



Skull Base Osteomyelitis Presenting with Facial Paralysis, Low Cranial Nerve Palsies and Bilateral Carotid Involvement: A Case Report

Fasyal Paralizi, Alt Kranyal Sinir Felçleri ve İki Yanlı Karotis Tutulumu ile Kendini Gösteren Kafa Tabanı Osteomyeliti: Olgu Sunumu

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Summary

Skull base osteomyelitis (SBO) typically presents with severe otalgia and unilateral otorrhea in immune-compromised, particularly in elderly diabetic patients. Skull base osteomyelitis usually presents with external otitis but it can also occur as a complication of acute otitis media and mastoiditis. Complications of SBO are venous sinus thrombosis, meningitis, abscess, cranial neuropathies and carotid invasion with or without ischemic stroke. Here we report a case with SBO presenting with facial paralysis, lower cranial nerve palsies and bilateral carotid involvement which occurred following sore throat and bilateral otalgia. (Turkish Journal of Neurology 2015; 21:27-30)

Key Words: Skull base osteomyelitis, external otitis, pseudomonas aeruginosa, internal carotid artery

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Özet

Kafa tabanı osteomyeliti (KTO) tipik olarak immün sistemi baskılanmış kişilerde, özellikle yaşlı diyabetik hastalarda şiddetli otalji ve tek taraflı otore ile kendini gösterir. Genelde eksternal otitle birlikte ama akut otitis media ve mastoiditin komplikasyonu olarak da görülebilir. Kafa tabanı osteomyelitinin komplikasyonları arasında sinüs ven trombozu, menenjit, abse, kranyal nöropatiler, iskemik infarktların eşlik ettiği veya etmediği karotis invazyonu bulunmaktadır. Bu olgu sunumunda boğaz ve iki yanlı kulak ağrısı sonrası gelişen fasyal paralizi, alt kranyal sinir felçleri ve iki yanlı karotis tutulumu ile kendini gösteren bir kafa tabanı osteomyeliti olgusu bildirilmektedir. (Türk Nöroloji Dergisi 2015; 21:27-30)

Anahtar Kelimeler: Kafa tabanı osteomyeliti, eksternal otit, psödomonas aeruginosa, internal karotis arter

Çıkar Çatışması: Yazarlar bu makale ile ilgili olarak herhangi bir çıkar çatışması bildirmemiştir.

Introduction

Skull base osteomyelitis (SBO) is divided into two subtypes: Typical and atypical (central) (1). The typical form is also called "malignant external otitis". In immunocompromised people, especially the elderly diabetic people, it is seen as severe otalgia and unilateral otorrhea (2). The infection starts from the soft tissue on the outer ear canal and spreads onto temporal bone, occipital bone and skull base through Santorini fissure and tympanomastoid

line (3). Typical SBO, however, can also be seen secondary to bone erosion, thrombophlebitis and infection spreading to temporal bone and intracranial compartments through the hematogenous route in the absence of external otitis, in addition to being a complication of the previously mentioned acute otitis media and mastoiditis. Pseudomonas aeruginosa is the most common pathogen (4,5,6).

Atypical SBO involves clivus. Without external otitis, it is generally caused by paranasal infections such as sphenoidal

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or ethmoidal sinusitis (7). Infection, moves to the skull base through the compact bone's Haversian system, causing osteitis or osteomyelitis. *Aspergillus*, *Fusobacterium necrophorum*, *Pseudomonas*, *Salmonella*, *Streptococcus* and *Staphylococcus* strains are detected in atypical osteomyelitis (7,8).

