CASE REPORT

Pseudoathetosis and ataxia – a rare presentation of multibacillary leprosy in a non-endemic area

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Introduction

This report details the case of a patient with pseudoathetosis and sensory ataxia secondary to multibacillary leprosy. To the best of our knowledge there is only one previously reported incident of pseudoathetosis associated with leprosy.

Case Report

A 51 year old female presented to an aged-care facility in North-Western Panama, where for the last three decades the prevalence and incidence rate of leprosy has been similar to most OECD countries at below 20 new cases per year.1 Symptoms arose at the age of 29 and progressed over decades. Medical assessment revealed a malnourished, cognitively intact and otherwise healthy post-menopausal woman on no medications. Her neurological examination was significant for the following:

Marked athetoid writhing of the trunk, arms and legs - worse distally and with eyes closed, disappearing while asleep; an ataxic, wide-based gait with left foot drop (power 2/5); a complete right peripheral facial nerve palsy; right-sided fixed-flexion deformity of the first, third and fourth digits and fixed extension of the wrist (Figure 1); anaesthesia of the lower limbs to the level of the mid-tibia, with preservation of sensation on the dorsum of the right foot and loss of proprioception and vibration in the upper limbs to the elbow and in the left lower limb to the knee.

Numerous (more than five) hypopigmented, anaesthetic lesions were noted around the trunk, knees, and hands, with sparing of the fifth digit of the left hand. Deep tendon reflexes were absent at the left ankle, right elbow and wrist. The right plantar reflex was down-going. Finger-nose and heel-shin tests were impaired in all limbs more-so in the right. The right great auricular and left common fibular nerves were markedly thickened. Additional findings...
included the absence and/or deformity of multiple digits; a deep, painless, $2 \times 2 \times 3$ cm ulcer on the medial volar aspect of the right foot, immediately proximal to the first digit; a painless $3.5 \times 2.5$ cm decubitus ulcer over the left ischial tuberosity; wasting of the right periocular muscles and a right-sided cataract.

Differential diagnoses included Friedreich’s ataxia, multiple sclerosis and diabetic, alcoholic, autoimmune or toxic peripheral neuropathy. After 2 weeks the diagnosis of multibacillary leprosy was made on clinical findings and smears collected from both ulcers and hypopigmented lesions, which stained positive for acid-fast bacilli.

**Discussion**

In this case, signs pathognomonic for leprosy included the hypopigmented, anaesthetic lesions; slowly erosive ulcers and distal loss of digits; thickened peripheral nerves with corresponding neuropathy and a cataract. Involvement of the facial, greater auricular and fibular nerves in particular is typical, as these traverse bony prominences, providing the relatively cooler environments preferred by *M. leprae*. Of note in this case was the presence of pseudoathetosis. The only previously reported incident of leprous pseudoathetosis was an acute case of paucibacillary leprosy, with rapid deterioration, pain and Type-I lepra reaction following 12 days of anti-leprosy treatment. Although a similar pattern of pseudoathetosis and sensory ataxia was described in our patient, the development of her symptoms occurred over a longer time-course, without pain, and was not associated with the administration of antibiotics. The cause of pseudoathetosis is significant proprioceptive deafferentation at any stage of the sensory pathway from peripheral sensory nerves to the parietal cortex. In the case of leprosy the pathology is limited to peripheral nerves. The patient’s ataxic gait is proposed to have similar origins, as described in previous reports of patients with polyneuritic leprosy,

![Figure 1. Fixed flexion deformity of the first, third and fourth digits, impaired flexion of the right hand and an irregularly shaped anaesthetic, hypopigmentated lesion on the volar surface of the forearm.](image)
absent tendon reflexes in the ataxic limbs and no other cause for ataxia. Follow-up after 4 months of MDT treatment showed moderate improvement of ulcers and foot drop (left ankle dorsiflexion power increased to 4/5) but all other symptoms persisted.

Due to the late diagnosis, the facility’s location in a non-endemic area and limited supplies, appropriate antibiotic therapy was delayed for several weeks. Transmission becomes negligible within 24 hours of therapy. This prompted questions regarding confidentiality, and the need for personal protection and/or patient isolation to protect staff and residents of the aged-care facility. Current research suggests that transmission requires a genetic susceptibility coupled with sustained close proximity to a patient with active disease. As such, isolation and use of respiratory precautions was deemed unnecessary. Confidentiality was strictly enforced.

Conclusion

This case presented with unusually severe neurological symptoms, masking an otherwise obvious diagnosis. This case adds to the spectrum of clinical presentations of leprosy, however medical practitioners should also remain mindful of the disease’s stigma and social implications.

References