less common. Relatively urgent nerve decompression must be considered in each case presenting with neurologic deficit. Options for decompression include medical management, open cranial and cranio-facial approaches, and endoscopic sino-nasal approaches.

ACKNOWLEDGEMENTS

The authors would like to thank Holly Wagner for editorial assistance.

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