Case

Sixty-two-year-old female patient came to us with the complaints of inability to close her right eye, left-sided mouth drooping, inability to swallow and speech difficulty, emerging after throat and ear pain that started 2 months ago. The patient had lost 20 kilos in the last month. It was learned that he was admitted with stroke diagnosis to another facility and received Coraspin treatment. Being diabetic for 7 years with hypertension, she had been using insulin, metformin and acetylsalicylic, and for the past two days cefdinir. It was learned that she had been using various antibiotics for the past 2 months. In her neurological examination, she was dysarthric with peripheral facial paralysis on the right (grade 5); she was unable to move her tongue and the tongue was atrophied; soft palate would not elevate during phonation; gag reflex was absent on both sides. Based on these findings, the patient who had right 7th, bilateral 9th, 10th and 12th cranial nerve paralysis was further evaluated for multiple cranial neuropathy etiology. In the contrast cranial magnetic resonance imaging (MRI) including diffusion-weighted imaging (DWI), there were heterogeneously hyperintense lesions in the clivus, bilateral petrous bones and frontal half of the occipital bone seen in T2 slices and hypointense and dense contrast-holding lesions seen in T1. Retropharyngeal muscle structures adjacent to the lesions had appearances congruent with necrosis (Figures 1a,b). The lesion areas were assessed as skull base osteomyelitis. In addition, there was T2 hyperintensities congruent with dense effusion in the mastoid cells, especially on the right. There was no signal at the level of right internal carotid artery (ICA) petrous and cavernous segments and the appearance was congruent with thrombus. Both centrum semiovale had several regions smaller than 1 cm that seemed mildly hyperintense in DWI and isointense in ADC (Figure 2a). Extracranial and intracranial MR angiographies were clogged after the right ICA bulbous level; cervical, petrous and cavernous segments were not apparent but supraclinoid segment and the rest was visible. There was a diffuse, advanced narrowness for 1 cm at the left ICA segment distal (Figure 2b). Cranial computerized tomography (CT) showed destruction at the level of clivus and bilateral petrous bones (Figure 3). In the otorhinolaryngology examination, there were pulsatile purulent discharge on the bilateral acute otitis media and nasopharynx. Empirical meropenem and gentamicin treatment was started. *Staphylococcus aureus*, *serratia marcescens*, *escherichia coli* and yeast reproduced in the purulent discharge sample taken from nasopharynx. In the patient's hemogram, leukocyte count was 7280/ μ L (4000-11000), neutrophil rate was 63%, CRP level was 27.68 mg/dl (0-5), sedimentation rate was 72 mm/hour (0-20) and HBA1c level was 9.4% (4, 8-6). Her treatment was reorganized as meropenem for 3x2g/day and fluconazole 1x400 mg/day. A hypermetabolic region in the clivus was seen in the Fluorine-18 fluorodeoxyglucose (FDG)-PET examination. In combination with clinical and MRI data, the hypermetabolic locus in clivus

seen in FDG-PET was evaluated as infectious process (Figure 4). The patient's mastoid effusion was congruent with serous effusion, and her the infection locus was possibly not on mastoid. Thus, mastoidectomy was not performed. The patient was started on meropenem and fluconazole for 3 months with central skull base osteomyelitis diagnosis. The patient had right ICA occlusion and left ICA stenosis in the acute stage due to the infectious involvement and was started on enoxaparin 2x0.6 cc/day. After the completion of antibiotherapy, the patient was planned to be followed with antiaggregant treatment. Following the treatment, the leukocyte count dropped to 5200/ μ L, and CRP to 3 mg/dl. In the post-treatment neurological examination, there was still grade 5 peripheral facial paralysis; she could not move her tongue which was atrophied; soft palate elevation was present. The nasogastric tube was removed after she started swallowing again.

Discussion

In the differential diagnosis for skull base osteomyelitis, MRI or CT should be used to rule out other conditions, especially malignancy (9). In the cranial MR signal changes in the skull base bone marrow take place as the loss of cortical definition and contrast-holding appearance in the affected regions. Diffuse infiltration and contrasting of the soft tissues along the deep and superficial surfaces surrounding the skull base, posterior propagation of the infection towards the carotids causing obliteration of skull base foramen are frequently seen (10). In this situation, clinical findings indicating lower cranial nerve involvement are seen: Stylomastoid foramen involvement causes facial nerve paralysis, jugular foramen involvement causes 9th, 10th or 11th cranial nerve paralysis and hypoglossal canal involvement causes hypoglossal nerve paralysis (11). Our patient had right 7th, bilateral 12th, 9th and 10th cranial nerve paralyzes and there were bilateral invasion in ICAs in the MRI. The patient was diagnosed with atypical (central) SBO due to the MR images showing heterogeneous T2A hyperintense, T1A hypointense regions in clivus, bilateral petrous bones and occipital bone frontal part, and a hypermetabolic region in clivus in FDG-PET.

