

Skull Base Osteomyelitis: A Rare Cause of Multiple Cranial Nerve Palsies—A Case Report from Ghana

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How to cite this paper: Kuuzie, E., Peki-Boateng, P.K., Yennah, A. and Duodu, F. (2023) Skull Base Osteomyelitis: A Rare Cause of Multiple Cranial Nerve Palsies—A Case Report from Ghana. *World Journal of Neuroscience*, 13, 257-265.

<https://doi.org/10.4236/wjns.2023.134018>

Received: September 21, 2023

Accepted: November 25, 2023

Published: November 28, 2023

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Abstract

Introduction: Skull Base Osteomyelitis (SBO) is an infectious inflammation of the skull bones that is often caused by malignant otitis externa (MOE) and affects the temporal bone. This condition commonly affects immunocompromised individuals and the elderly, particularly those with a history of diabetes mellitus. Diagnosis is challenging because of non-specific symptoms that lead to late detection and complications. This report discusses a case of SBO with multiple bilateral cranial nerve abnormalities and highlights the diagnostic and management challenges in high-risk individuals with subtle clinical signs. **Case presentation:** This report describes a 63-year-old patient with hypertension and diabetes who underwent surgical debridement of the left ear due to malignant otitis externa 4 months prior to presentation. The patient presented with significant dysarthria, dysphagia, ptosis of the left eye with double vision, and hearing impairment in the left ear. Examination revealed bilateral CN VI palsy, right CN VII palsy, left CN VIII palsy, and a right CN XII deficit. Initial tests were unremarkable, but a high Fungitell assay and a second review of the CT scan and MRI revealed a pathological process in the base of the skull involving bony structures and cranial nerves bilaterally, which helped diagnose SBO. The patient was subsequently discharged with oral voriconazole and continued his usual medications. The patient requested further management abroad, because he did not notice resolution of his symptoms. Surgical treatment was employed abroad to relieve his symptoms, as he recovered slowly. **Conclusion:** This case report underscores the importance of a multidisciplinary approach to address SBO. Collaboration between specialists in infectious diseases, otolaryngology, radiology, and neurology plays a pivotal role in achieving an accurate diagnosis and developing a tai-

lored treatment plan. Although SBO may be infrequent, this case report highlights the need to maintain heightened clinical suspicion in high-risk individuals.

Keywords

Skull Base Osteomyelitis, Cranial Nerves, Malignant Otitis Externa, Bulbar Palsy, Fungal Infection

1. Introduction

Skull Base Osteomyelitis (SBO) is a rare, complex, and fatal infection of the skull bones that commonly involves parts of the temporal, sphenoid, and occipital bones [1]. The disease was first described in 1959 by Meltzer and Keleman in a patient with pyocyanus chondritis and osteomyelitis of the external auditory canal [2]. Due to non-specific symptomatology, a long clinical course, and non-specific imaging, it is often initially misdiagnosed as a malignancy [3]. The clinical course is often a direct complication of sinogenic, odontogenic, otogenic, and rhinogenic infections when improperly treated in high-risk, immunocompromised patients. It is most commonly observed in immunocompromised patients and the elderly with pre-existing conditions, such as diabetes, arteriosclerosis, and hematological conditions, such as leukemia and lymphoma [1] [4] [5]. A myriad of non-specific findings such as headaches, severe otalgia, facial pain, conductive hearing loss, meningism, intracranial abscesses, and raised intracranial pressure, as well as cerebral venous thrombosis and strokes could be identified on presentation [6]. Malignant otitis externa (MOE) is the most common cause of SBO, particularly in temporal regions. SBO at atypical sites indicates other possible etiologies. Numerous studies have revealed that the predominant etiological agent responsible for MOE is *Pseudomonas aeruginosa*, a gram-negative bacterium, although instances of fungal origin have also been documented [1] [7]. A 2023 case-control study revealed that among patients with MOE, the prevalence of diabetes mellitus was 54.8%, and the adjusted odds ratio vs. control was 10.07 (95% CI) [8]. Despite several advances in antibiotics, diagnostic modalities and surgical techniques, the management of SBO is still a hard nut to crack [9]. In advanced cases, progression of the pathological process from the external ear to the skull can lead to cranial nerve palsy. This is because of the close proximity of the cranial nerves to the base of the skull. Mortality rates range from 28% to 60% among individuals with multiple cranial nerve deficits [10]. Patients typically experience lower cranial nerve palsy because the inflammatory process is located near neural foramina. When the stylomastoid foramen or mastoid process is affected, involvement of CN VII is inevitable, causing patients to present with facial pain and/or Bell's palsy. Sensorineural hearing loss is often accompanied by CN VIII, which is adjacent to CN VII in the internal acoustic meatus. Involvement of the petrous apex presents as binocular diplopia,

