Radiology Case Reports 15 (2020) 1512–1517

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Case Report

Central skull base osteomyelitis manifesting with a preclival mass and internal carotid artery mycotic aneurysm ☆,☆☆,★

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ARTICLE INFO

Article history:
Received 13 May 2020
Revised 28 May 2020
Accepted 29 May 2020

Keywords:
Skull
Base
Central
Osteomyelitis
Clivus

ABSTRACT

Central skull base osteomyelitis is a rare entity that can demonstrate confounding radiologic, clinical, and laboratory data leading to a delay in diagnosis. The morbidity and mortality for skull base osteomyelitis are both high, thus a rapid diagnosis is required for appropriate treatment. In this case report, we discuss a 68-year-old male who presented with acute left facial nerve paralysis in the setting of chronic headache and left mucoid middle ear effusion. Radiologic evaluation revealed abnormal hypointense marrow of the central skull base on T1 weighted magnetic resonance imaging, preclival mass-like tissue, and short segment luminal narrowing of the left cervical ICA with mycotic aneurysm formation.

Extensive workup via a multidisciplinary approach, including neurology, otolaryngology, neurosurgery and radiology led to a diagnosis of central skull base osteomyelitis. A familiarity of this disease process is important for the radiologist in order to facilitate appropriate patient referral and treatment. This case emphasizes the importance of considering this diagnosis in the setting of headache, cranial neuropathy, and abnormal skull base imaging with adjacent preclival soft tissue mass.

Introduction

Central skull base osteomyelitis is an uncommon diagnosis which is defined by the involvement of sphenoid bone, occipital bone and the clivus, and is distinct from osteomyelitis arising from malignant otitis externa in the adjacent temporal bone [1]. Paraclival soft tissue masses can occur with central skull base osteomyelitis which can be a confounding finding requiring biopsy for histopathologic confirmation [2]. An

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☆☆ The investigators have adhered to the policies for protection of human subjects as prescribed in 45 CFR 46.
★ Competing Interests: The authors have no special interests to declare.
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https://doi.org/10.1016/j.radcr.2020.05.069

accurate and rapid diagnosis is vital in order to obtain a multidisciplinary treatment regimen for these patients. We present a case of central skull base osteomyelitis manifesting with a preclival soft tissue mass and cervical ICA mycotic aneurysm.

Case report

A 68-year-old male with a past medical history notable for poorly controlled type two diabetes mellitus, hypertension and hyperlipidemia presented to the emergency department with acute onset left-sided facial droop and changes in taste. Upon further query, the patient endorsed a low-grade chronic headache for several months, for which he had not sought medical care. Additionally, he had a chronic left mucoid middle ear effusion which underwent myringotomy and fluid sampling approximately 6 weeks prior to presentation which revealed a thick, clear fluid. Cultures returned negative for the sample at that time.

In the emergency room, his vital signs demonstrated mild hypertension with a blood pressure of 169/98, without tachycardia, fever, or hypoxia. On physical exam, note was made of complete left sided facial asymmetry at rest which persisted with eyebrow elevation, eye closure and grimace. No other neurologic deficits were noted on physical exam. Additionally, there had been re-accumulation of a left sided mucoid middle ear effusion. Initial laboratory markers were notable for a normal complete blood count, glucose elevated to 249, CRP elevated to 1.40 and erythrocyte sedimentation rate (ESR) elevated to 112.

Initial radiologic evaluation included a noncontrast CT head and a CT angiogram of the head and neck. The unenhanced CT demonstrated ground glass mineralization of the central skull base with subtle erosions of the bilateral, left greater than right, sphenoid bones (Fig. 1) as well as left middle ear and mastoid effusions which were stable to prior comparison examinations (Fig. 2). The follow on CT angiogram showed focal narrowing of the left distal cervical internal carotid artery with an indistinct, preclival soft tissue mass (Fig. 3).

The patient was admitted for expedited workup of his acute onset, atypical Bell's palsy. ENT performed a left myringotomy which obtained mucopurulent fluid. Given the purulent middle ear effusion and elevated inflammatory markers the patient was started on broad spectrum intravenous antibiotics, Unasyn and Vancomycin, as well as Ciprofloxacin ear drops.