Magnetic resonance is the best imaging technique when it comes to viewing soft tissue boundaries around the skull base and detecting bone marrow abnormalities. Computerized tomography

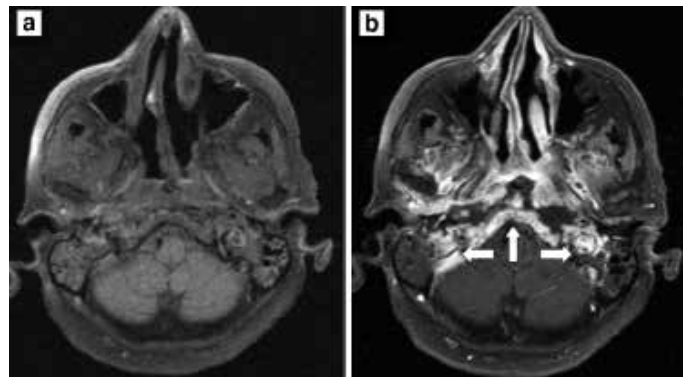


Figure 1. a, b. Uncontrasted (a) and contrasted (b) T1 slices showing dense contrast-holding lesions in clivus, bilateral petrous bones and occipital bone frontal part (white arrows)

may display a normal appearance due to the lack of bone erosion in the early stages (12). However, since the changes may persist in either modality even when the infection is cleared up, neither of these modalities should be used to evaluate the response to treatment (13).

Among the complications of SBO are venous sinus thrombosis, meningitis, abscesses, cranial neuropathies, and carotid invasions

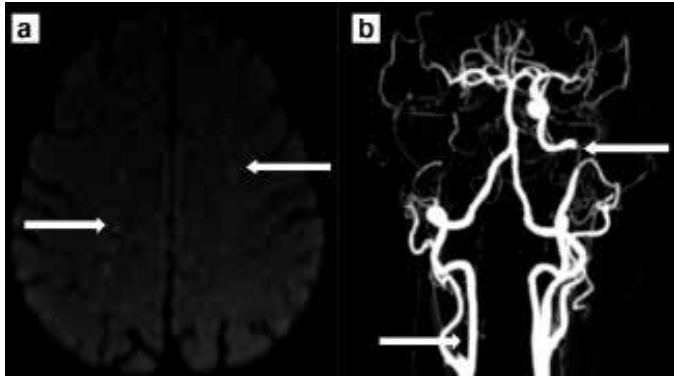


Figure 2. a, b. <1 cm areas showing mild diffusing-restriction, on both centrum semiovale (white arrows) (a). In extracranial MR angiography, right ICA is clogged starting from the bulbous level (white arrow). Right ICA cervical, petrous, cavernous segments cannot be seen. Right ICA cannot be visualized past the supraclinoid segment. 1-cm-long, severe, diffuse narrowness in the distal cervical segment of left ICA

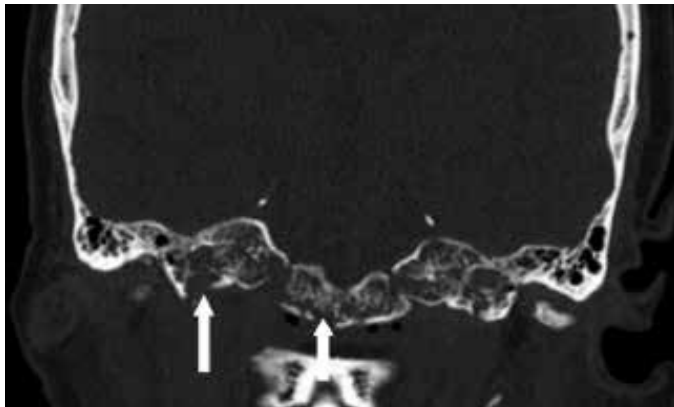


Figure 3. Bone destruction in the clivus and bilateral petrous apex shown in cranial CT (white arrows)

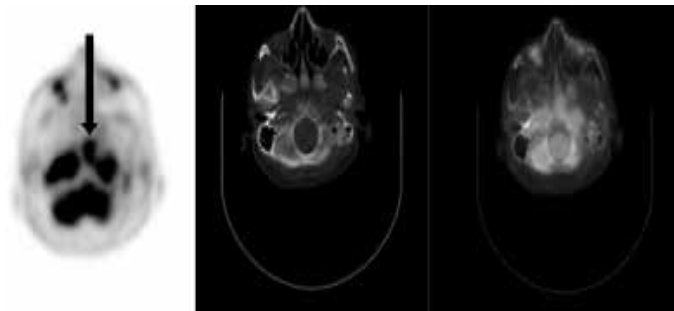


Figure 4. Hypermetabolic locus in the clivus, shown in FDG-PET, interpreted as infectious process (marked with black arrow)

