# Rhinogenic Meningitis Caused by Congenital Petrous Apex Cholesteatoma: Simultaneous Surgical Treatment through Transotic and Transsphenoidal Approach

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Case Report

# RHINOGENIC MENINGITIS CAUSED BY CONGENITAL PETROUS APEX CHOLESTEATOMA: SIMULTANEOUS SURGICAL TREATMENT THROUGH TRANSOTIC AND TRANSSPHENOIDAL APPROACH

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SUMMARY – A 66-year-old male patient was admitted due to high fever, severe headaches and disturbance of consciousness. Meningitis was confirmed by lumbar puncture and intravenous antimicrobial therapy was started. Since he had undergone radical tympanomastoidectomy 15 years before, otogenic meningitis was suspected, so the patient was referred to our department. Clinically, the patient manifested watery discharge from the right nostril. Microbiological analysis verified Staphylococcus aureus in a cerebrospinal fluid (CSF) sample acquired by lumbar puncture. Radiological work-up, including computed tomography and magnetic resonance imaging scans, showed an expanding lesion of the petrous apex of the right temporal bone disrupting the posterior bony wall of the right sphenoid sinus with radiological characteristics indicating cholesteatoma. Those findings confirmed rhinogenic meningitis caused by expansion of the petrous apex congenital cholesteatoma into the sphenoid sinus, allowing nasal bacteria to enter the cranial cavity. The cholesteatoma was removed completely by the simultaneous transotic and transsphenoidal approach. Since the right labyrinth was already non-functional, there was no surgical morbidity after labyrinthectomy. The facial nerve remained preserved and intact. The transsphenoidal approach enabled removal of the sphenoid portion of the cholesteatoma and two surgeons met together at the level of the retrocarotid segment of the cholesteatoma, completely removing the lesion. This case represents an extremely rare condition in which a petrous apex congenital cholesteatoma expanded through the petrous apex to the sphenoid sinus, causing CSF rhinorrhea and rhinogenic meningitis. According to available literature, this is the first case of petrous apex congenital cholesteatoma causing rhinogenic meningitis successfully treated with the simultaneous transotic and transsphenoidal approach.

Key words: *Petrous apex; Congenital cholesteatoma; Rhinogenic meningitis; Simultaneous approach, transotic, transsphenoidal*

## Introduction

Petrous bone cholesteatomas are defined as cholesteatomas localized medially to the otic capsule involving petrous portion of the temporal bone[1]. They can be distinguished as congenital or acquired. Congenital

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cholesteatomas arise from embryonic rests deposited in the temporal bone following abnormal development of the first branchial groove, whereas the acquired ones originate from the tympanomastoid region and invade medially through the labyrinth[2].

Cholesteatomas show locally aggressive nature by eroding the ossicular chain and surrounding bones, causing conductive hearing loss, and subsequently sensorineural loss due to damage to the inner ear structures. Further expansion of cholesteatoma may cause sigmoid sinus thrombosis[3], mastoiditis, facial nerve palsy[4], cerebrospinal fluid (CSF) leak, meningitis, or even brain abscess[5].

According to their extension, petrous bone cholesteatomas can be divided into 5 types: supralabyrinthine, infralabyrinthine, massive labyrinthine, infralabyrinthine-apical, and apical[6]. In order to include extratemporal extension sites, three subclasses are subsequently added to the initial classification, comprising petrous bone cholesteatoma extension to the clivus, sphenoid sinus, and nasopharynx[7]. These subclasses are of high significance in choosing the right surgical approach. Furthermore, Moffat *et al.* proposed a more comprehensive classification which is somewhat expanded Sanna's classification, comprising supralabyrinthine, supralabyrinthine-apical, infralabyrinthine, infralabyrinthine-apical, massive labyrinthine, massive labyrinthine-apical, and apical cholesteatomas[2].

## Case Report

A 66-year-old male patient was admitted to the Emergency Department due to high fever, severe headaches and disturbance of consciousness. Meningitis was confirmed by lumbar puncture and intravenous antimicrobial therapy was started initially. Since he had undergone radical tympanomastoidectomy 15 years before, otogenic meningitis was suspected so the patient was referred to our department. Clinically, the patient manifested watery discharge from the right nostril. Otoscopic examination showed a normal trepanation cavity without secretion or signs of inflammation. Microbiological analysis verified *Staphylococcus aureus* in a CSF sample acquired by lumbar puncture. Computed tomography (CT) scan revealed inflammatory changes in the right sphenoid sinus showing communication with the apex of the temporal bone and a liquid content within the remaining right mastoid cells (Figs. 1 and 2).