a rare presentation when the CN VI is affected. In rare cases, involvement of the jugular foramen leads to CN IX, X, and XI palsy, resulting in soft palate immobility, vocal cord paralysis, and inability to raise the arms above the horizontal plane, respectively. Furthermore, patients who develop problems with tongue movement show involvement of the hypoglossal canal [6] [11] [12].

Diagnosing Skull Base Osteomyelitis (SBO) is challenging due to its rarity and the similarity of symptoms to other neurological conditions. Treatment requires a multidisciplinary team collaboration, a long course of antibiotics, and in some cases, surgical debridement [3].

2. Case Presentation

We present the case of a 63-year-old male with known hypertension and diabetes who was referred to our facility for neurological consult four months after undergoing debridement surgery for malignant otitis externa involving his left ear. Two months post-surgery, the patient developed a sore throat, headache, dysphagia, dysarthria, left-sided hearing impairment, and ptosis of the left eye. Further examination revealed blurry double vision and weight loss, but no oral ulcers, skin rash, fever, chills, or prior head trauma. Owing to his inability to swallow, a percutaneous endoscopic gastrostomy (PEG) tube was inserted for feeding two weeks prior to his referral to our facility. The patient's family history was unremarkable and he was compliant with antihypertensive and antidiabetic medications. Additionally, he was administered neuroprotective medications from the referring facility (citicoline and piracetam). The patient worked as an engineer at a power plant and had no history of alcohol consumption, smoking, or recreational drug use. On presentation, he appeared chronically ill and wasted, and was clinically pale, anicteric, and afebrile with no features of dehydration. No lymphadenopathy was observed. Neurological examination revealed a conscious and alert man who was well-oriented in time, person, and place. He could actively move all limbs spontaneously, but was unable to speak and communicate through written notes. Detailed examination of the cranial nerves revealed bilateral CN VI palsy, predominantly affecting the left side, as well as palsy of the lower motor neuron right CN VII. A left CN VIII deficit revealed mixed-type, conductive, and sensorineural hearing loss. In addition, a right CN XII palsy was observed. All other cranial nerves were intact. Cardiorespiratory and abdominal examination results were unremarkable. Extensive research has been conducted to rule out systemic and autoimmune conditions. These include lumbar puncture, serum ACE, autoimmune panel, urine calcium tests, HIV serology, and D-dimer levels. All tests yielded normal results (**Table 1**), and the fungal assay revealed significantly elevated levels (120). Barium swallow showed normal esophageal motility, with no masses present.

Initial CT scan report: mild diffuse cerebral atrophy with mastoiditis.

Initial MRI report: mild periventricular seepage with sinusitis.

Chest and Neck CT scans: normal findings.

Table 1. Results of laboratory investigations.

Laboratory test	Value
C-reactive protein (CRP)	6 mg/L
ESR	35 mm fall/hour (4-7)
Hemoglobin	12.7 g/dL
White cell count	$4.95 \times 10^9/L$
Neut#	$3.45 \times 10^9/L$
Lymph#	$1.00 \times 10^9/L$
Mono#	$0.37 \times 10^9/L$
Platelet	$371 \times 10^9/L$

2.1. Differential Diagnosis

A working diagnosis at the time of admission was motor neuron disease, with differential diagnoses of carcinomatous meningitis, chronic basilar meningitis, neurosarcoidosis, and mono-neuritis multiplex.

