

## Case Report

# Cervical spinal cord compression: a rare and serious complication of *Actinomadura pelletieri* actinomycetoma

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**Introduction:** Mycetoma is a chronic granulomatous inflammatory disease predominantly affecting the foot and hand. The cervical region is an uncommon site for mycetoma and spinal cord compression is a rare complication.

**Case presentation:** This communication reports on a 40-year-old male farmer from Western Sudan who presented with quadriparesis due to cervical spine cord compression caused by *Actinomadura pelletieri* actinomycetoma. His condition started with a small painless subcutaneous swelling in the right shoulder region that gradually increased in size to involve the right side of the neck and the cervical spinal cord ending in progressive quadriparesis. He made a good response to an extended course of antibiotics, but was left with mild disability.

**Conclusions:** *A. pelletieri* is an uncommon cause of actinomycetoma, and the clinical presentation of the reported patient is a rare and serious sequela of mycetoma. The literature contains only a very few reports on such presentation, and our case report will add to the knowledge and experience in managing such a presentation.

**Keywords:** actinomycetoma; clinical presentation; complications; management; mycetoma; quadriparesis; spinal cord; Sudan.

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## Introduction

Mycetoma is a neglected tropical disease, which is endemic in many tropical and subtropical areas in what is known as the mycetoma belt (Fahal, 2004). It has devastating medical and socio-economic impacts on patients and communities in endemic areas (Fahal, 2011). The reported patient presented with quadriparesis due to cervical spinal cord compression caused by actinomycetoma, which is a serious and potentially fatal condition. Cervical actinomycetoma with cord compression is a rare sequela of mycetoma, and this case report intends to share experience and advance knowledge on the disease.

## Case presentation

A 40-year-old male farmer from Western Sudan was referred to the Mycetoma Research Centre, Khartoum, Sudan with a history of bilateral upper and lower limb severe weakness rendering him immobile for 45 days. The weakness was gradual but progressive. Initially he developed right-sided weakness, progressing within a

matter of a few days to involve the left side; he subsequently became bed-bound. He denied neither sensory nor sphincteric deficits. He had no change in vision, or difficulty in swallowing or in breathing. There were no symptoms suggestive of raised intracranial pressure or higher cortical function disturbances.

Six years prior to presentation he noted a small painless right shoulder swelling. It progressively increased in size, and involved the right aspect of the neck and extended posteriorly towards the back of the neck. A localized dark discoloration of the skin over the swelling with multiple sinuses discharging red grains then developed. The patient could not recall a history of trauma at the swelling site.

He initially presented to a district hospital where he received some medication for 2 months. There was a reduction in the size of the swelling; however, the patient was not aware of the diagnosis or medication given.

He had no medical co-morbidities, no previous surgical intervention and he was not on regular medications. He is a farmer of low socio-economic status and there was no family history of mycetoma.

Abbreviation: MRI, magnetic resonance imaging.

Clinical examination revealed an ill bed-bound male. He was not pale, icteric or cyanosed. His pulse rate was  $80 \text{ min}^{-1}$  regular, normal volume and not collapsing. Blood pressure supine was 120/80, respiratory rate was  $19 \text{ min}^{-1}$  and temperature was  $37.7^\circ\text{C}$ .

The cardiovascular, respiratory and abdominal examinations were normal. On local examination, there was a large swelling on the right shoulder region extending to the right supraclavicular fossa, right aspect of the neck and posteriorly to the back of the cervical region. The swelling was about  $20 \times 15 \text{ cm}$  in size with a nodular surface. The skin over the swelling was dark in colour with multiple healed and active sinuses discharging serous fluid and small red grains. The swelling was firm in consistency and slightly hot compared with the adjacent tissue. It was firmly attached to the skin and fixed to the underlying structures (Fig. 1). Regional lymph nodes were not palpable. There was a normal carotid pulsation bilaterally.

Neurological examination revealed an patient orientated to time, place and persons, with intact recent and remote memory. No apparent cranial nerve abnormalities. No facial asymmetry. Pupils were normal in size, and reactive to direct and consensual light reflex. Eye movements were normal with no diplopia. Tongue movement was normal. Soft palate and uvula were central. Hearing was

normal. Neck movements were restricted. Hearing assessment was reported as normal.

