CASE REPORT

An intriguing case of locked jaw secondary to melioidosis

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SUMMARY

A 56-year-old woman presented with fever, pain and restriction of movement of the right temporomandibular joint. She was premorbidly diagnosed to have type 2 diabetes mellitus and rheumatoid arthritis. Local examination revealed a poorly demarcated severely tender, erythematous swelling in the right preauricular region. All haematological and biochemical investigations were within normal limits. MRI of the neck revealed the presence of a masticator space infection with intramuscular abscess involving the masseter and the temporalis muscles along with intracranial extension. Osteomyelitic changes were detected in the right mandibular condyle, temporal bone and in the temporomandibular joint. Melioidosis was suspected due to this unique clinical presentation of an abscess at an unusual and atypical site. Blood cultures identified the Gram-negative bacilli *Burkholderia pseudomallei*, which established the diagnosis of Melioidosis. Remarkable improvement was attained with antibiotics meropenem and cotrimoxazole, deferring the need for any surgical intervention.

BACKGROUND

Melioidosis caused by *Burkholderia pseudomallei* is a rare but potentially life-threatening infection with a high mortality rate. The disease is predominantly seen in Australia and southeast Asia, including parts of India,^{1 2} where the organism is commonly found in water and soil, especially in areas receiving heavy rainfall, and is transmitted by inoculation, ingestion and, rarely, by inhalation. The hallmark of melioidosis is formation of abscess, most commonly in the lungs, but it may involve any organ.³ Central nervous system (CNS) melioidosis, though rare, comprising only 3% of all cases, is associated with higher morbidity and mortality than other presentations of this bacterium.⁴ Moreover, as noted by Dance, the available case reports perhaps represent just the tip of the iceberg, as the disease poses a diagnostic challenge to the physician.¹ The disease is rare and often manifests with atypical presentations, and unless the laboratory test is forewarned of its possibility, the organism may easily be misidentified. We report a case of extensive head and neck melioidosis presenting as locked jaw, in which secondary neurological spread was picked up incidentally. Locked jaw due to melioidosis has never been described before in the literature. The case highlights the importance of suspecting melioidosis in individuals presenting with abscess at uncommon and atypical sites.

CASE PRESENTATION

A 56-year-old woman presented to us with a 1-week history of fever, pain in the region of the right temporomandibular joint (TMJ), associated with swelling in the right preauricular region, and inability to open her mouth. She gave no other positive history. Her medical history was positive for rheumatoid arthritis (RA), for which she was only taking symptomatic treatment. She was also recently diagnosed with type 2 diabetes mellitus for which she was on insulin.

Systemic examination was unremarkable. Local examination revealed severely tender, erythematous, poorly demarcated swelling in the right preauricular region. Mouth opening was painful and severely restricted to inserting one finger, essentially causing locked jaw.

INVESTIGATIONS

Routine investigations including blood counts and biochemical parameters were normal. Serological testing for mumps antibodies performed to rule out parotitis secondary to mumps was negative. Markers of active rheumatoid disease, erythrocyte sedimentation rate and C reactive protein, were within normal limits. Ultrasound of the neck revealed the possibility of an early abscess in the right masseter muscle. Since the patient continued to spike fever and reported increasing pain, MRI of the neck was carried out, with perplexing results. It showed the presence of a masticator space infection with an intramuscular abscess involving the masseter and the temporalis muscles with sparing of the parotid gland (figure 1). There was an intracranial extension of the infection with involvement of the leptomeninges and focal cerebritis in the temporal lobe (figure 2), and associated osteomyelitic changes detected in the TMJ, right mandibular condyles and in the underlying temporal bone.

The clinical picture of an abscess developing at an unusual site such as the masseter and temporalis muscle made us consider the diagnosis of melioidosis. Blood culture grew Gram-negative bacilli with repeat cultures identifying the Gram-negative organism as *B. pseudomallei*, thus confirming our diagnosis.

DIFFERENTIAL DIAGNOSIS

The presence of fever along with the location of the swelling and restricted mouth opening prompted us to think of the possibility of an infective aetiology involving the parotid, TMJ or regional lymph nodes. Hence, the initial differentials included:

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Figure 1 T1-weighted image with contrast: suggestive of abscess in the right masseter.

1. Acute parotitis possibly secondary to mumps;
2. Preauricular lymphadenitis;
3. Flare-up of RA with TMJ involvement;
4. Neck muscle abscess.

However, MRI and blood cultures proved otherwise and helped us clinch the diagnosis of a masticator space infection secondary to melioidosis.

TREATMENT

The patient was managed with parenteral meropenem, 1 g, administered intravenous every 8 hours for 14 days, with aggressive control of sugars and other supportive care. Eradication therapy with cotrimoxazole 240/1200 mg orally twice a day for

Figure 2 T1-weighted image with contrast: leptomeningeal and pachymeningeal enhancement along the right temporal lobe with focal cerebritis in basitemporal lobe and osteomyelitic changes of the overlying right temporal bone.

a period of 6 months was initiated. Since there was a remarkable improvement with antibiotics, surgical intervention was deferred.

