Case Report

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Central Skull Base Osteomyelitis with Ischemic Infarct: A Case Report and Literature Review

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Abstract

Skull base osteomyelitis (SBO) defined as an inflammation of bony structures of the cranial base, is rare condition with substantial morbidity and mortality. It typically involves temporal bone and manifested as otologic symptom and cranial neuropathy. Central skull base osteomyelitis (CSBO) is atypical form of SBO that involves the sphenoid and clivus and related to non-otogenic conditions. We present an unusual case of CSBO presented with ischemic stroke and multiple cranial nerve involvement. A diabetic women of 45 years old, with a previous history of right otalgia without otorrhea and severe headache, admitted with sudden left hemiplegia and controlateral multiple cranial nerve (CN) palsies. Cerebral imaging investigations (CT and MRI scans) showed ischemic infraction secondary to the occlusion of the right internal carotid artery (ICA) and radiological evidence of invasive osteomyelitis on the right side of the central skull base with contiguous lateral sinus thrombosis. Broad spectrum antibiotics and anticoagulant therapy was initiated with unfavorable outcome. Based on thorough review of the literature, arterial cerebrovascular complications revealing CSBO are extremely rare and the diagnosis is often challenging for the clinician.

Keywords: Skull base osteomyelitis; Central skull base osteomyelitis; Stroke; Cranial neuropathies

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Introduction

Skull base osteomyelitis (SBO) is a rare, life-threatening infection of the bones and soft tissues at the base of the skull. It predominantly occurs in elderly, diabetic, and/or immunocompromised patients. The diagnostic is often delayed given rise to potential serious complications [1]. Pseudomonas aeruginosa is the most common pathogen implicated, although there have been reports of other micro-organisms such as Aspergillus, Gram positive organisms, Mycobacterium and Candida [2,3]. SBO can present as either typical or atypical form. The typical SBO involve the temporal bone and is usually initiated by malignant otitis external and adjacent soft-tissues infection. Patient frequently complains of severe and profound otalgia with purulent otorrhea complicated by multiple cranial nerve palsies: The facial nerve is the most common and first one involved followed by the lower cranial nerves (IX, X, IX, XII) [4]. In the other hand, the atypical form, as in our case, also called central skull base osteomyelitis is characterized by the involvement of sphenoid and occipital bones and the clivus. Hence, it can emerge from contiguous paranasal infection, such as sphenoidal or ethmoidal sinusitis, without associated external otitis. It often presents with non-specific symptoms such as headache, facial pain and cervicalgia that may evolve to more severe clinical complications depending on the local spread of the infection/inflammation [5]. Given the critical boundaries of the central skull base, serious cerebrovascular complications might arise. Although, thrombosis of transverse-sigmoid sinus, cavernous sinus and the internal jugular vein is thought to be one of the typical complications of skull base osteomyelitis, arterial cerebral infracts have been exceptionally reported [6-9]. We present an uncommon case of CBOS revealed by ischemic stroke and we have undertaken through review of the literature.

Case Report

A 45-year-old woman was admitted to the emergency department for sudden left hemiplegia, difficulty of speech and swallowing. She had a 27-year history of diabetes mellitus with poor metabolic control and micro vascular complications (retinopathy and diabetic nephropathy without renal failure). In the past four months, she had complained of right otalgia and neck pain. She never had external otitis or otorrhea. Several weeks later, she develops severe headache and vomiting, an episode of mild right epistaxis, visual disturbances and facial deviation. On examination, she was febrile and her neck was blocked rightward. She has a total left hemiplegia and multiple right cranial nerve palsies: facial nerve, complete ophtalmoplegia, and motor trigeminal nerve with trismus (which limited the clinical evaluation of the lower cranial nerves). The first cerebral CT scan showed right infraction with right lateral sinus thrombosis **(Figure 1)** and retropharyngeal abcess. Laboratory data indicated poor glycemic control (HbA1c 8, 1%) and high levels of inflammation markers with elevated white blood cell (WBC) counts (21,680 /μL), CRP levels (125 mg/dL, normal <0.5 mg/dl), and sedimentation rate (134 mm/1st hour). The diffusion-weighted image (DWI) indicated a sub-acute infraction in the right middle cerebral artery (MCA) territory with right internal carotid occlusion and thrombosis of the right transverse-sigmoid at angiographic sequences **(Figure 2)**. Moreover, T2 weighted and FATSAT MRI demonstrates a large right mass lesion of the petrous apex bone that expanded to the clivus, the retropharyngeal space and soft tissues. This process invaded the carotid artery and the jugular vein which are not visualized after contrast injection **(Figure 3)**. CT scan revealed erosion of the skull base bone with destruction of clivus but not of the bony wall of the external ear canal. Opacification of the right mastoid air cells was also noted **(Figure 3)**. An otolaryngeal examination identified a bulging posterior pharyngeal wall and a pharynx biopsy was negative. No culture was obtained at that time. The patient was treated by high dose of empirical broad spectrum intravenous antibiotic (ceftriaxone and metronidazole), but unfortunately showed a deterioration in her mental status culminated in her death on day 7.