The following morning, a magnetic resonance imaging (MRI) of the skull base and magnetic resonance angiogram (MRA) of the neck was obtained which demonstrated abnormal T1 hypointense marrow of the central skull base and enhancing preclival mass-like soft tissue with associated restricted diffusion on diffusion weighted imaging (Figs. 4-6). The MRA confirmed narrowing of the distal cervical and petrous segments of the left ICA and more clearly depicted an eccentric outpouching consistent with a pseudoaneurysm (Figs. 7a and b).

Given the constellation of these clinical and radiologic findings a diagnosis of central skull base osteomyelitis with adjacent inflammatory mass was considered. The patient was subsequently transferred to an outside facility to be evaluated by a subspecialist skull base otolaryngologist. Biopsy was subsequently performed of the preclival soft tissue mass which yielded polymicrobial growth of *Pseudomonas aeruginosa, Staphylococcus epidermidis, and Cutibacterium* (formerly Propionibacterium) with no evidence of malignancy. A diagnosis of polymicrobial osteomyelitis of the skull base was made and the patient was started on a 12-week regimen of broad spectrum intravenous antibiotics. Endovascular and surgical management of the patient's ICA mycotic aneurysm are to be deferred until after completion of antibiotic therapy given the aneurysm's location at the craniocervical junction and adjacent active inflammation.

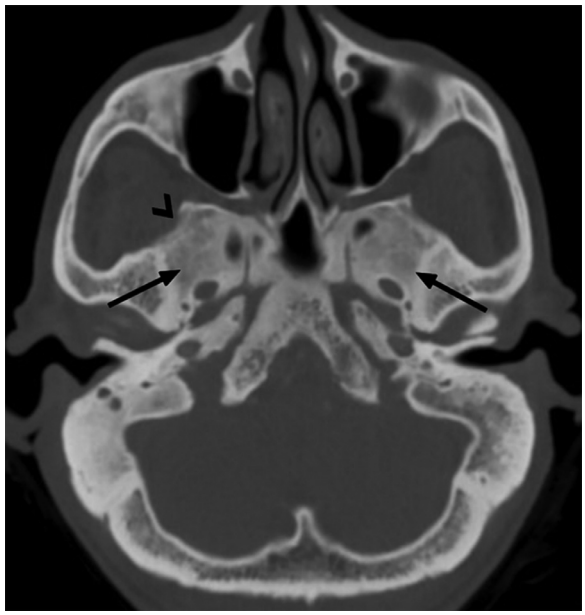

Fig. 1 – Noncontrast axial head CT, bone window demonstrates groundglass mineralization of the central skull base (arrows) with subtle cortical irregularity (arrowhead) suggesting an underlying inflammatory or proliferative process.

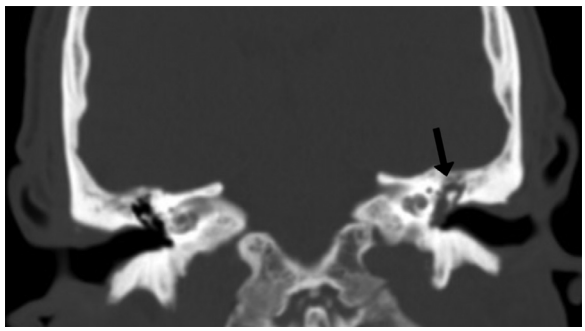

Fig. 2 – Noncontrast coronal head CT, in a bone window, demonstrates a left middle ear effusion (black arrows). ENT performed a left myringotomy which returned mucopurulent fluid; cultures were negative.

