

A rare case report on unilateral cranial nerve palsy (9th and 10th) with skull base osteomyelitis

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ABSTRACT

Skull base osteomyelitis (SBO) is a rare disease affecting predominantly diabetic and immunocompromised adults and has a high mortality rate of about 10–20%. Here, we describe the case of SBO with progressive cranial nerve palsy involving 9th and 10th cranial nerves in a 30-year-old female patient who presented with a history of dysphagia and nasal regurgitation of food. The patient had no weakness and ataxia but over the following weeks, she developed 9th and 10th cranial nerve palsy. Magnetic resonance imaging brain report shows SBO and the patient was started on i.v. antibiotics. After which, the patient responded to treatment. SBO is a life-threatening condition and prolonged administration of antibiotics is a valuable option among these patients.

Key words: Cranial nerve palsy, Dysphagia, Skull base osteomyelitis

Skull base osteomyelitis (SBO) is a rare disease with a high mortality rate of about 10–20% [1]. It was first described in 1959 [2]. It affects predominantly diabetics and immunocompromised patients. Typical cases arise as a complication of malignant otitis externa with temporal bone involvement. However, there are atypical cases of SBO that affects the sphenoid or occipital bone that is not associated with malignant otitis externa. It can be associated with meningitis, cerebritis, or abscess formation. Early treatment is the key to success. Empirical treatment should be based on anti-pseudomonas agents. We describe the case of SBO with soft tissue and bone involvement and complicated by progressive cranial nerve palsy.

CASE REPORT

A 30-year-old female admitted to the hospital with a 20 days history of dysphagia to both solids and liquids with nasal regurgitation of food on attempted swallowing and dysphonia. Her medical history reveals no diseases in the past or any comorbidities.

On physical examination, her vitals were normal and the patient was afebrile throughout the period. Neurological examination revealed the absence of gag reflex and loss of arching of the palate on the left side with uvula deviated toward the right side. There was no evidence of other cranial nerve involvement and peripheral neurological examination was normal. Throat examination revealed left vocal cord palsy.

Her blood investigation revealed a total leukocyte count of 11,200/mm³ with 75% neutrophils and 20% lymphocytes. C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) were elevated. The chest X-ray was normal. Magnetic resonance imaging (MRI) brain showed diffuse edema and post-contrast enhancement involving the posterior pharyngeal wall which was more on the left side including left lateral nasopharyngeal wall, left side of clivus, left occipital condyle, left petrous apex, and edema involving hypoglossal and jugular foramen. Based on the MRI findings, a final diagnosis of SBO was made.


The patient was started on i.v. ceftriaxone 1 g 12th hourly, i.v. vancomycin 1 g 8th hourly, and other supportive treatment. The neurosurgeon did not recommend any active intervention and advised to continue i.v. antibiotics. The patient's condition improved over 12 days, dysphagia, and dysphonia reduced. The patient was referred to SGPGI Lucknow on the 14th day. The patient's contrast-enhanced computed tomography scan of the paranasal sinuses was done after 2 weeks which revealed small peripherally enhancing collection, the largest of 24 mm × 10 mm involving the posterior pharyngeal and parapharyngeal spaces, nasopharynx, and oropharynx suggestive of parapharyngeal abscess. The patient was switched to oral linezolid and cefuroxime for 2 weeks and then changed to voriconazole for 1 month. The patient was found asymptomatic after a follow-up of 1 month.

DISCUSSION

We describe the case of SBO affecting the skull base with multiple and progressive cranial nerve palsies. The VII cranial nerve is

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most commonly affected, whereas, in our patient, the VII cranial nerve was not involved. A meta-analysis conducted by Ridder *et al.* found that 59% of patients were affected by cranial nerve palsy (that persisted after therapy in 41%) [3]. There is some controversy related to the terminology as some authors refer SBO as the entity resulting from complications of malignant external otitis [4-7]; whereas, others have included atypical or central SBO also in the definition. The atypical or central SBO affects sphenoid and/or occipital bones and it may be secondary to an otogenic, sinus, odontogenic, or hematogenous infection, although there are also cases with no obvious infective source [3,8-10]. Some authors prefer to avoid the term SBO and define it as “a lateral temporal bone and medial temporal bone osteomyelitis” [11].