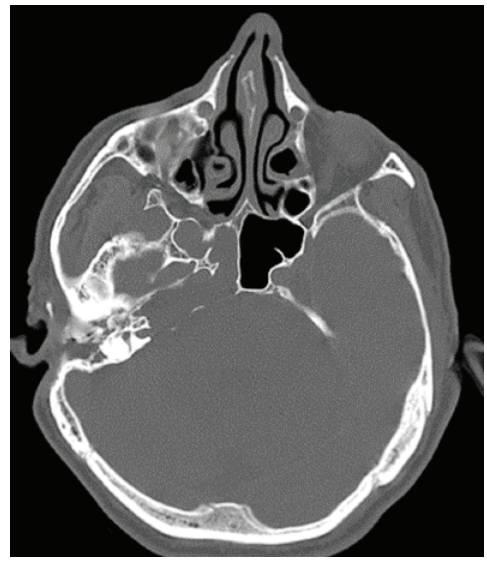

*Fig. 1. Axial CT scan showing opacity within the right sphenoid sinus with the posterior wall disruption, communicating with the apex of the right temporal bone. A liquid content is visible in the right mastoid cells along with the lateral wall disruption.*

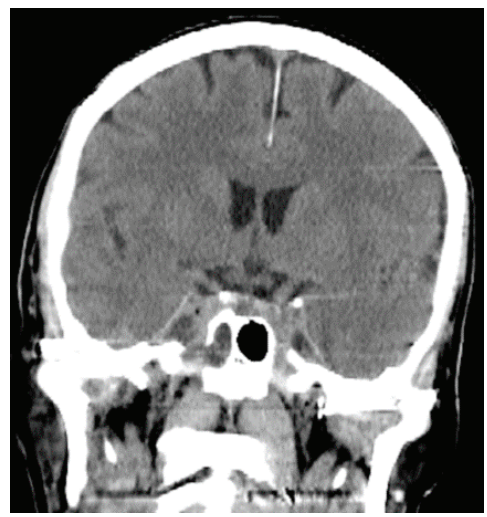

*Fig. 2. Coronal CT scan shows opacity within the right sphenoid sinus with a disrupted posteroinferior wall.*

Magnetic resonance imaging (MRI) scan showed an expanding lesion of the petrous apex of the right temporal bone disrupting the posterior bony wall of the right sphenoid sinus. It also disrupted the anterior wall of the internal acoustic meatus (Fig. 3). The lesion measured approximately

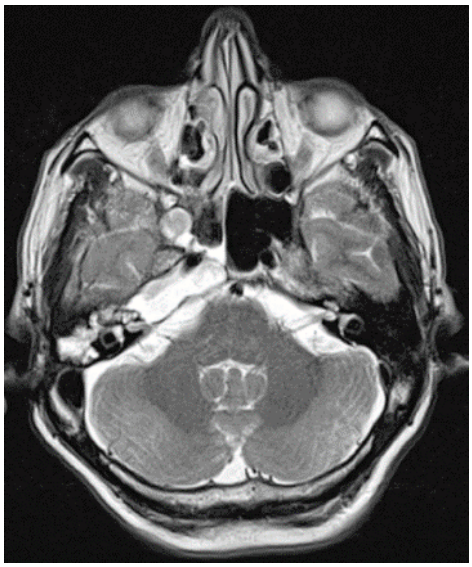

*Fig. 3. Axial MRI scan showing a lesion of the petrous apex of the right temporal bone disrupting the right half of the posterior wall of the sphenoid sinus and expanding within the right half of the sphenoid sinus. The lesion also disrupts the anterior wall of the internal acoustic meatus but without involving the internal acoustic canal.*

38x16x20 mm in diameter. MRI characteristics of the lesion were indicative of cholesteatoma.

These findings confirmed rhinogenic meningitis caused by expansion of the petrous apex congenital cholesteatoma into the sphenoid sinus, causing CSF rhinorrhea and allowing nasal bacteria to enter the cranial cavity. A surgical procedure combining a simultaneous transotic and transsphenoidal approach was indicated. The transnasal approach included elevation of a nasoseptal flap, wide right sided sphenoidotomy, and removal of the anterior part of the inter-sphenoid septum and mucosa of the sphenoid sinus. A defect in the posterior wall of the sphenoid sinus was observed along with CSF leak and a protruding cholesteatoma (Fig. 4).