The muscles of the upper limbs were wasted, spastic, had grade 1 power and were hyper-reflexic. No fasciculation was noted. Sensation to pain, touch and temperature was intact, but it was not possible to assess coordination.

Lower limb examination was similar, but power grade was 0 bilaterally with up going planter reflexes. Sensation to pain, touch and temperature was intact. Abdominal reflex was absent.

Investigations revealed haemoglobin of  $10.6 \text{ mg dl}^{-1}$ ; renal and hepatic profiles were within normal. Chest X-ray revealed a soft tissue mass between the right shoulder and neck, and otherwise normal lungs. There was thickening of the right clavicle with periosteal reaction. Cervical and shoulder magnetic resonance imaging showed ill-defined soft tissue mass extending from the right shoulder and supraclavicular areas, and infiltrating all soft tissues, muscles and deep structures all the way down to the cervical spine vertebrae. There was large long epidural component noted from C2 down to T1 causing significant spinal cord compression, encasement and displacement of the cord to the left alongside spinal canal stenosis (Fig. 2).

An incisional biopsy was obtained from the swelling which established the diagnosis of *Actinomyadura pelletieri* surrounded by neutrophils, lymphocytes and multinucleated giant cells representing type I and II host tissue reactions (Fig. 3). Grains were cultured in Lowenstein-Jensen media and growth was typical of *A. pelletieri*, which confirmed the diagnosis.

The patient was hospitalized and commenced on a combination of intramuscular amikacine sulphate  $15 \text{ mg kg}^{-1} \text{ day}^{-1}$  and co-trimoxazole  $8/40 \text{ mg kg}^{-1} \text{ day}^{-1}$  orally. He developed ototoxicity due to the amikacine sulphate and it was stopped. The medication was resumed with co-trimoxazole  $8/40 \text{ mg kg}^{-1} \text{ day}^{-1}$  orally combined with amoxicilltn/clavulanic acid  $2 \text{ g day}^{-1}$  for 6 weeks. He showed good response. He regained power grade 4 in both upper limbs, grade 3 in the right lower limb and grade 2 in the left lower limb. Six months later he continued to improve and was able to mobilise independently with a minor disability.

## Discussion

Mycetoma is one of the notoriously neglected tropical diseases worldwide and the disease prevalence in the Sudan is considered the highest globally (Ahmed *et al.*, 2004). It is a chronic, granulomatous subcutaneous specific infection, caused by certain fungi (eumycetoma) or bacteria (actinomycetoma). Mycetoma is characterized by the formation of a painless subcutaneous mass, multiple sinus formation and discharges that contain grains (Fahal, 2004). The foot is the commonest site affected by mycetoma and accounts for 70 %, followed by the hand (12 %) (Fahal, 2004, 2011). Spinal cord involvement is rare, but serious



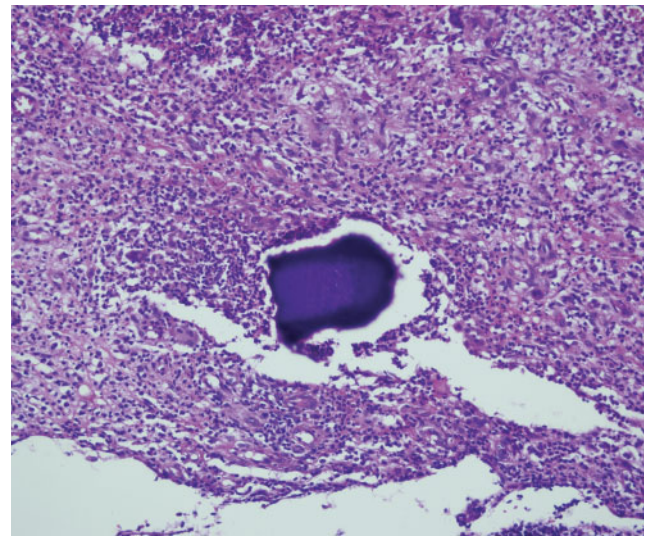
**Fig. 1.** Photograph showing the right shoulder lesion.