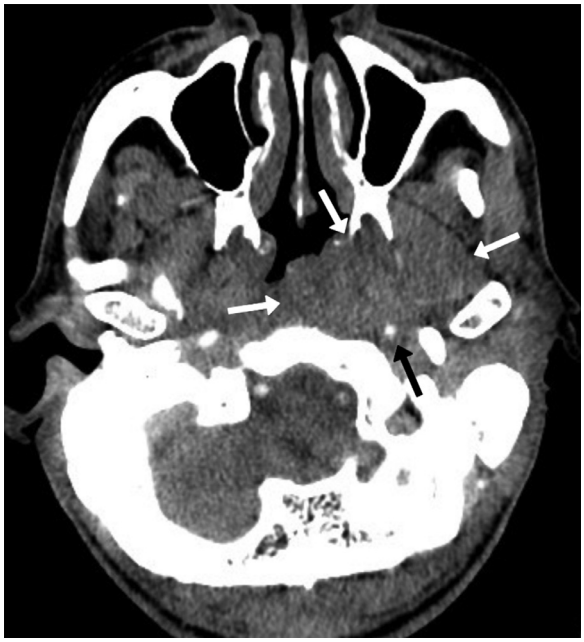

Fig. 3 – Axial CT Angiogram, demonstrates a left preclival soft tissue mass (white arrows) with mild effacement of the nasopharynx. There is also narrowing and irregularity of the adjacent distal cervical ICA (black arrow).

Discussion

Central skull base osteomyelitis is a rare pathological process that, if left untreated, leads to significant morbidity and mortality. Osteomyelitis is characterized by progressive inflammation and destruction of bone which may extend into the adjacent soft tissues [3]. Skull base osteomyelitis can be subdivided into three distinct groups, skull base osteomyelitis, central or atypical skull base osteomyelitis and pediatric clival osteomyelitis [4]. Classic skull base osteomyelitis is often associated with malignant otitis externa, also known as necrotizing otitis externa, and classically involves the temporal bone. Central skull base osteomyelitis is considered a distinct entity due its location, involving the sphenoid bone, occipital bone, as well as the clivus, in addition to absence of malignant otitis externa [1].

Central skull base osteomyelitis is most often diagnosed in patients with underlying comorbidities, such as diabetes mellitus, immunocompromised status or chronic sinonasal inflammation [2]. Bacterial and fungal pathogens have been described in the literature with Pseudomonas aeruginosa as the classic, most common bacterial source. Additional less common bacterial species include Staphylococcus spp, Salmonella spp, Proteus spp, nontuberculous Mycobacterium spp, Treponema pallidum, and Klebsiella spp. The most common fungal organism implicated in skull base osteomyelitis is Aspergillus spp, although Candida spp, Cryptococcus neoformans, Blastomycosis, and Mucormycosis have also been documented in the literature [4].

