A rare presentation of central skull base Osteomyelitis in a case of severe headache

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ABSTRACT

A 75-year-old fit and well patient with an old mastoid cavity presented to the ENT outpatient clinic with a four-week history of right-sided hemicranial pain with neuralgic characteristics.

Head and neck examinations via otoscopy and flexible nasoendoscopy were essentially unremarkable without evident cause for the patient's symptoms. Hence an urgent computed tomography (CT) neck with contrast was arranged, revealing an abnormal appearance of the petrous apex and nasopharynx. A subsequent magnetic resonance image (MRI) of the skull base including a cholesteatoma diffusion-weighted imaging (DWI) protocol confirmed abnormal appearances of the central skull base, with a differential diagnosis between osteomyelitis and an invasive nasopharyngeal neoplasia invading the petrous apex.

The patient was referred to a tertiary multi-disciplinary skull base team, and a consensus diagnosis of central petroclival skull base osteomyelitis was made. Treatment with oral antibiotics was commenced promptly with guidance from the microbiology team, and sequential follow-up MRI scans documented progressive improvement in the osteomyelitis with corresponding gradual resolution of the headache.

BACKGROUND

Skull base osteomyelitis (SBO) is a complex and relatively rare inflammatory disorder, predominantly found as a complication of malignant/necrotizing otitis externa (NOE) with involvement of the mastoid, temporal, and basal skull bones, with pseudomonas aeruginosa being the most common causative pathogen. It is primarily found in diabetic and immunocompromised patients, with a higher prevalence in males.

In NOE, the hallmark of clinical diagnosis is granulations arising from the floor of the ear canal, and in advanced cases, there are often cranial nerve palsies (vIIth nerve being the commonest) due to osteomyelitis of the lateral temporal bone (lateral SBO).

In contrast, central skull base osteomyelitis (CSO) is exceedingly rare and presents in a very different mode compared to the distinctive pattern of lateral SBO. CSO is often unrelated to ear pathology and tends to present initially with just a headache without any other clinical symptoms or signs, making clinical diagnosis difficult and often delayed. In addition, as this case demonstrates, the only way the diagnosis can be made early is on radiological grounds.

Even so, the radiological diagnosis of CSO is extremely challenging, even with a combination of CT and MRI scans when it is limited to the petroclival skull base, with inflammatory and neoplastic pathology often proving hard to differentiate in an anatomical area which is very difficult to biopsy for confirmatory diagnosis. However, early diagnosis is of paramount importance before the infection progresses to cranial nerve palsies and even, potentially, death.

The aim of this case report is to highlight the features of central/petroclival skull base osteomyelitis, which was diagnosed relatively early, with early treatment instituted, thus avoiding serious complications. Although very rare, it is a diagnosis worth remembering for all physicians managing headaches, and we would emphasize that discussion with tertiary-level experienced skull base radiologists is vital as an initial step to guide clinical management in the right direction from the start.

CASE PRESENTATION

A 75-year-old male, who had been attending an ENT clinic for many years for routine mastoid cavity clearance, presented with a four-week history of right-sided severe hemicranial pain with neuralgic characteristics, affecting the upper division of the trigeminal nerve, and specifically, increased lacrimation. He reported that the pain was primarily located at the centre of the head and posterior aspect of the nose. Also, he had an unremarkable mastoid cavity on the right with minimal discharge, although over the past two years ears swabs had grown pseudomonal swabs on more than one occasion. Neurological examination, including cranial nerve examination, was normal. The patient was reasonably fit and well for his age, without any background of diabetes or immunosuppression.

Although flexible nasoendoscopy was unremarkable during the initial examination as the rest of his ENT assessment, subsequently, oedema of the right post-nasal space was found on subsequent visits. The patient was commenced on a diagnostic trial indomethacin for ruling out hemicrania (as it is a criterion for diagnosis of this condition in unilateral headaches with lacrimation), and an urgent CT head and neck scan with contrast was arranged.