2.2. Clinical Progress and Outcome

The patient's condition showed a progressive deterioration of vision. Ophthalmological assessment revealed binocular diplopia without evidence of optic nerve damage. Examination by the ear, nose, and throat team could not be performed because of the inability to visualize the tympanic membrane due to extensively impacted wax. However, no masses or granulation tissues were observed bilaterally in the external auditory canal. The patient initially received intravenous ciprofloxacin and clindamycin, which were later switched to voriconazole based on the results of the fungal assay. Despite a month of hospital stay, the patient's condition showed minimal improvement. The patient underwent PEG for feeding and received a hearing aid, which improved his hearing. However, the patient still experienced a persistent headache.

On discharge, the patient's blood pressure and blood sugar levels were well controlled with prescribed medications. He was discharged home with routine antihypertensive and antidiabetic medications, as well as oral voriconazole and analgesics which he was compliant on. The patient, although unable to verbalize, appeared to be depressed, as there was no observed improvement in his condition despite undergoing all recommended investigations and adhering to prescribed treatments.

2.3. Follow-Up and Review

During a follow-up visit two weeks after discharge, biopsy samples were obtained, and the results were negative for malignancy. However, the patient did not experience any symptom relief, despite compliance with oral voriconazole. Consequently, he requested further management abroad because he did not notice any improvement in his symptoms. Approximately one year after discharge, during a follow-up phone call with the patient's son, it was reported that the pa-

tient had undergone surgical treatment in the United States. It was reported that the patient had some significant improvement in symptoms and was scheduled for a second surgical procedure.

3. Discussion

Skull base osteomyelitis, a rare complication of malignant otitis externa, is a serious infection that affects the temporal, sphenoid, and occipital bone [1] [5] [11] [13]. Malignant otitis externa, as described by James Chandler in 1968 [14], primarily manifests in elderly diabetic and immunocompromised patients [1] [4] [5]. While often stemming from untreated or persistent otitis externa [15] [16], other routes of infection include rhinogenic, sinogenic, and odontogenic. *Pseudomonas aeruginosa* is the most frequently implicated pathogen, accounting for 50% - 90% of cases, whereas fungal infections, particularly *Aspergillus fumigatus*, are occasionally observed [13]. Notably, *Pseudomonas aeruginosa* typically lacks the ability to cause infection in immunocompetent individuals; however, in the presence of immunocompromised and iatrogenic trauma resulting from ear manipulations, it can cause infection [17]. The symptoms of skull base osteomyelitis vary and may initially present as seemingly benign, such as headaches, which can progress to cranial nerve palsies as the disease progresses. Severe otalgia is disproportionate to clinical signs of external ear infection and is a common complaint among patients [18].

In the case of our patient, although the classic symptoms of otalgia and otorrhea were absent at the time of presentation, he had experienced such symptoms four months prior and was diagnosed with an ear infection. Antibiotics were prescribed and subsequent debridement surgery was performed. The non-specific nature of symptoms, coupled with the absence of pronounced inflammatory signs, especially in fungal etiologies, poses challenges in the diagnosis of skull base osteomyelitis [16]. Therefore, a high index of suspicion is crucial, particularly for patients with risk factors.

Imaging plays a pivotal role in the diagnosis and monitoring of treatment response in skull base osteomyelitis [16]. Patients, particularly the elderly and those with diabetes, presenting with headaches, cranial nerve palsies, and radiological evidence of bony erosion, should be considered as potential cases, even in the absence of an obvious infective source [4]. Our patient fulfilled all the aforementioned criteria and had an identifiable source of ear infection. The diagnosis in our patient was made one month after discharge, approximately five months after symptom onset. This was performed after a detailed review of his investigations, coupled with the biopsy results and a second analysis of his initial imaging scan results. MRI demonstrated extensive bilateral base of the skull T1-post contrast enhancement, indicating a bilateral spreading pattern. Dural enhancement was also observed (Figure 1). CT also revealed cortical destruction of the petrosal apex with medial extension and cortical irregularities of the clivus (Figure 2).