The cholesteatoma was subsequently removed up to the level of the retrocarotid area. Previously, canal wall down tympanomastoidectomy was performed because of chronic otitis media. Retro auricular approach was made; a blind sac closure of the right external auditory canal was performed. Revision of the trepanation cavity was made; mucus retention was found in the remaining mastoid cells but without cholesteatoma. Labyrinthectomy was subsequently performed, the cochlea was removed without rerouting the facial

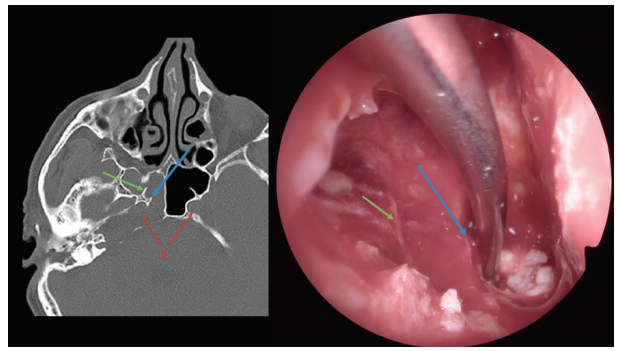

*Fig. 4. Endoscopic view of a cholesteatoma within the right sphenoid sinus protruding through a disrupted posterior wall. Blue and green arrows show operative landmarks correlated with CT scan (C, internal carotid artery; curved suction tube is inserted behind the carotid artery).*

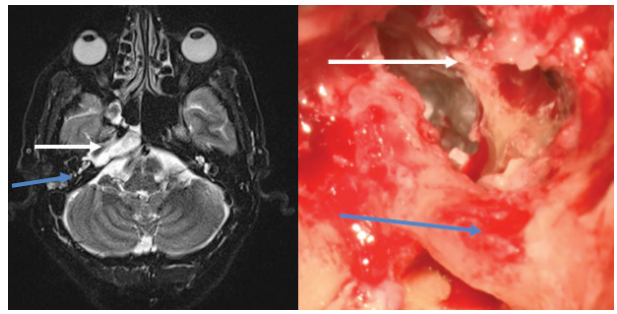

*Fig. 5. Microscopic view of the cholesteatoma surrounding the internal carotid artery correlated with axial MRI scan section (blue arrow represents the prominence of the facial canal and white arrow represents the internal carotid artery surrounded by cholesteatoma).*

nerve. A cholesteatoma was observed medially to the cochlea, spreading back to the internal auditory canal and anteriorly to the sphenoid sinus. The cholesteatoma eroded the carotid canal and surrounded the internal carotid artery (Fig. 5).

The transsphenoidal approach enabled removal of the sphenoid portion of the cholesteatoma and both surgeons met together at the level of the retrocarotid segment of the cholesteatoma, completely removing the lesion. The fused surgical cavity and the sphenoid sinus were obliterated by autologous fat tissue. The sphenoid sinus was also covered with a nasoseptal graft.

There were no complications during the operation or in the postoperative course. CSF leak was successfully solved. A follow up CT scan was performed on

postoperative day 4 showing a satisfactory result without intracranial hematoma. The patient was receiving inpatient antimicrobial treatment for the next 2 weeks and was subsequently discharged in a good general condition. Since the right labyrinth was already non-functional, there was no surgical morbidity after labyrinthectomy. The facial nerve remained preserved and intact.

## Discussion

This article presents an extremely rare case of a patient with a petrous apex congenital cholesteatoma involving the right sphenoid sinus, who arrived at the Emergency Department with rhinogenic meningitis.

Algorithms in petrous bone cholesteatoma treatment emphasize 5 important considerations, including complete eradication of the lesion, preservation of the facial nerve function, CSF leak and meningitis prevention, cavity obliteration, and hearing preservation when possible[7].

Patients with petrous bone cholesteatoma most frequently present with hearing loss in the affected ear or facial nerve paralysis[2,7-9]. By thoroughly reviewing the available literature, we conclude that no similar case has been published so far. To the best of our knowledge, we found two somewhat similar cases in which two patients with petrous bone cholesteatoma presented with CSF rhinorrhea, but the pathophysiology of their condition involved a fistula in the internal auditory canal, which was to be seen intraoperatively and probable CSF leakage through the eustachian tube and nasopharynx[10,11]. Neither of the patients presented in these two studies had rhinogenic meningitis.

Since the patient had an irreversible hearing loss, the transotic approach was indicated[6,7], combined with the transsphenoidal approach[12], in order to completely remove the sphenoid portion of the cholesteatoma. The facial nerve function was normal preoperatively, so we decided to use the transotic approach and were able to completely remove the pathology without rerouting the facial nerve. Preoperative and postoperative facial nerve function was assessed according to the House-Brackmann grading system and showed a completely normal result, i.e., grade I, before and after the procedure[13]. CSF leak that occurred after the cochlea was drilled, was repaired by fat tissue obliteration, so there was no CFS leakage to be seen in the postoperative period.