INVESTIGATIONS

An urgent CT head and neck with contrast (Figure 1) reported findings consistent with soft tissue inflammation in an otherwise normal mastoid cavity, without evidence of bony erosion or skull base osteomyelitis. Additionally, there was an incidental finding of bilateral parathyroid nodules, but normal parathyroid hormone (PTH) levels (5.9 ppmol/L) ruled out functioning parathyroid adenomas. Due to the limitations of CT, a cholesteatoma could not be excluded, and the radiologist arranged for an MRI neck with contrast and MRI internal acoustic meatus (IAM) as per the diffusion-weighted imaging MRI (DWI) cholesteatoma protocol.

Based on the initial CT report, the patient was commenced on oral Augmentin and ciprofloxacin for any underlying mastoiditis. Additional blood tests failed to confirm an infective picture with normal white cell counts (WBC 5.2 10*9/L, Neutrophils 3.7 10*9/L) and C-reactive protein only very mildly elevated (10.7 mg/L).

The subsequent DWI scan report did not flag up any cholesteatoma in the mastoid cavity. However, the MRI neck with contrast and IAM (Figure 2) flagged up a lesion in the petrous temporal bone extending to the clivus, which was reported to be of uncertain aetiology with regards to being infective or neoplastic, and a post nasal space biopsy under anaesthetic was proposed for diagnosis. However, in view of the fact that the nasoendoscopy was relatively normal, the

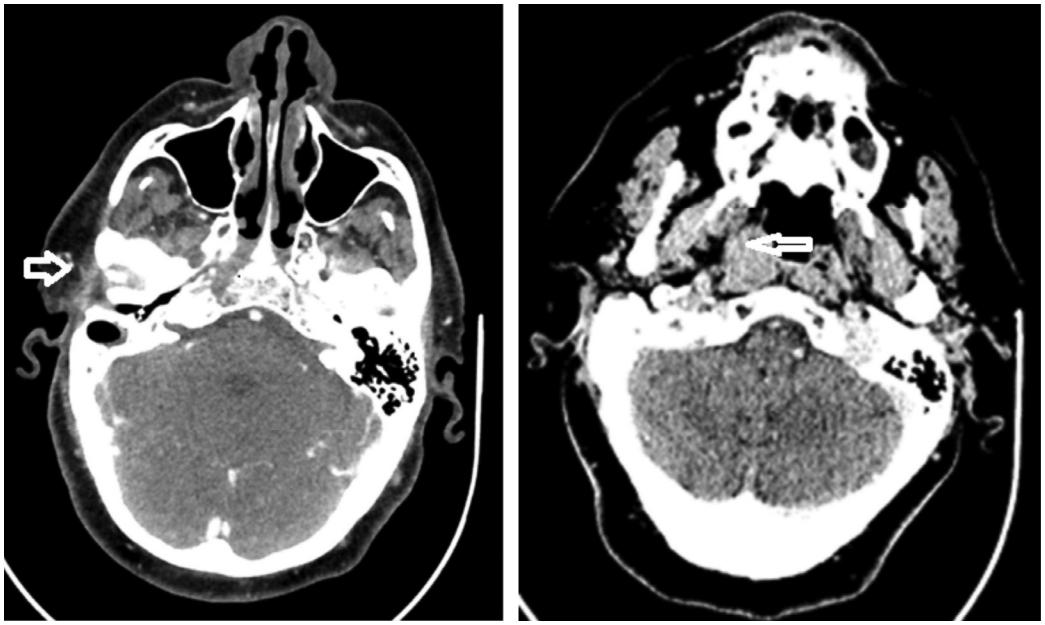

Figure 1: CT mastoids (left) showing previous right mastoidectomy cavity, with some soft tissue opacity (white arrow), which prompted a DWI MRI scan to assess any recurrent cholesteatoma. CT post-nasal space (right) shows some asymmetry in the nasopharyngeal tissues, more prominent on the right (white arrow).