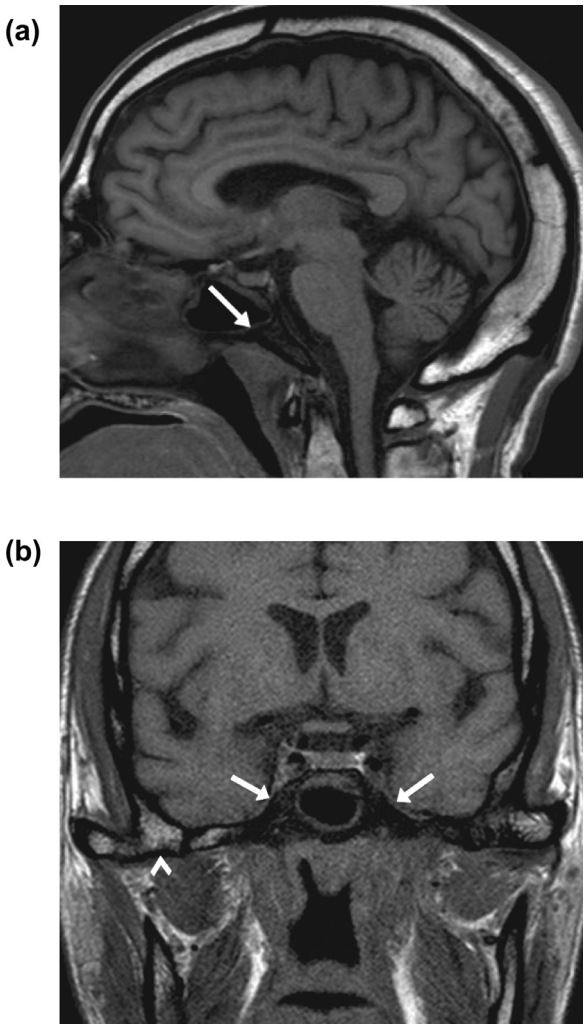

Fig. 4 – a. MRI of the skull base was performed on day 1 of admission. Sagittal MRI T1-weighted noncontrast image clearly demonstrates abnormal clival marrow signal hypointensity (white arrow). b. Coronal MRI T1-weighted noncontrast image shows abnormal hypointense marrow in the central skull base and clivus (white arrows). Note the normal marrow signal within the uninvolved bone (arrowhead).

Patients can present with a broad range of signs and symptoms secondary to the innate and complex anatomy of the central skull base. In the literature, reported initial symptoms have included headaches, atypical facial pain and various cranial neuropathies [5,6].

The laboratory analysis is an important factor to consider in the early workup of these patients. In the context of an aggressive underlying osteomyelitis, inflammatory markers, for example, acute phase reactants, are expected to be elevated and oftentimes, the patient will present with elevated inflammatory markers, leukocytosis and fever, thus raising the clinical suspicion for infection [4]. However, up to 25% of patients with central skull base osteomyelitis may present without an elevated ESR, leukocytosis, fever or other clinical signs suggestive of infection [7].

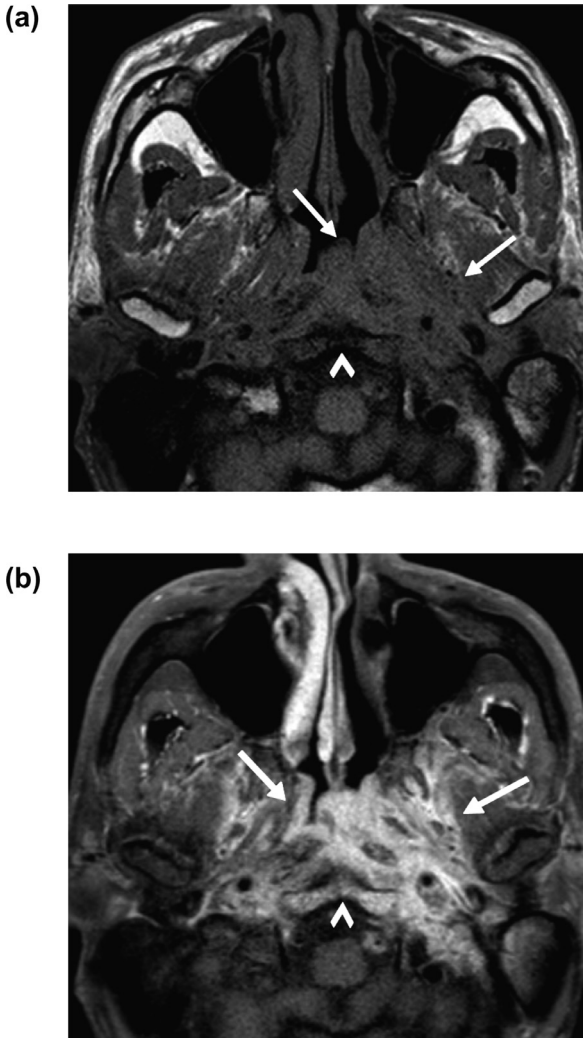

Fig. 5 – a. Axial MRI TI-weighted noncontrast image demonstrates abnormal soft tissue, isointense to muscle infiltrating submucosally into the nasopharyngeal tissues and surrounding the left cervical ICA (white arrows). T1 hypointense marrow signal is again seen within the clivus (arrowhead). b. Contrast-enhanced axial T1 weighted image with fat saturation demonstrates marked enhancement within the infiltrative tissue (white arrows) and underlying clivus (arrowhead).