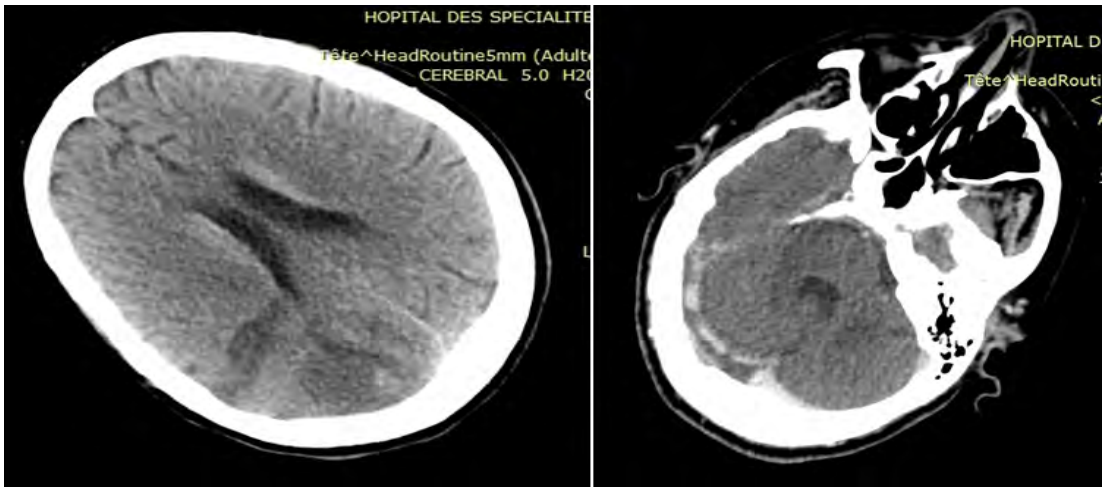

Figure 1 CT scan showing right cerebral infraction and after injection right lateral sinus thrombosis.