OUTCOME AND FOLLOW-UP

The patient showed remarkable improvement with antibiotics along with aggressive glycaemic control, and was discharged in good general condition. She was counselled regarding the need to continue cotrimoxazole for a period of 6 months and to report at the earliest in case of any adverse drug reaction such as skin rash, jaundice, seizures, weakness or recurrence of fever. She was followed up on outpatient department basis and was asymptomatic.

DISCUSSION

Abscess formation is a trademark of *B. pseudomallei*. Existing case reports describe prostatic abscess, parotid abscess, liver and splenic abscess secondary to melioidosis, however, an extensive literature search yielded no results on abscess primarily involving the masseter and, hence, this is the first case report of a locked jaw due to melioidosis.³ Moreover, the CNS extension of the disease in our patient highlights the propensity of this saprophyte to cause rapid worsening, even from a relatively benign presentation, if not picked up promptly.

The disease can have a myriad clinical presentations ranging from pneumonia, which is the most common manifestation, to suppurative parotitis, CNS involvement, septic arthritis and osteomyelitis, as well as from disseminated disease and septic shock.³ Neurological involvement secondary to melioidosis is rare with under-50 cases reported over 30 years.⁵ However, the disease has a high morbidity and mortality. According to a 20-year prospective study on the subject by Currie *et al*,⁴ it carries a 21% mortality rate, as opposed to the more general 14% mortality rate associated with other presentations of melioidosis. CNS involvement can occur in the form of primary meningoencephalitis or secondary encephalitis and brain abscess following haematogenous or contiguous spread. Presenting symptoms may include a near normal CNS evaluation, neck stiffness, ataxia, dysphasia, limb weakness, urinary retention, flaccid paralysis and cranial nerve palsies.⁴⁻⁷ Secondary neurological melioidosis likely follows haematogenous spread following pneumonia, or spread from contiguous structures such as orbital cellulitis or facial sinuses.^{8,9} It is typically associated with a higher rate of positive blood cultures than primary neurological melioidosis.⁴ In this patient, since the lung fields were clear and the disease was limited to the head and neck, the neurological spread could have resulted from contiguous spread from the masticator space infection and osteomyelitis of the underlying bones. The positive blood culture also fits the clinical picture of secondary neurological disease. A combination of both extracranial involvement as masseteric abscess, asymptomatic intracranial involvement as a brain abscess and osteomyelitis of underlying bone masquerading as lock jaw, has never been described before in the literature. Moreover, an incidentally picked up intracranial spread reiterates the need for an MRI of the brain even in cases of extracranial head and neck lesions.

Risk factors for the disease include conditions with impaired neutrophilic function such as diabetes, as seen in this case and also renal disease, excessive alcohol consumption and thalassaemia.¹⁰⁻¹³ However, HIV is not reported to be a risk factor and hence the role of CD4-mediated immunity is unclear. Humoral immunity is far less significant, which is evident from the fact that the antibody response resulting from repeated natural exposure to the organism is insufficient to provoke a protective

response. Exposure to wet soil and rice fields where the organism is found in abundance are other risk factors.

Rapid diagnostic tests are unreliable and the disease is diagnosed mainly by positive cultures. Although Gram staining of the organism typically shows Gram-negative rods with bipolar staining, the morphology in actual clinical specimens is extremely variable, hence the microbiologist must be cautioned or the diagnosis may be missed.¹⁴ Brain imaging is a must and brain MRI has proven to be better than CT in terms of sensitivity, especially during the early stages, allowing for the recognition of even subtle abnormalities. Typically, MRI signs of neurological melioidosis include calvarial osteomyelitis, leptomeningeal enhancement and abscess, as also seen in our patient. Ring-enhancing lesions, oedema and brainstem involvement may also be seen.⁷

Patients diagnosed with melioidosis should be started on initial intensive therapy using ceftazidime or meropenem for 14 days, which may be extended in critically ill patients. This should be followed by eradication therapy with cotrimoxazole for 3–6 months. A randomised comparative trial in Thailand revealed that not only was there no significant difference in mortality between groups using high-dose imipenem and ceftazidime, the group that received imipenem also suffered fewer treatment failures.^{15–18} Hence, carbapenems are preferred in intensive care unit settings. Prostate abscess and, rarely, parotid abscess, may need surgical drainage.

Learning points

- ▶ *Burkholderia pseudomallei* should always be considered in the differential diagnosis of patients presenting with abscess at unusual sites.
- ▶ Brain imaging should be considered in melioidosis of the head and neck due to propensity of the disease to spread from contiguous structures.
- ▶ Neurological melioidosis is associated with a high mortality rate of 21%, which is higher than the general mortality rate associated with other clinical presentations of the disease.
- ▶ Early diagnosis and aggressive treatment with antibiotics is a must, as an initial benign presentation can rapidly spread to contiguous structures or disseminate and lead to high morbidity and mortality.
- ▶ The case highlights the importance of awareness of melioidosis among clinicians and microbiologists to avoid missing the diagnosis.

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