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Abstract

For a diagnosis of polymyositis (PM) to be deemed probable or definite, affected patients must fulfil 3 or 4, respectively, of the Bohan & Peter 1975 diagnostic criteria (i.e. proximal muscle weakness, elevated muscle-specific enzyme levels, myopathic EMG and characteristic muscle histology)¹. We report here a patient who was initially misdiagnosed and treated as having PM, and whose diagnosis of sarcocystosis myopathy was only confirmed when specialist muscle histology was undertaken in a tertiary laboratory.

Introduction

Sarcocystosis, previously known as sarcosporidiosis, is a rare intracellular zoonotic protozoal parasitic infection of human beings caused by *Sarcocystis* spp, in the phylum Apicomplexa². *Sarcocystis* spp has a two-host life cycle of a sexual reproduction stage in a definite host (gametogony followed by sporogony) in the intestine of a carnivore or omnivore, and asexual multiplication stage (schizogony) in the tissue of an herbivore as an intermediate host. Human beings may serve as both intermediate and definitive hosts². Humans can be dead-end intermediate hosts for *S. nesbitti*, which is likely acquired via consumption of food or water contaminated with oocysts from snakes or monkeys³. This organism in humans will cause muscular sarcocystosis. Manifestations may be nonspecific and include fever, myalgia, and headache.

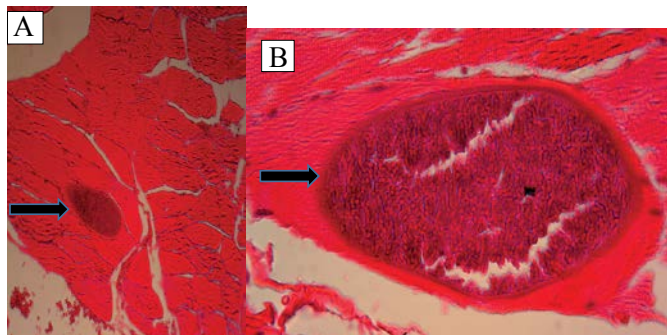
Case report

A 29-year-old previously well male labourer presented to a rural Kenyan clinic with myalgia and muscle weakness, in association with generalized malaise and fever. Examination revealed obvious proximal muscle weakness.

A presumptive diagnosis of PM was made, and he was commenced on prednisolone at daily doses ranging from 10-20mg. This gave no benefit even after five months of therapy. The patient discontinued the drug, and sought herbal medicines, again without benefit. After ten months of further continued symptoms, the patient thus journeyed to a specialist rheumatology clinic in Nairobi for a second opinion. Physical examination again revealed proximal weakness, but with no other signs of a connective tissue disease. Laboratory investigations revealed a white blood cell count of 12.6x 10⁹/L, and with an eosinophilia of 9%. It is unknown whether his eosinophil count was checked in the rural clinic. Muscle enzymes were also elevated: lactate dehydrogenase 270 U/L (NR 100-250) and aldolase 24.2 U/L (NR <7.5). Antinuclear factor was negative, and thyroid function tests normal. Neurophysiological examination was unavailable even in the Kenyatta National Hospital, University of Nairobi. As PM was still suspected muscle biopsies were obtained from the deltoid and quadriceps muscles, and corticosteroids were restarted. In view of the eosinophilia, and the possibility of a yet unproven infestation, albendazole was also recommenced, again with no improvement.

Muscle histology took a number of weeks, as the samples were sent to a tertiary laboratory in Italy, but eventually confirmed the diagnosis as one of sarcocystosis myopathy (Figure 1). The patient had meanwhile tested serologically negative for toxoplasmosis. As retreatment with steroids and albendazole was of no benefit, he was switched to high dose trimethoprim-sulfamethoxazole co-prescribed with steroids. He then recovered to normal within a matter of weeks, with normalisation of the muscle enzyme levels and resolution of the eosinophilia.

Figure 1: Cross-section of a sarcocyst in this patient's quadriceps muscle (arrow, H&E, x100) in A). The sarcocyst measures 320 μm x 180 μm and has a thin, oval, smooth cystic wall, and with numerous bradyzoites seen within the sarcocyst (arrow, H&E, x400) in B). Inflammatory cell infiltrations are conspicuously absent



Discussion

Sarcocystosis is a rare cause of human proximal myopathy, and is due to infestation by a coccidian parasite of the sarcocystis genus. It is spread by the faecal-oral route, from snakes and monkeys to pigs and cattle, and thence to man. Humans can be intermediate and/or end hosts, via accidental faecal/oral contamination by oocysts³. His initial diagnosis of PM was made in a rural clinic setting, but his chronic symptoms of fever, myalgia and proximal muscle weakness, in combination with an obvious chronic eosinophilia, widened the differential to include infectious and parasitic myopathies, including with viral infections (e.g coxsackie), trichinosis, cysticercosis, trypanosomiasis and HIV-associated myositis related to toxoplasma, cryptococcal or fungal infections. These myopathic diagnoses, and that due here to sarcocystosis, can currently be confirmed only by muscle histology, as serological testing is unavailable. Sarcocystosis has a worldwide distribution, and is generally harboured by livestock. Symptomatic and asymptomatic gastrointestinal infection occurs, with the highest prevalence actually being reported in Europe⁴. Most cases of human sarcocystosis are reported during outbreaks, e.g in Southeast Asia⁴⁻⁵, and an autopsy study of 100 Southeast Asian patients dying of other causes reported a prevalence of 21%⁶. Sarcocystosis manifestations include myositis, myalgia, localized muscular swellings, low grade fevers, weakness, vasculitis and eosinophilia⁴. Serum muscle enzyme levels are usually elevated⁴. Sarcocystosis myopathy symptoms are due to secondary inflammatory cell infiltrations, and usually composed of lymphocytes and eosinophils⁸. The incubation period for muscular sarcocystosis is 9-13 days³, initial symptoms usually lasting for a median of 17 days. They may however persist for months, as occurred in our case, or even years³⁻⁵. Sarcocystosis may also have a brief, and self-limiting course with myalgia lasting less than a week³⁻⁵. The early clinical features of

muscular sarcocystosis are non-specific, so in an African setting raise the possibility of other tropical conditions, Sarcocystosis myopathy has no specific treatment, but trimethoprim-sulfamethoxazole, clindamycin and pyrimethamine have all been used with success^{4,5}. Corticosteroids and albendazole have also been used, but with limited success^{7,8}. A reported paucity of secondary inflammatory cell infiltrations in muscle sarcocystosis⁴ may explain the limited effectiveness of corticosteroids when used alone. Proper disposal of animal and human faeces, and careful animal husbandry, are prerequisites to avoid human sarcocystosis parasitism. Public health education regarding transmission, combined with proper diagnosis, are therefore vital. If a neurophysiological examination had been undertaken here, and had shown myopathic changes, it would have been possible to misdiagnose his case as "probable PM", illustrating the crucial importance of undertaking diagnostic muscle biopsies in the African setting, to avoid missing this and other myositis mimics. Although rare, this diagnosis should clearly be considered in patients developing apparent PM after recent trips to rural Africa or Asia, and especially in the presence of an eosinophilia.

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