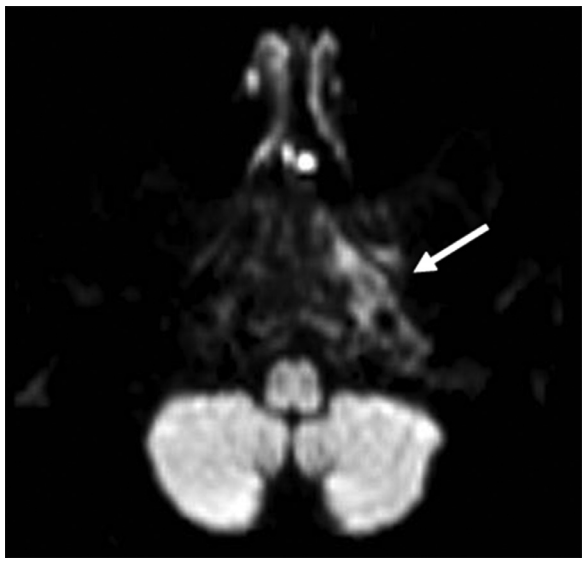

Fig. 6 – Axial diffusion weighted image demonstrates restricted diffusion within the mass-like tissue (white arrows) and clivus which can be seen with both inflammatory and neoplastic processes.

Radiology plays a critical role in the evaluation of these patients and is essential to the timely and accurate diagnosis of these patients in order to expedite treatment. CT is often the initial study and can show destructive bony changes and remodeling; however, it frequently underestimates the extent of disease and involvement of adjacent structures. Moreover, findings may be absent on CT and are often seen only in advanced cases [8] with no osseous erosion observed in 30%-37% of cases [7,9].

Contrast enhanced MRI is the study of choice for these patients as it provides superior soft tissue contrast and has the ability to assess the involvement of adjacent intra and extracranial structures such as the cranial nerves [10]. Notable positive findings on MRI include abnormal T1 hypointense marrow signal within the clivus and involved skull base, bone and soft tissue enhancement, and development of soft tissue masses within the preclival space, cavernous sinus and Meckel's cave [2]. Marrow T1 hypointensity is nonspecific and only implies the presence of underlying pathology, which encompasses a broad differential including marrow regeneration as well as infectious and neoplastic etiologies. Mass-like preclival soft tissue abnormalities are commonly reported in central skull base osteomyelitis, and is important to note as this can easily mimic malignancy [11]. When biopsies are obtained, specimens should be sent for cultures and gram staining, in addition to cytological evaluation in order to prevent delays in diagnosis [12].

Vascular complications, such as mycotic aneurysm formation, of the adjacent arterial structures have been reported in the literature [13–15]. This highlights the importance of scrutinizing the nearby vessels, and the threshold to obtain angiographic studies should therefore be low. Treatment options for infectious vascular complications include initial long-term antibiotic therapy with potential open versus endovascular intervention. If the pseudoaneurysm is amenable to endovascular intervention these may be treated with the use of detachable coils, balloons, glue, or thrombin [16].

Given its proximity to several important structures, an infection involving the central skull base can progress involving the cranial nerves, cavernous sinus, and dura leading to intracranial complications. Cranial neuropathy is often a presenting symptom, as previously mentioned and highlighted in our patient presenting with facial nerve paralysis [7]. Cranial nerve involvement is variable based on the origin and spread of the infection. In central skull base osteomyelitis, the central and posterior cranial nerve foramina are most commonly affected involving the VII-XII cranial nerves [7].

