Chronic suppurative cryptococcal extensor tenosynovitis in a patient with Castleman's disease: a case report

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Abstract *Cryptococcus neoformans* is an ubiquitously occurring encapsulated fungus that is commonly found in the environment. It is also an opportunistic pathogen that has potential to cause systemic fungal infection, predominantly in the immunocompromised host with cell-mediated immunological defects. Cryptococcal tenosynovitis is an extremely rare condition, with only a few cases previously documented in the literature. We report a case of chronic suppurative cryptococcal extensor tenosynovitis in a patient with Castleman's disease who was successfully managed with surgical debridement and antifungal therapy.

Keywords Chronic suppurative tenosynovitis · *Cryptococcus neoformans* · Castleman's disease

Abbreviations

PET positron emission tomography
HIV human immunodeficiency virus
TB tuberculosis
MRI magnetic resonance imaging
CNS central nervous system

Introduction

Cryptococcus neoformans is an ubiquitously occurring encapsulated fungus that is commonly found in the environment. It is also an opportunistic pathogen that has potential to cause systemic fungal infection, predominantly

in the immunocompromised host with cell-mediated immunological defects. Cryptococcal tenosynovitis is an extremely rare condition, with only a few cases previously documented in the literature. We report a case of chronic suppurative cryptococcal extensor tenosynovitis in a patient with Castleman's disease who was successfully managed with surgical debridement and antifungal therapy.

Case Report

A 48-year-old, left hand dominant male presented with an 8-month history of a progressively enlarging mass on the dorsum of his left hand. The lesion initially developed as a painless small lump. Two months prior to his presentation, he noted the mass gradually increased in the size and he also developed pain and stiffness at his wrist. He controlled his symptoms with rest and anti-inflammatories. There were no neurological or vascular symptoms. He denied any recent weight loss or fevers. There was no history of previous trauma, inoculation, or previous surgery to the region.

He had a significant past medical history of unicentric Castleman's disease. This was diagnosed 5 years prior on a positron emission tomography scan which showed increased uptake in the left external iliac, right supraclavicular, and the anterior and posterior triangles of his neck. The diagnosis was then confirmed on supraclavicular node excision. He was managed conservatively without chemotherapy or radiotherapy.

The patient was born in Ghana but spent most of his childhood in London after the age of 5. He worked in the fire brigade prior to emigrating to Australia in 1996. His current occupation as a police officer did not expose him to any unusual environments. He is a smoker with moderate

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alcohol intake. There was no history of intravenous drug use, HIV, TB, sexual promiscuity, or homosexual partners. There was no significant family history.

At presentation, there was a 6×4-cm, firm, slightly irregular, tender mass arising from the dorsal aspect of the left hand, just distal to the wrist joint and overlying the extensor tendons. The mass was not tethered to the overlying skin but seemed involved with the underlying extensor tendons. It did not transilluminate and was not fluctuant. He had a notable reduction in wrist dorsiflexion compared to the contralateral side, especially with reduction of middle finger extension. Normal pronation and supination as well as radial and ulnar deviation were noted. From a systemic point of view, he had palpable lymph nodes in the right posterior triangle of his neck. Plain radiographs of his wrist and hand indicated that the mass lesion did not arise nor involve the underlying metacarpals or wrist joint.

An MRI scan revealed a lobulated soft tissue mass (50×16×10 mm) arising from the extensor tendon sheath over the wrist joint, centered in line with the middle metacarpal (Fig. 1). It was hyperintense compared to muscle on T1 images and enhanced on T2-weighted sequences. Gadolinium scan revealed moderate enhancement with a distal area of cystic or necrotic change. There was also generalized synovitis of the tendon sheath of the fourth dorsal compartment. From these findings a provisional diagnosis of a giant cell tumor (GCT) arising from the extensor tendon sheath was made.

A decision to perform an excisional biopsy was made. This was performed under general anaesthesia using a tourniquet. An extensile approach to the dorsum of the wrist was utilized via a longitudinal incision over the fourth dorsal compartment (Fig. 2). A complex tumor-like mass was encountered and seen to be very adherent to or possibly arising from the tendon sheath of the fourth compartment. The mass was removed en bloc, following careful dissection of the extensor tendons which had been partially encapsulated by the tumorous mass. The distal portion of the tumor was inadvertently opened, and this released a necropurulent discharge, which seemed more consistent with possible infection rather than GCT. The fluid and the mass were sent for microbiological and histopathological analysis. The wound was closed in layers with meticulous hemostasis. The wrist was immobilized in a resting splint for 14 days, and then a gentle home-based range of motion program was commenced.