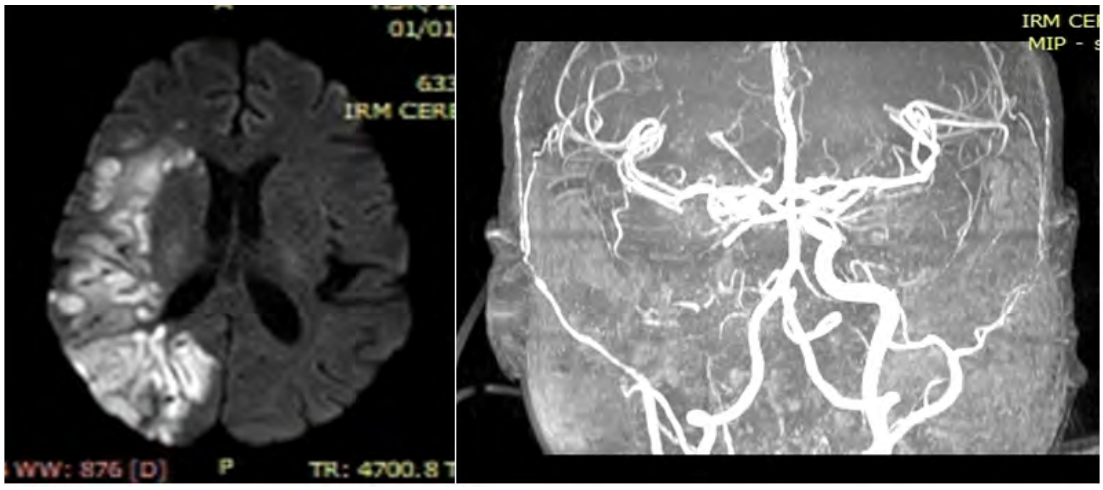

Figure 2 **(a)** The diffusion-weighted image (DWI) showing infraction in the right middle cerebral artery (MCA) territory and **(b)** right internal carotid occlusion at angiographic sequences.

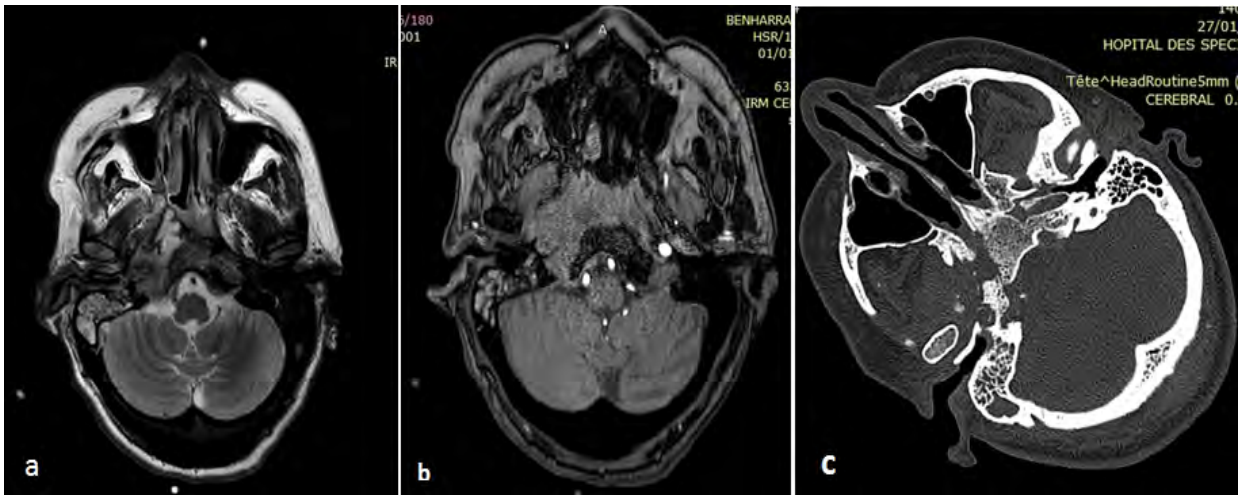

Figure 3 **(a)** MRI T2 weighted revealed a large right mass lesion of the petrous apex bone that expanded to the clivus, the retropharyngeal space and soft tissues. **(b)** FATSAT with contrast: This mass invaded the carotid artery and the jugular veins which are not visualized after contrast injection compared to the contra lateral side. **(c)** CT showed erosion of the skull base bone with destruction of clivus but not of the bony wall of the external ear canal. Opacification of the right mastoid air cells was also noted.

Discussion

In the era of antibiotics, SBO became a rare event but is still potentially fatal condition if not promptly recognized and properly treated. CSBO, also called atypical SBO, is less frequent and distinct from the typical form [1,3]. Similar to the classical form, CSBO typically afflicts elderly patients with immunocompromised conditions such as diabetes mellitus, corticosteroid use or HIV infection [1]. Our patient had uncontrolled diabetes with microangiopathic complications (diabetic retinopathy and nephropathy). Pathologically, CSBO is characterized by sphenoid bone osteomyelitis (classically the clivus) as a result of a direct spread, via the Haversian system, of an infectious process involving the paranasal sinuses, external auditory canal, mastoid, middle ear, or oral cavity [10].