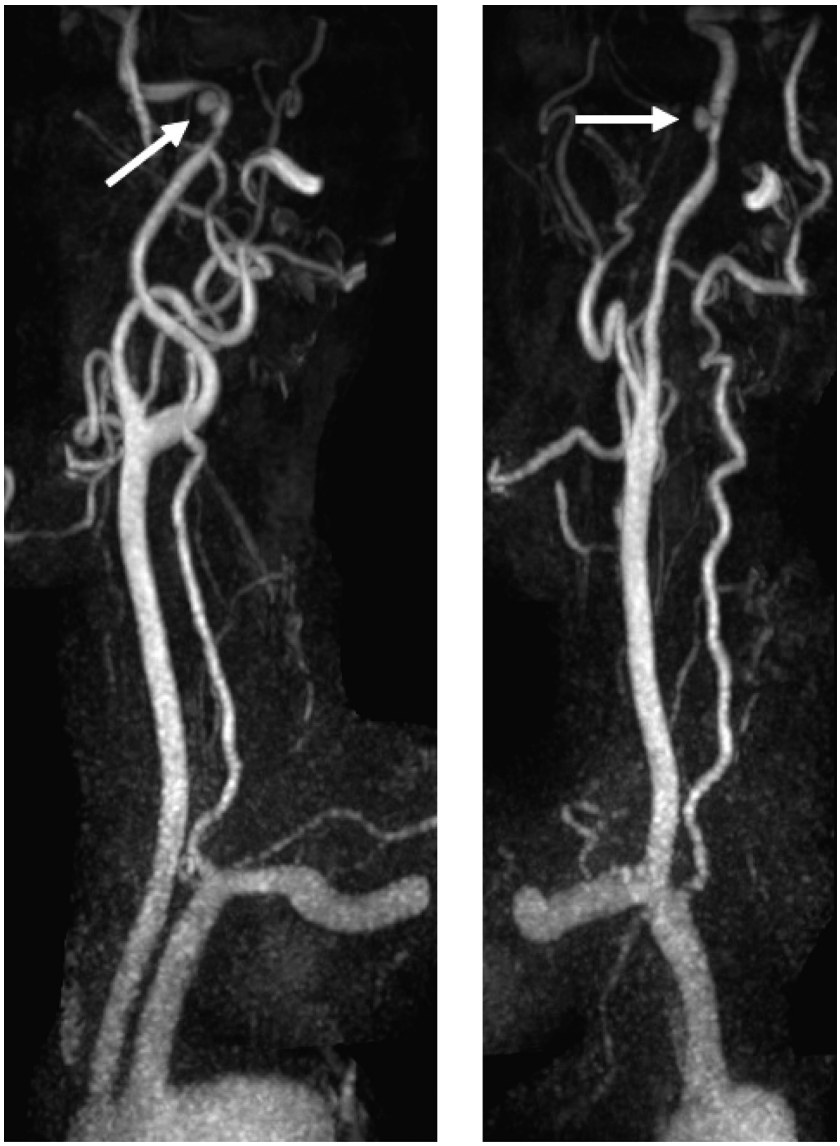

Fig. 7 – a. Maximum intensity projection (MIP) MRA of the left carotid circulation demonstrates short segment irregularity and narrowing of the left distal internal carotid artery at the level of the skull base, with an eccentric saccular aneurysm (white arrow). b. Oblique MIP MRA better delineates the eccentric saccular morphology typical of a mycotic aneurysm of the distal left internal carotid artery (white arrow).

Radiologic follow up of these patients is essential given the required long-term antimicrobial regimens. CT imaging is limited in evaluating the underlying osseous involvement and only shows reactive osseous change, which may resolve slowly, or never at all [9]. MRI is the preferred imaging modality for follow-up, as MRI more accurately evaluates the osseous and soft tissue involvement concurrently [17]. However, it is important to note that marrow signal abnormalities may persist up to six months after appropriate antimicrobial treatment [7]. Nuclear medicine studies, including indium-111 white blood cell and technetium-99m methylene diphosphonate bone single photon emission computed tomography have been shown to be beneficial adjunct studies with conventional CT and MRI in assessing response to treatment [8].

In patients presenting with headache, cranial neuropathy, and abnormal skull base imaging with preclival soft tissue mass, it is critical to include central skull base osteomyelitis within the differential diagnosis which may drastically improve patient morbidity and mortality through prompt diagnosis and treatment. This is of particular importance in patients presenting without classic signs of infection and potentially normal inflammatory lab markers so that biopsied specimens are sent for culture as well as cytological evaluation. Additionally, consideration for angiographic studies with careful scrutiny of the surrounding vasculature is recommended to

assess for secondary vascular pathology including vascular narrowing and mycotic aneurysm formation. Rapid diagnosis allows for timely administration of antimicrobial treatment with subsequent improvement in patient outcomes.

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