

A case of Atlanto-axial instability following Skull base osteomyelitis: tips for diagnosis and management

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Abstract

Here we discuss a diabetic 54 years old female presented with malignant otitis externa and bilateral facial paresis followed by cervical spondylitis and C1-C2 instability. Skull base osteomyelitis confirmed by clinical presentation, imaging, and lab data. Fortunately, she responded well to anti-bacterial and antifungal therapy.

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A case of Atlanto-axial instability following Skull base osteomyelitis: tips for diagnosis and management

Abstract

Introduction: Skull base osteomyelitis is a rare but dangerous consequence of untreated malignant otitis externa. *Pseudomonas aeruginosa* is responsible for most cases with typical presentation.

Case report: Here we discuss a diabetic 54 years old female presented with malignant otitis externa and bilateral facial paresis followed by cervical spondylitis and C1-C2 instability. Skull base osteomyelitis confirmed by clinical presentation, imaging, and lab data. Fortunately, she responded well to anti-bacterial and antifungal therapy.

Discussion and conclusion: Due to limited data, there is no confirmed standard of treatment for instability secondary to SBO. It seems antibiotic therapy is the mainstay of treatment. In case of poor response to

antibiotic therapy surgical intervention is inevitable. This article introduces the first case of SBO-related AAI managed with a conservative treatment modality.

key words:

Otitis externa, Osteomyelitis, Skull base, Cervical instability

Introduction

Skull base osteomyelitis (SBO) is an uncommon condition. It can be divided into two groups using origin and bony anatomic involvement. The typical group is mainly related to malignant otitis externa, which usually involves the temporal bone. The atypical or central group commonly involves sphenoid and occipital bone without a determined otologic infection¹.

Delay in treatment is related to poor prognosis. The physician should be familiar with this condition; otherwise, it can be missed, and extensive morbidity and mortality will be forced onto patients². SBO is more common in immunocompromised patients or elderly with comorbidities like renal failure or diabetes mellitus(DM)³. SBO is usually bacterial; the most common germ is *Pseudomonas aeruginosa*, especially in otic-origin infections. Due to vital vascular and neurological element proximities, complicated conditions are not rare in these patients⁴.

Here we present a case of complicated fungal skull base osteomyelitis in a female with uncontrolled diabetes mellitus.

Case presentation

A 54 years old female presented to the otolaryngology department with a complaint of bilateral facial palsy. The patient developed diabetes twenty years ago and was not well controlled (last HbA1c was 9.2 %). she had multiple otitis externa in the preceding six months, which has been partially treated with short courses of oral and topical antibiotics. She complained of severe headaches and bilateral ear pain for the last four months and had developed right-side facial palsy two months ago, followed by left-side facial palsy two weeks before our visit. She had low-grade fevers, and external canal redness, inflammation, and granulation tissue were evident on the ear exam. She was admitted to the infectious disease department for more evaluation. CRP and ESR were high (63 and 95, respectively). Leukocytosis was evident with a WBC count of 15300. A smear sample of ear discharges was obtained, and after blood culture specimens, intravenous broad-spectrum antibiotics started (Meropenem 1 gr TDS and Vancomycin 1 GR BD). Brain and temporal bone CT scans and MR imaging were done. CT scan revealed evidence of skull base osteomyelitis as bilateral temporal bone sclerosis. In MR imaging, Soft tissue hyperintensity extending from the middle ear to the skull base was seen in T2 MRI with contrast enhancement after gadolinium injection (figure 1). Brain MRI was interpreted as an inflammatory/infectious process. After two weeks, the patient continued to have periodic low-grade fevers and no significant decrease in inflammatory markers. The blood and discharge specimens' cultures were negative. Anti-fungal Intravenous Voriconazole was added, and the Patient continued to receive intravenous antibiotics for additional four weeks, followed by oral antibiotics (Levofloxacin 750mg daily and Voriconazole 200 mg q 12 hours) after discharge. Left side facial palsy improved completely in follow-up visits, and right side paresis decreased but was still persistent as House Brackman grade of 3. CRP and ESR levels decreased (to 24 and 46, respectively), but she continued to complain of headaches. After six months, the patient was referred to the neurosurgery department, where she reported progressive cervical pain. Cervical ROM was severely limited, and the patient insisted on keeping her head motionless. There was no radiating pain, and the sensory and motor examinations were normal. Deep tendon reflexes were normal, and there was no Gait or balance disturbance on examination. There was no midline tenderness. No cervical lymphadenopathy was palpated. There was no jaw tenderness or jaw movement limitation. She denied any history of trauma. Ear pain and discharges were improved. Right-side hearing loss was explicit.