**Fig. 2.** Cervical region MRI showing ill-defined lesion extending from right shoulder and supraclavicular areas infiltrating the soft tissues, muscles and deep structure all the way to reach the cervical spine vertebrae. Large long epidural component noted from C2 down to T1 causing significant cord compression, encasement and displacement of the cord to the left alongside spinal canal stenosis.

and potentially fatal, and only a few cases have been reported in the medical literature (Cascio *et al.*, 2011; Fahal *et al.*, 2012).

The disease presents initially with a small, localized painless mass which is confined to the subcutaneous plane; however, with disease progression it reaches a considerable size and extends to involve the underlying structures and bones, causing disability and deformity (Fahal, 2013). The natural history and outcome of the disease depend on the underline causative agent, disease site and immune status of the patient (Mahgoub & Murray, 1973). In general, actinomycetoma is more aggressive, destructive and invasive than eumycetoma. *A. pelletieri*, the causative organism encountered in the reported patient, is a rare causative agent of actinomycetoma. It is characterized by the absence of cement substance, and this may explain the massive and aggressive disease we report in this case report (Fahal *et al.*, 1994). Several factors



**Fig. 3.** Histopathological section of the surgical biopsy showing a grain of *A. pelletieri* with type I and II tissue reactions. Haematoxylin and eosin stain,  $\times 200$ .

contributed to the rather long disease duration encountered in this report, amongst which were the patient's poor health education, low socio-economic status, painless disease nature and unavailability of local health facilities.

Mycetoma management depends on isolating the causative agent and assessing the extent of the disease. The identification of the causative organism can be established by direct culturing of viable grains, fine needle aspiration for cytology, histopathological examination of surgical biopsy and PCR identification of the causative agents (Abd El Bagi, 2003; Ahmed *et al.*, 1999; El Shamy *et al.*, 2012; Fahal *et al.*, 1997).

Imaging studies play an important role in determining of the extent of the disease. MRI usually delineates the extent of soft tissue and bone involvement; it is characterized by the dot-in-circle sign, conglomerated low signal intensity foci, and macro and micro-abscesses on the background of a hypo-intense matrix. A mycetoma MRI grading system (Mycetoma Skin, Muscle, Bone Grading System) was set up to grade disease severity and therefore facilitate patient management accordingly (El Shamy *et al.*, 2012). Radiography, ultrasonography and computed tomography scan are commonly used, but are less accurate than MRI (Abd El Bagi, 2003; Fahal *et al.*, 1997). In the reported patient the extent of the disease was determined accurately by MRI examination.

Actinomycetoma is characterized by good response to pharmacological therapy. Combination therapy is preferred to reduce drug resistance (Welsh *et al.*, 2012). The common drugs in use are intramuscular amikacin sulphate, at a dose of  $15 \text{ mg kg}^{-1} \text{ day}^{-1}$  in combination with oral co-trimoxazole at a dose of  $8/40 \text{ mg kg}^{-1} \text{ day}^{-1}$ . They are given in cycles, each cycle lasting 5 weeks. Amikacin sulphate is given for only 3

weeks and co-trimoxazole is given throughout the 5 week cycle. Audiometric and renal function must be monitored in between cycles. The number of cycles to be administered depends on the clinical response and presence of side effects. Second-line treatment includes rifampicin, sulfadoxine/pyrimethamine, sulphonamides, amoxicillin/clavulanic acid and imipenem (Welsh *et al.*, 2012).

Our reported patient responded well to treatment despite the advanced disease. This is in line with our experience that, in general, the response to medical treatment is better in actinomycetoma compared with eumycetoma.

To date there are no effective preventative and control measures, and thus health education is crucial to reduce the enormous deformity, disability and high morbidity, which our reported patient has demonstrated.

## Conclusions

Cervical spinal cord compression due actinomycetoma caused by *A. pelletieri* is a rare but a serious medical problem. The recognition of this condition and active and proper treatment is essential to avoid its grave and fatal consequences.

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The patient gave written consent to publish his case. The authors declare that they have no competing interests.

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