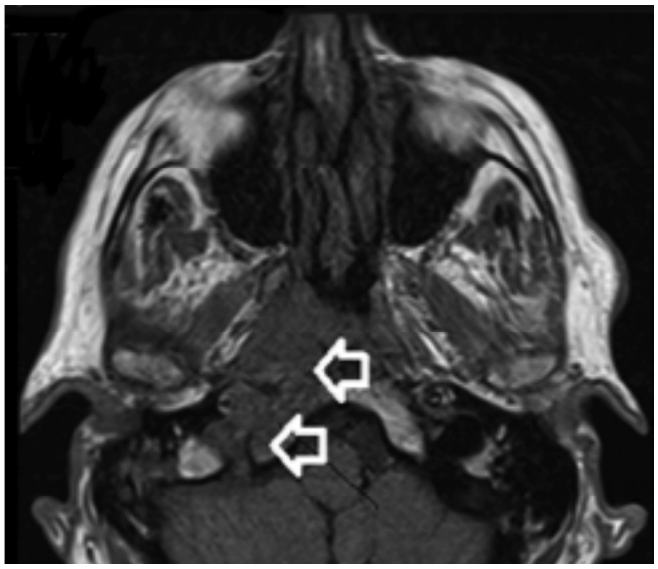

Figure 2: MRI: The abnormal signal extending along the petrous apex into the right side of the clivus and involving the right side of the nasopharynx (white arrows). Based on the above appearances, a biopsy from the right nasopharynx was suggested.

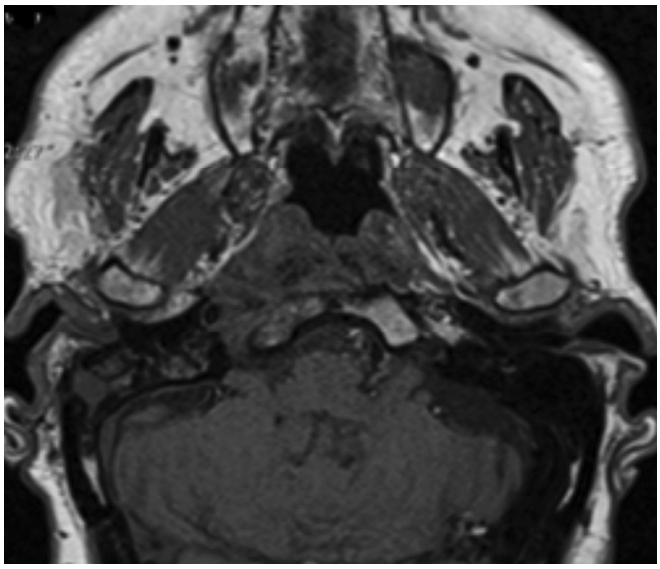

Figure 3: Surveillance MRI at six weeks: There is significant improvement of the inflammatory changes in the right side of the central skull base. There is a small area of marrow oedema and enhancement within the right petroclival region, which as previously demonstrated involves the walls of the carotid canal. There is a small area of enhancing soft tissue in the right nasopharynx. This still obstructs the right eustachian tube as evidenced by effusion in the right middle ear.

ENT clinician decided to obtain a tertiary opinion with a skull base multidisciplinary team. The skull base MDT promptly took place after careful evaluation of the preceding scans with clinical correlation, and a provisional diagnosis of a central petroclival skull base osteomyelitis was made after a general consensus that a possible submucosal malignancy from the nasopharynx invading the petroclival region was less likely.

TREATMENT

The patient was commenced on a combined oral antibiotic regimen of ciprofloxacin and clindamycin, as per the advice of the microbiologists, for an initial period of six weeks.

A routine surveillance MRI neck with contrast and IAM (Figure 3) was performed five weeks through the oral antibiotic therapy to assess treatment response, revealing the marked improvement in the appearance of the skull base osteomyelitis, and there was a clinical correlation with the pain becoming less frequent and intense.

Another surveillance MRI was repeated after a further six weeks (Figure 4), which revealed stable disease with no further progression of the SBO. The patient was reviewed in the clinic after the completion of ten weeks of antibiotic therapy, and not only was his headache entirely resolved, but his right mastoid cavity was also completely dry.

The patient will continue to be monitored in outpatients even though he remains asymptomatic.