with or without ischemic infarcts (1). Arterial complications of SBO are less frequently seen and often overlooked. Unilateral or bilateral stenosis in the ICAs can be seen in both CT and MR angiography. The inflammation in the cavernous sinus in central SBO can easily spread onto carotid siphon, causing inflammatory arteritis. ICAs are often surrounded by inflammatory tissue; contrast involvement and wall thickening is seen. Stenosis may be seen in the petrous segment of the carotid due to petrositis (14) and in the extracranial segment due to retropharyngeal abscess (15). In addition, ICA stenosis can also be caused due to the mass effect or due to the spasms caused by the surrounding inflammatory tissue. ICA stenosis can be clinically silent and temporary (15). In some patients with SBO, cerebral infarcts may develop due to critical ICA stenosis or septic embolism. Some patients may exhibit “watershed” infarcts due to cerebral hypoperfusion caused by bilateral ICA occlusion (16,17). In the subacute stage cranial MR examination of the case, the lesions seen in both centrum semiovale were thought to be asymptomatic infarcts due to bilateral ICA invasion and the patient was started on first anticoagulant and later antiaggregant treatment.

There is not sufficient information on the use of anticoagulation or antiaggregants for the thromboembolic events due to carotid artery infection. In one patient, cavernous sinus and right carotid artery involvement caused a.cerebri media (MCA) infarct, resulting in left hemiparesis, aphasia, homonym hemianopsia, and the treatment used antibiotherapy and aspirin. Hemiparesis and homonym hemianopsia improved but the aphasia remained (17). A patient with SBO causing right ICA bifurcation complete occlusion and right MCA infarct, resulting in left hemiparesis, right 9th-12th nerve paralysis and right Horner syndrome, was treated only with antibiotherapy (18). A patient with meningitis underwent cerebral angiography after the development of 3rd-6th cranial nerve paralyses. Severe narrowing was observed in the bilateral ICA cavernous segment along with mural thrombus, which suggested cavernous sinus thrombosis. Antibiotherapy and 4 months of warfarin treatment was given. The 6th month follow-up angiography showed that the left ICA narrowness has improved but the right side remained unchanged (19).

Fifteen patients who were histopathologically diagnosed as acute, chronic osteomyelitis or inflammatory spondylitis were differentiated perfectly by FDG-PET. In two patients, normal or decreased FDG involvement was observed in FDG-PET in parallel with the clinical improvement. Therefore it was concluded that FDG-PET has immense utility in the diagnosis and monitoring of osteomyelitis, but it fails to detect the infection reacting to changes seen in early postoperative stages (20). While CT and MR are good at displaying fractures and pseudarthroses, FDG-PET, being a functional imaging method, can help differentiate fractures and pseudarthroses from infections by showing the increased glucose metabolism in the inflammatory cells (20). FDG-PET can also differentiate osteomyelitis from the infection on the soft tissues surrounding bones (20,24). However, since the FDG involvement would increase in the malignant tissues, it should be kept in mind that metastasis and infection discrimination can be difficult in patients with malignity (20). The hypermetabolic clivus area seen in FDG-PET supported SBO in addition to MRI and clinical findings. PET was not repeated.

There are 3 treatment regimes in sBO:

- 1) An aminoglycoside and beta lactamase.
- 2) A third generation cephalosporins like ceftazidime.
- 3) An oral quinolone like ciprofloxacin.

Treatment is a long process driven by culture-antibiogram results and it is conducted according to the clinical and imaging improvements (5). In a study, all blood cultures of the 20 SBO patients came out negative and the ear canal granulation cultures were insufficient to diagnose in 70% of the patients. It was possible to isolate *Pseudomonas aeruginosa* (in ear canal swab samples) in half of the patients. Only one of them was found to be resistant to ciprofloxacin. All were sensitive to ceftazidime. Fungal factors were isolated in 2 patients. Mortality rate was found to be 15%. Since there are no guidelines for optimal treatment duration and scintigraphy repetitions, this study applied a 6-week antimicrobial treatment and conducted gallium-67 scintigraphy. Antimicrobial treatment was continued as long as the scintigraphy remained positive and it was repeated every 6 weeks (9). Intravenous cephalosporin and oral ciprofloxacin combination was shown to be effective in culture negative SBO (25). Recurrence can be seen even 1 year after the treatment (11). Advanced hyperbaric oxygen was shown to be effective in the treatment of advanced SBO (26). Surgical interventions may be necessary for the debridement of bone and inflammatory tissue, biopsy and culture (14). *Staphylococcus aureus*, *Serratia marcescens*, *Escherichia coli* and yeast reproduced in the purulent discharge taken from our patient's throat. The patient received meropenem and fluconazole treatment for 3 months and while her infection markers responded to the treatment, the neurological deficits remained largely intact.

In conclusion, SBO should be included in the differential diagnosis if a patient with pre-existing ear or sinus infection develops cranial nerve paralysis. Anticoagulant application in the acute stage can be beneficial in case of intracranial venous thrombosis.

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