Microbiology from the fluid demonstrated polymorphs, but the gram stain and culture results were negative. The Ziehl–Nielson stain was negative for acid-fast bacilli as were the cultures for tuberculosis.

The histopathology result revealed a necrotizing suppurative granulomatous inflammation with identifiable fungal

Fig. 1 MRI scan showing the characteristics and location of the mass. **a** Proton density weighted coronal. Note the mass overlying the capitate. **b** T2 axial. Note the relationship of the extensor tendons to the dorsal mass

spores and associated chronic synovitis and reactive synovial hyperplasia (Fig. 3). Further testing and staining revealed these spores to be consistent with *Cryptococcus neoformans* infection. This result was confirmed by pathologists at two different centers.

The patient was then referred to an infectious disease clinic in a tertiary hospital for review and further investigation into a possible immunodeficiency. The results of his HIV and TB testing were negative. Lumbar puncture for cryptococcal meningitis was negative.

He was commenced on high-dose fluconazole therapy, 400 mg orally bd, for a period of 3 months. Fluconazole is a triazole antifungal drug used in the treatment and prevention of superficial and systemic fungal infections. He is currently being investigated for an occult immunodeficiency. At 12-month follow-up, the patient has normal function of his left hand and wrist with no identifiable recurrence of disease.

Fig. 2 Intraoperative photograph demonstrating the size and location of the mass and its proximity to the extensor tendons. Phalanges are to the left of the picture

Discussion

C. neoformans infection is an opportunistic systemic fungal pathogen found predominantly in immunodeficient patients [1, 2, 6]. Common sites of infection include respiratory, CNS, skin, and bone. Cryptococcal tenosynovitis is a very rare manifestation of the disease. In 2002, Bruno et al. reported a case of cryptococcal flexor tendon tenosynovitis and carpal tunnel syndrome, but in the literature, extensor tendon involvement has not previously been reported.

Castleman's disease was first described in 1956 by Benjamin Castleman. It is a benign, hyperplastic condition affecting the lymph nodes and has also been referred to as angiofollicular hyperplasia. Like cryptococcal infection, it

Fig. 3 Histopathological photograph showing a hematoxylin and eosin stain showing the presence of chronic granulomatous inflammation (LI lymphocytic infiltration, GC multinucleate giant cells)

Fig. 4 Histopathological photograph showing a Giemsa stain demonstrating the cryptococcal spores (CS)

is often associated with immunodeficiency syndromes [3, 4]. Castleman's disease is a non-clonal disease of the lymph nodes which is classified by the following: (a) unicentric or multicentric, based on clinical and radiological findings; (b) hyaline vascular vs. plasmacytic vs. mixed cellularity variety based on histopathology; and (c) HIV status of the patient.

Unicentric Castleman's disease is characterized by a slow-growing solitary mass typically in the mediastinum or mesenteries. There are no constitutional symptoms and normal acute phase reactants. Symptoms are caused by a mass effect of lymphadenopathy, and in the majority of cases, surgical resection is curative and usually there is no progression to lymphoma or association with other tumors. The multicentric type especially with an HIV-positive status is much less clinically favorable. This case is interesting, because while infection is always a possible aetiology, the chief differential diagnoses of a progressively enlarging mass on the dorsum of the wrist include ganglion, GCT of the tendon sheath, lipoma, or neurofibroma.

The MRI demonstrated an appearance consistent with GCT of the tendon sheath, and therefore the decision was made to proceed to excision rather than biopsy. However, intraoperatively, an area of necrosis accompanying purulent discharge was encountered while dissecting the distal margin of the mass off the extensor tendons. This is an atypical feature of GCT. At this point, it was clear that an infective aetiology was responsible for this mass.

Given the patient's history and background, TB was considered as a highly probable pathogen and this has been widely reported in the literature [5, 9]. However, he had no previous contacts and all subsequent investigations were negative. The unlikely diagnosis of cryptococcal infection was made via histopathology (Fig. 4).

Because of the high neurotropism of *C. neoformans*, CNS involvement is always a possibility, particularly in disseminated disease [1, 2, 6, 8]. However, in this case, the lumbar puncture was negative and the cryptococcal immunoglobulin titre was relatively low. While these investigations may only be positive 25–50% of the time, even in an immunocompetent patient, it seems likely that this case represents a localized rather than a disseminated systemic manifestation of the disease. Oral fluconazole has been reported as effective treatment of cryptococcal infections [7, 8], and he was commenced on this as recommended by the infectious diseases unit.

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