Dynamic Cervical radiography was done. C1- C2 instability was diagnosed as an increased Atlanta-dental interval at flexion (about 5 mm) (figure 2, 3). Bed rest, analgesics, and a rigid cervical collar were prescribed, and a cervical spine CT scan and MRI was requested. Sclerosis of upper cervical vertebrae and erosions

in the odontoid process was reported in the CT scan. MRI revealed bone marrow edema of bodies of upper cervical vertebrae without the involvement of intervertebral disks indicating cervical spondylitis. The patient was admitted to the infectious ward Again. CRP and ESR levels were high (49 and 61), and PPD and PCR for TB infection and wright and coombs-wright for brucellosis were tested, which were negative. Hard Cervical Collar was applied, and Broad spectrum intravenous antibiotics were restarted (Meropenem and Vancomycin) in addition to intravenous antifungal (IV Voriconazole 200 mg q 12 h), which continued for four weeks. Then oral antibiotics continued for twelve months. At the 12-month follow-up, the pain was relieved significantly. At 1-year imaging, cervical spine edema and spondylitis disappeared in MRI, and C1-C2 stability was retained (figure 4).

Discussion

Skull base osteomyelitis (SBO) is a rare condition. Origins of infection in the skull are usually malignant otitis externa (MOE), paranasal sinusitis, odontogenic infections, and chronic mastoiditis. The primary germ causing SBO is *P. aeruginosa* in overall. In fungal SBOs, *Aspergillus* is the leading pathogen. MOE involving the temporal bone is the most common cause of SBO. MOE mainly involves the sphenoid and temporal bone. Although male to female ratio is more than 1 in MOE, it seems that SBO prevalence is the same in both sex⁵. MOE finds its way to the skull base from the external auditory canal(EAC) through the osseocartilaginous junction of the EAC and Santorini fissure. Skull base extension can affect cranial base foramina, leading to cranial neuropathies and temporomandibular joint involvement^{6 7}. The mastoid and middle ear cleft are known as the Lateral temporal bone, and the petrous apex is the medial temporal bone; both can be involved during MOE course⁸. The main complaint in MOE is disproportional otalgia. Tender EAC, preauricular cellulitis, and pinna woody induration are the most common signs in MOE⁹¹⁰.

On imaging, Patients with the erosion of the mastoid part of the facial canal are seven times more at risk of incidence of facial paresis¹¹. The most common cranial neuropathy in SBO is facial nerve involvement in the stylomastoid foramen. Jugular foramen involvement and lower cranial neuropathies manifestation are less common. Spreading to petrous apex can lead to 5th and 6th cranial nerves paresis⁶¹². WBC count, ESR, and CRP can be elevated. These inflammatory markers can be helpful as markers of treatment response¹³. MRI and SPECT are better studied for detecting the first stages of SBO. CT scan is not sensitive for detecting early SBO phases. However, it can help reveal periosteal reactions and bony erosions in the base of the skull¹⁴¹⁵. Tissue samples are essential for treatment. Using broad-spectrum antibiotics must be culture based; otherwise, they may not be effective. The best prognosis achieves through early diagnosis and intervention.