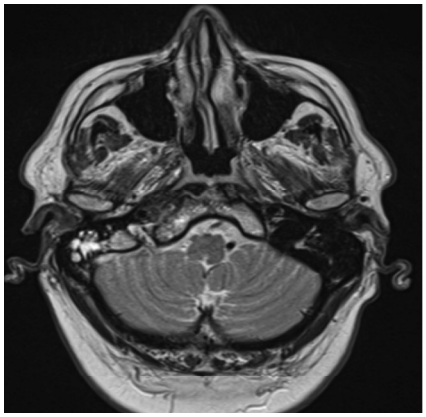

Figure 4: Surveillance MRI at twelve weeks after completing ten weeks of antibiotic treatment shows symmetric appearance of petroclival region, despite some marrow signal changes and abnormal enhancement in the right petroclival region extending into the right side of the nasopharynx.

CASE DISCUSSION

Although central skull base osteomyelitis is a known rare entity, the vast majority of cases are seen as an extension of lateral skull base osteomyelitis due to necrotizing otitis externa,[1] quite often resulting from failure to control the infection, leading to potentially serious and fatal consequences of cranial skull base involvement in the central compartment.

The alternative terminology for central skull base osteomyelitis is 'Atypical Osteomyelitis of the Skull Base' (AOSB), and a recent paper[2] has pointed it out that it is an emerging clinical entity often seen without any prior otological or rhinological infection.

In the absence of otitis externa, the diagnosis of this entity is quite tricky on clinical grounds, especially in the early stages, as this case report illustrates. It presented with just central headache with lacrimation, with a normal ENT examination and no change in ear symptoms with regards to a mastoid cavity that had been discharging for decades. Therefore, as has been reported in large case series, mostly in radiology journals, the early diagnosis of this condition often depends upon a radiological assessment of possible causes of headaches.[3] In this regard, the gold standard radiological diagnostic tool is an MRI scan.

However, the radiological interpretation is rarely straightforward, and quite often a biopsy of the nasopharynx and the clivus is recommended. In many instances swelling in the posterior nasopharynx is noted on endoscopy, and endoscopic biopsies of the nasopharyngeal lining are not always straightforward. Sometimes biopsies of the clivus have also been carried out using endoscopic techniques, as discussed in the literature.[4]

Therefore, it has been pointed out in many instances that diagnosis has been delayed in central skull base osteomyelitis due to the logistics involved in obtaining good quality radiological scans[5] and often followed by histological assessment after a biopsy. In rapidly progressive skull base infections, usually with pseudomonas aeruginosa, such delays can lead to irreversible neurological complications through cranial nerve damage. In our case, the blood tests, particularly the inflammatory markers and white cell counts, did not point out any significant systemic inflammatory response at the onset of the headache, which illustrates another important feature of skull base infections – pain is often an important marker of the infection, and even response to treatment.

Another key message of this paper which has also been pointed out by other authors,[4] is that the need for biopsies can be obviated by a combined meticulous clinical approach combining clinical examination and expert radiological assessment and, sometimes, assessing the therapeutic response to ciprofloxacin. Pseudomonas aeruginosa is the usual agent in most cases, apart from some rare instances of aspergillus fumigatus and similar fungal or mixed fungal infections.[6]

The role of experienced neuroradiologists is invaluable, as was the case with the patient illustrated in this paper. Early treatment is the key to preventing serious complications, and sound clinical decision-making with radiological diagnosis is perhaps the quickest route to instituting prompt treatment in a generally quite morbid condition like CSBO. This case also points out that in early cases, oral antibiotic treatment can be adequate for a complete clinical response, although the patient in this case report was not diabetic or immunosuppressed, and this would be another argument in favour of an early diagnosis and start of antibiotic treatment.

In this particular case, the source of infection was almost certainly the mastoid biofilms that had led to discharge on and off from the cavity for decades. Certainly, this case could be considered one of the first reports of isolated central skull base osteomyelitis related to an old mastoid cavity chronic biofilm infection.

However, it must be pointed out that in the literature, central skull base osteomyelitis without any ear infection has been reported to be also associated with nasal and paranasal sinus infections and, in one of the largest systemic reviews in recent years,[7] was described as an emerging clinical entity. The need for surgical debridement is now considered relatively rare compared to long-term antibiotic treatment.

Finally, it has been pointed out in the literature that radiological abnormalities persist way longer than the clinical resolution of the symptoms, and that is the reason why the patient in our case report will undergo repeated clinical evaluation despite the full resolution of his current symptoms, as long-term follow-up is always advised.[8]

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