The available literature proposes the transtemporal or middle fossa approach in treating petrous apex cholesteatoma in patients with an irreversible hearing loss due to good visualization and efficient removal of the lesion[2,7]. Since complete removal of the petrous apex cholesteatoma expanding to the sphenoid sinus may be difficult by using only the transtemporal approach, Aubry *et al*. proposed the combined transtemporal and transsphenoidal approach[8,14]. Compared to the middle fossa approaches, these two techniques show less morbidity and complications in terms of craniotomy avoidance, faster recovery, and shorter hospitalization[8,15]. Transotic, infracochlear, as well as retrocarotid transsphenoidal approach are used and very well described for evacuation of cholesterol granulomas of the petrous apex. However, in cases of congenital cholesteatoma, radical removal of the cholesteatoma is mandatory, so wider exposure of the petrous apex is usually required.

In our case, since the facial nerve was normal preoperatively, we decided to use the transotic approach which was sufficient for complete removal of the pathology. Furthermore, endoscopic petrous bone surgery enables direct visual control of microscopically hidden areas, ensuring better removal and lower recurrence probability[16].

## Conclusion

This was an extremely rare case of a patient with petrous apex congenital cholesteatoma presenting with rhinogenic meningitis. Given its localization and expansile potential, the petrous apex cholesteatoma extended to the sphenoid sinus, disrupted the posterior wall of the sphenoid sinus and involved the right sphenoid sinus, enabling nasal bacteria to ascend to the cranial cavity, causing rhinogenic meningitis. According to our knowledge, there has not been any similar case published so far. Since the patient was successfully treated by combined transsphenoidal and transotic approach, we endorse that endoscopic assistance should be implemented in petrous bone cholesteatoma treatment due to better visualization of hidden areas, less morbidity compared to the open surgical procedures, and faster recovery of the patient. Further studies on this topic could be helpful in order to include endoscopic approach in petrous bone cholesteatoma treatment guidelines. Moreover, in order to implement new endoscopic surgical approaches to the petrous apex pathology and minimize the possible risks of damaging

important anatomic structures, it would be of major importance to conduct further studies on the anatomy and morphological variations of the sphenoid bone as well[17].

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Sažetak

## RINOGENI MENINGITIS UZROKOVAN KONGENITALNIM KOLESTEATOMOM APEKSA PIRAMIDE: SIMULTANO KIRURŠKO LIJEČENJE TRANSOTIČKIM I TRANSSFENOIDNIM PRISTUPOM

*M. Čukman, J. Ajduk, L. Bukovac* i *M.V. Grgić*

Bolesnik u dobi od 66 godina primljen je zbog visoke temperature, jake glavobolje i poremećaja svijesti. Lumbalna punkcija potvrdila je meningitis i započeta je intravenska antibiotska terapija. Budući da je 15 godina ranije kod bolesnika rađena radikalna timpanomastoidektomija, postavljena je sumnja na otogeni meningitis te je upućen na našu Kliniku. Nakon primitka je uočena desnostrana rinolikvoreja, a ponovljenom lumbalnom punkcijom u likvoru je izoliran *Staphylococcus aureus*. Radiološka obrada uključujući kompjutoriziranu tomografiju i magnetsku rezonancu pokazala je ekspanzivnu leziju vrha piramide desne temporalne kosti s destrukcijom stražnje stijenke sfenoidnog sinusa i radiomorfološkim osobinama kolesteatoma. Ovi nalazi potvrdili su da se radi o rinogenom meningitisu koji je uzrokovan širenjem kongenitalnog kolesteatoma vrha piramide u sfenoidni sinus, što je omogućilo prodor bakterija iz nosa u endokranij. Kolesteatom je u cijelosti odstranjen simultanim kombiniranim transotičkim i transsfenoidnim pristupom. Budući da je desni labirint od ranije bio nefunkcionalan, nije bilo kirurškog morbiditeta nakon labirintektomije. Očuvani su integritet i funkcija ličnog živca. Transsfenoidni pristup omogućio je odstranjenje sfenoidnog dijela kolesteatoma, a oba operatera susrela su se na razini retrokarotidnog segmenta kolesteatoma koji je tako odstranjen u cijelosti. Ovaj bolesnik predstavlja iznimno rijedak slučaj kod kojega se kongenitalni kolesteatom vrha piramide proširio u sfenoidni sinus uzrokujući rinolikvoreju i rinogeni meningitis. Prema dostupnoj literaturi, ovo je prvi takav slučaj koji je ujedno uspješno liječen simultanim transotičkim i transsfenoidnim pristupom.

Ključne riječi: *Apeks piramide; Kongenitalni kolesteatom; Rinogeni meningitis; Simultani pristup, transotički; transsfenoidni*