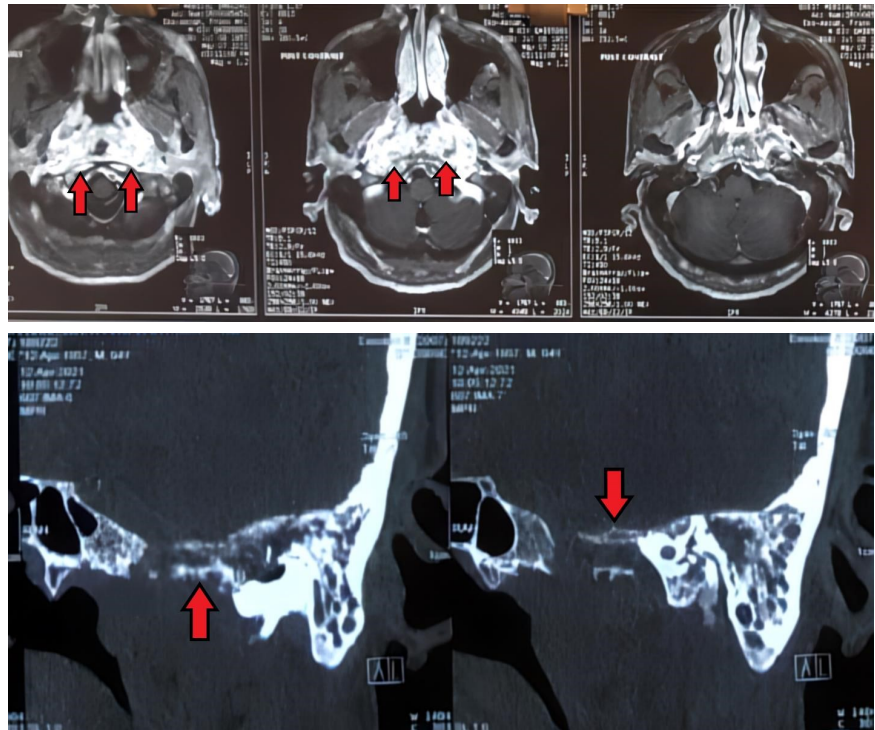


Figure 1. MRI—Extensive bilateral base of skull T1-post contrast enhancement, indicating a bilateral spreading pattern with dural enhancement.



Figure 2. CT scan—Cortical destruction of the petrosal apex with medial extension and cortical irregularities of the clivus.

The delay in diagnosis is not unique to our case, but is consistent with previous studies that reported an average time to diagnosis of approximately six months for this potentially life-threatening condition [5].

The primary limitations of this study were the delay in obtaining investigation results and the unavailability of surgical notes from the initial procedure conducted at the peripheral facility where the patient was initially referred.

4. Conclusion

Skull base osteomyelitis is a challenging condition to diagnose because of its

nonspecific symptoms and lack of pronounced inflammatory signs. Maintaining a high index of suspicion, particularly in patients with risk factors, is crucial for an early diagnosis. Imaging, including computerized tomography (CT) and magnetic resonance imaging (MRI), plays a vital role in the diagnosis and monitoring of treatment response. Delays in diagnosis are common, highlighting the need for increased awareness and timely intervention for this potentially life-threatening condition.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent form is available for review by the Editor-in-Chief of this journal upon request.

Acknowledgements

We would like to express our sincere gratitude to Dr. Lieke van der Meer of Maastricht University, Amsterdam, for her invaluable assistance in analyzing the images, which played a crucial role in the accurate diagnosis of our patient.

Data Availability Statement

Data sharing is not applicable to this article, as no datasets were generated or analyzed during the current study.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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Abbreviations

SBO = skull base osteomyelitis
MOE = malignant otitis externa
CN = cranial nerve
CT = computerized tomography
MRI = magnetic resonance imaging
PEG = percutaneous endoscopic gastrostomy
CRP = C-reactive protein
ACE = angiotensin converting enzyme
ESR = erythrocytes sedimentation rate

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