In immune-compromised patients, improving the immune system should be considered. In a diabetic patient, strict blood sugar level control is essential^{8 16}. Diabetes mellitus increases infection rate, but no data suggests more prolonged antibiotic therapy in these patients¹⁷. Hyperbaric oxygen(HBO) therapy is an adjunct treatment for refractory SBO. It can be used in a daily manner for several weeks. Important side effects are perforation of the tympanic membrane and barotrauma¹⁸¹⁹. Surgical debridement is helpful in extensive fungal SBOs, but in usual cases, the surgical role is limited to biopsy. In some cases of MOE, bone and granulation tissue debridement may be needed. Meatoplasty is indicated in extensive EAC involvement. Broad-spectrum antibiotics are the mainstay of treatment, and surgical intervention could be helpful in refractory conditions^{20 21}.

Typical SBO mainly involves the temporal bone, and the atypical entity may affect the occipital bone, especially the clivus. The clival area is connected to the fourth thoracic vertebra via epidural space²²²³. Clinical presentation can be different due to the extension of the infection. If it extends superiorly, cranial neuropathy, facial pain, and headache are the prominent symptoms. Inferior extension of SBO to the paravertebral region can affect the Occipitoatlantoaxial junction. It may present cervical pain, fever, suboccipital region tenderness, paresis, and craniovertebral junction instability^{2425 26}.

With aggressive treatment, the Neurologic morbidity rate has been reported in 31% of cases and mortality in about 10%. Diabetes mellitus is related to poorer prognosis^{27 28}.

Atlantoaxial instability, in our case, can be compared with Grisel syndrome. This syndrome is secondary

to an infection in the head and neck regions, leading to non-traumatic rotatory atlantoaxial subluxation. It is more common in the pediatric population and presents with torticollis. Adult cases are rare²⁹. The exact mechanism is unknown; however, it has been hypothesized that hyperemia secondary to infection spread through pharyngovertebral veins to periodontoid veins and cervical lymphadenitis may have roles in transient ligament laxity^{30 31}. In Grisel syndrome also, early detection is essential and delayed diagnosis after three weeks may increase the need for fusion surgery. It seems that with inflammatory process resolution, stability will be recovered. Recommended treatment is conservative management consisting of bed rest, anti-inflammatory therapy, muscle relaxants, and antibiotic therapy. Failure of Conservative management necessitates surgery and appropriate fusion^{29 30 31}.

Determining the optimal time to do surgery could be challenging. In case of progressive deformity, progressive neurologic decline, and refractory pain or instability despite appropriate antibiotic therapy, surgical management is inevitable^{32 19}. A summary of previous similar cases in the literature is shown in table 1.

In our case, there was no absolute indication for early surgery, so we chose non-surgical treatment and close follow-up.

Table 1. A summary of previously reported AAI following skull base osteomyelitis

Year	author	Age	Sex	Co-morbidity	Neurological exam	Discharge smear/culture	Response to antibiotic therapy	Surgical intervention
2020	Low et al. ³²	67	Male	DM type2	Limited cervical ROM	MSSA P.A	Poor response	Occipital arthrodesis
2020	Chefi et al. ¹⁹	35	Male	Negative	Normal	Coagulase-negative staphylococci	Poor response	Occipital arthrodesis
2022	Current study	54	female	DM type2	Limited cervical ROM, Cervical pain	Negative smear and culture	Good response	No

AAI: at- lantoax- ial instabil- ity, DM: diabetes mellitus, ROM: range of motion, MSSA: methicillin- resistant staphy- lococcus aureus, P.A: pseu- domonas aeruginosa								
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Conclusion

Skull base osteomyelitis is a rare but severe disease. Symptoms usually are not specific. It can be life-threatening if not diagnosed early and treated appropriately. Skull base foramina, cranial nerves, and vascular structure can be involved. Infection extension to the craniovertebral junction may lead to bone and ligament injuries and instability. In this case, proper antibiotic therapy with appropriate orthosis leads to the good outcome, but in some cases, fusion may be inevitable. This article introduces the first case of SBO-related AAI managed with a conservative treatment modality.

Disclosure of interest

The authors report there are no competing interest to declare.

Consent statement

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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