The main causative organism of SBO is *Pseudomonas aeruginosa*; although, other bacteria such as *Staphylococcus aureus*, *Klebsiella* spp., *Proteus* spp., and *Mycobacterium chelonae* have also been reported [1,3]. Apart from bacteria, some fungal species such as *Candida*, *Aspergillus*, *Rhizopus*, and *Mucor* have also been involved in SBO [3]. Hence, a possibility of fungal infection should always be kept in mind among patients with underlying chronic sinusitis, sinus pain, facial/periorbital swelling, or in the absence of ear discharge [8]. A review conducted by Blyth *et al.* found that *Zygomycetes* contributes to more than 50% of fungal SBO, and as a result, amphotericin B was suggested as the preferred empirical treatment when fungal SBO is suspected [8].

An exact and early diagnosis of the entity is very important which can be achieved with the help of the latest diagnostic techniques. Usually, CRP and ESR are used as a diagnostic adjunct among patients with SBO. CT and MRI are used to evaluate the degree of bone and soft-tissue involvement. The use of imaging tests has a role in the diagnosis and follow-up of these patients as we can measure the activity of infection by close radiological and clinical follow-up of SBO. The follow-up of these patients can be achieved with the help of gallium scans that have been proposed at 1 week and 12 weeks after antibiotics cessation to search for recurrence [12].

The single-photon emission computed tomography (SPECT) scan has also shown good predictive value for long-term outcomes [6]. The use of SPECT-CT with ⁶⁷Ga or ¹¹¹In-labeled leukocytes has shown higher specificity than scintigraphy, CT scan, or SPECT alone [13]. In a recent study by Sharma *et al.*, SPECT-CT was the most sensitive technique for SBO diagnosis with sensitivity and specificity of 100% while CT was the most specific technique with a sensitivity of 73% and specificity of 100% [14]. Filippi and Schillaci found that the specificity of SPECT-CT was higher than SPECT alone (89% vs. 78%) [15]. Obtaining tissue specimens are paramount to determine the microbial etiology but it needs proper microbiological and histopathological processing of samples.

The differential diagnosis of soft-tissue masses at the skull base is wide and comprise malignant lesions (nasopharyngeal carcinoma, squamous cell carcinoma of the external auditory canal, schwannoma, metastases, and multiple myeloma), infectious and inflammatory diseases (SBO, inflammatory pseudotumor,

granulomatosis with polyangiitis, tuberculosis, and sarcoidosis), and others, such as cholesteatoma, dural arteriovenous fistula, fibrous dysplasia, and Paget disease [1,10,16,17].

The treatment of SBO includes broad-spectrum antibiotics along with antifungal therapy that has good anti-*Pseudomonas* activity [4]. Initial intravenous (i.v.) treatment is usually followed by long-term oral therapy. Antibiotic regimens comprise an initial 4–6 weeks course of i.v. treatment followed by 6–12 months of oral medication [9]. Since *P. aeruginosa* osteomyelitis has been associated with lower cure rates and higher recurrence risk [18], antimicrobial therapy should be continued for several weeks to minimize relapses [9,19]. In a meta-analysis by Ridder *et al.*, the median length of inpatient treatment was 32 days (range: 7–83), while the median length of subsequent oral treatment was 1.5 months (range 0–16 months) [3].

Continuous infusion of antibiotics allows outpatient administration of parenteral therapy, thereby reducing health-care resources consumption [20]. Administration of beta-lactams by continuous i.v. infusion generates higher blood and tissue concentrations that allow longer time above the minimum inhibitory concentration (MIC) as compared with intermittent dosing, which is particularly useful for bacteria with high MIC values such as *P. aeruginosa* [21]. The role of hyperbaric oxygen is not well-defined but may provide some benefit by reversing the hypoxia, improving the phagocytic activity against aerobic microorganisms, and stimulating angiogenesis. A positive effect on the cranial nerve palsies recovery has been reported in various studies [9,12,22].

The role of surgical debridement remains controversial as it has no clear impact on survival, antimicrobial treatment duration, or prognosis [6,19,22], except in the case of fungal SBO [8]. Surgery may be needed for decompressing cranial nerves only in selected cases [11]. Surgical debridement could remove devitalized tissue and reduce infection load to improve the penetration of antibiotics. A few authors also advise early and forceful surgical approaches, especially in patients with protracted ear infections and signs of cranial nerve involvement [8].

CONCLUSION

In the present case, the patient does not have any fever, any toxic symptoms, or osteomyelitis and responded well to antibiotics. Outpatient empirical parenteral antibiotic therapy administered by continuous infusion could be a suitable option to prolong intravenous treatment in this severe disease, especially when difficult-to-treat organisms like *P. aeruginosa* are suspected. Clinical suspicion, even in the absence of an obvious infective source, and microbiological processing of samples are the key to achieve a correct and timely diagnosis.

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