The clinical diagnosis is often difficult due to lack of specific symptoms. Headaches is the most frequent symptom and might be the only one [11,12]. Fever is relatively uncommon on presentation in the literature [13]. Other presenting features may include otalgia, atypical facial pain or painful neck stiffness, such our patient [5,13]. CN neuropathies occur later as the infection extends to the soft tissues surrounding the basal skull foramina. Contrary to the typical SBO, which primary affects the seventh nerve, CSBO seems to impact the more centrally located CN: III, V, VI, and the lower CNs (IX, X, XI, XII). This particular pattern indicates a clival pathology [5,6,13]. Chang *et al.* have reported six cases of atypical SBO that all have cranial neuropathy at the time of presentation with CN VI in five patients and CN IX, X et XII palsy in three. Moreover, a rare case of Collet Sicard syndrome have been described (palsy of cranial nerves IX, X, XI, and XII) as an ultimate complication of SCB originating from otitis media. [14]. The diagnosis delay may lead to serious complications including meningitis, abscess formation. Cerebrovascular complications of SBO are predominantly represented by sinovenous thrombosis, such as thrombosis of sigmoid sinus, cavernous sinus and internal jugular vein.

Arterial complications of SBO are less common and are usually found as asymptomatic radiological abnormalities [15,16]. A review of the literature reveals very few reports of cerebral infractions secondary to SBO. Accordingly, the internal carotid artery (ICA) is the more often involved artery, especially in its petrous and cavernous portion but also in its extracranial segment if a retropharyngeal abcess is associated. Stenosis of the ICA arise as the leading mechanism of cerebrovascular compromise [6,15,16]. From a large observational study of 41 cases of SBO, cerebral infraction, due to arteritis, occurred in 14 patients [17]. Interestingly, Miyabe *et al.* have described a case of CSBO that emerge with cerebral infarctions and radiological finding consistent with complete occlusion of the right ICA. They suggest that ICA stenosis result from contiguous inflammatory arteritis with artery to artery emboli. Our case seems to share similar pathogenesis. Others underlying mechanism of the ICA stenosis are mass effect or vasospasm related to the surrounding inflammatory process [7,15].

Moreover, pseudoanvrysms or anevrysms formation of the ICA represent another large artery complication of SBO that is limited to few case reports. They are more likely to be mycotic anevrysm and occur in the intrapetrous portion of the ICA [18-21]. A case of ICA blowout secondary to mycotic anevrysm, presented with massif epistaxis has been reported as an extremely rare and fatal complication [22].

Imaging plays an important role in diagnosing SBO. Various techniques have been studied in the literature, including contrast-enhanced CT, MRI and scintigraphy with Technetium-99m or gallium-67. MRI is the most accurate imaging modality owing to its superiority to evaluate the exact anatomical location and the full extent of the soft tissue abnormalities [10]. Given the potential

vascular involvement in SBO and its serious consequences, CT and MR angiography are important to assess arterial and venous vasculature, especially in the presence of cerebral infraction [23].

When imaging findings illustrates a destructive or infiltrative process in the central skull base, nasopharyngeal carcinoma, metastasis, and inflammatory diseases need to be considered. Hence, tissue biopsy is ultimately often indicated not only to exclude nasopharyngeal carcinoma but also for culture and sensitivity testing of the causative pathogens [12].

Broad spectrum antibiotics should be rapidly initiated which should cover P. aeruginosa. Possibles options include systemic quinolones (ciprofloxacin), ceftriaxone, piperacillin- tazobactam, meropenem or clindamycin. Recent data demonstrate carbapenems working synergistically with ciprofloxacin as a suitable adjunctive therapy. Also, antifungal therapy should be considered [1,5].

Conclusion

Vascular involvement is not surprising radiological finding in SCBO. However, subsequent ischemic stroke is an extremely rare presentation at the time of diagnosis. Our patient illustrates a case of undiagnosed CSBO presented with cerebral infraction due to critical ICA stenosis. As SCBO commonly present with non-specific symptoms, we emphasize the need of prompt investigations and treatment keeping in mind the screening of intracranial and extracranial vascular involvement especially in diabetic patients.

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