Open Journal of
Clinical & Medical
Case Reports

Volume 2 (2016)
Issue 10

ISSN 2379-1039

Aggressive Rheumatoid Arthritis Presenting as an Unusual Carpal Tunnel Syndrome

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Abstract

A 56 year old female underwent bilateral carpal tunnel decompression. The more symptomatic side was decompressed successfully with an unremarkable postoperative course. However, contralateral decompression required intraoperative induction of general anaesthesia to complete the resection of an unexpected soft-tissue lesion.

The mass was histologically consistent with a rheumatoid nodule. Interestingly, the patient had been thoroughly investigated by specialist rheumatologists over a period of years preceding her attendance at the orthopaedic clinic. There had not been definitive clinical or biochemical evidence of rheumatoid arthritis, other inflammatory arthritis, or connective tissue disease at these reviews. However, an aggressive phenotype of rheumatoid arthritis has subsequently developed with multiple joint involvement and strongly positive serological markers.

Carpal tunnel decompression is often performed under local anaesthetic by junior surgeons or trained general practitioners. However, unusual cases such as ours do occur and it is vital that experienced surgical and anaesthetic cover is available. Furthermore, this case reaffirms that carpal tunnel syndrome due to a space-occupying lesion can be a presenting feature of rheumatoid arthritis. Previous negative investigations do not preclude evolution of the disease.

Keywords

carpal tunnel syndrome; rheumatoid arthritis; rheumatoid, carpal tunnel decompression; hand surgery; tenosynovitis

Abbreviations

CTS: Carpal Tunnel Syndrome; RA: Rheumatoid Arthritis; ESR: Erythrocyte Sedimentation Rate; ANCA: Anti-Neutrophil Cytoplasmic Antibody; ANA: Antinuclear Antibody; Anti-CCP: Anti-Cyclic Citrullinated Peptide Antibody; SAP: Scintigraphy-Serum Amyloid P Scintigraphy; SOL: Space Occupying Lesion.

Introduction

Carpal tunnel syndrome (CTS) is the most common upper-limb compressive neuropathy. It peaks in the 4th and 5th decades, with a prevalence of 9.2% in women and 6% in men [1]. The aetiology is compression of the median nerve at the carpal tunnel, which transmits the four tendons of flexor digitorum superficialis, the four of flexor digitorum profundus, and flexor pollicis longus in addition to the nerve.

The clinical syndrome is usually described as pain and paraesthesiae in the median nerve distribution of the hand, with or without weakness of the muscles it supplies. In reality, it is more common to experience paraesthesiae in all of the digits rather than the median digits alone, [1-2] due to the communications amongst the digital nerves at the Berretini anastomosis [3]. In addition, the skin overlying the thenar eminence is often spared as the palmar cutaneous sensory branch is usually given off proximal to the carpal tunnel [1]. A final clinical point to be borne in mind is the prevalence of symptoms proximal to the carpal tunnel, most commonly in the forearm but as far proximally as the neck [1].

The majority of cases are idiopathic. However there are numerous potential secondary causes. A useful model is to consider them as abnormalities of the carpal tunnel itself (such as in traumatic displacement of the distal radius) or abnormalities of those structures which traverse it (such as tenosynovitis) [4]. There are also a number of systemic conditions associated with CTS, including hypothyroidism, diabetes and rheumatoid arthritis amongst others [5]. There is a recognised link with various occupational factors, [6-7] though these may be best considered provocative rather than causative [8].

The microscopic pathophysiology is complex. There is compromise of the capillary plexus formed by the vasa nervorum, with venocongestion and local oedema leading to ischaemia and local metabolic complications [4,8-9]. Disruption of the transmembrane ion pumps and axonal transport result in pain and paraesthesiae [4,8-9]. When the wrist is repositioned, or the fingers (and therefore their flexor tendons) moved, this oedema may disperse thus providing relief from the symptoms [4]. In more advanced disease the microcirculation cannot be restored leading to demyelination and/or axonotmesis [4].

Case Presentation

A 56 year old patient underwent nerve conduction studies for intractable neuropathic pain in the hands. Bilateral carpal tunnel syndrome was confirmed, 'moderately-severe' on the right and 'mild' on the left. Referral to an orthopaedic surgeon was made. On presentation at the orthopaedic clinic there were no atypical features in either hand and the clinical suspicion was of uncomplicated CTS. The patient was listed for decompression accordingly. The right side was decompressed under local anaesthesia with good results and an unremarkable post-operative course. As a result, the patient elected for contralateral decompression and attended as a day case.

The hand was prepped and infiltrated with local anaesthetic. The transverse carpal ligament was identified and divided under direct vision without complication. A small protrusion at first suspected to be a small haemangioma was noted amongst the flexor tendons and careful dissection was commenced in order to divide it at its source. However, as the dissection continued this lesion was noted to be more extensive than first presumed. The incision was extended in a step-wise fashion as the greater dissection revealed an extensive, highly-vascularised soft tissue mass. At this point, the patient was beginning to experience severe discomfort from the tourniquet and despite subsequent doses as the incision was extended, local anaesthesia proved inadequate. Due to the extended invasion of the lesion and its malignant appearance, we decided to convert to general anaesthesia intraoperatively in order to continue the procedure.

The findings were of a large lesion enveloping the flexor tendons and invading the epineureum of the median nerve. Proximally, it extended to the musculotendinous junction of flexor digitorum superficialis/profundus. The profundus slip to the index finger had ruptured (Figure 1). Distally, two extensions to the incision were performed: one to the junction of flexor zones 2 and 3 of the index finger and a second into zone 2 of the fifth finger where a second, macroscopically distinct nidus of the lesion was located (Figure 2).

The mass was carefully excised with tenosynovectomy of the affected tendons and epineurolysis of the median nerve. The ruptured profundus slip to the index finger was transferred to its equivalent from flexor digitorum superficialis, and the ruptured tendon of flexor digiti minimi was repaired. The carpal tunnel walls appeared eroded and these were curetted. Samples were sent for urgent histology as malignancy was strongly suspected and the patient admitted overnight as a precaution given the vascularity of the mass. The histology was not suspicious of malignancy, describing a reactive process with palisading histiocytes consistent with a rheumatoid nodule.

Interestingly the patient had been comprehensively investigated by rheumatology for arthralgia and chronic pain over a several-year period prior to referral to orthopaedics. Her initial presentation was 6 years previously, with progressive 'burning' foot pain. She had no tender or swollen joints and no extrarticular manifestations of inflammatory arthritis or connective disuse disease. The erythrocyte sedimentation rate (ESR) was noted to be raised at 86mmhr^{-1} with a normal C-reactive protein (CRP). Over the following 18 months she developed pain involving numerous other joints, though they remained non-tender and not clinically swollen.

She was subsequently investigated with radiographs of the hands and feet, rheumatoid factor, antineutrophil cytoplasmic antibodies (ANCA), antinuclear antibody (ANA), serum electrophoresis, anti-cyclic citrullinated peptide (anti-CCP) antibody titre, and serum amyloid P component (SAP) scintigraphy. A trial of intramusclular dexamethasone was attempted without benefit. All of these investigations were normal.

Her symptoms continued to worsen and therefore the autoimmune profile was repeated (at 3.5 years preoperatively). A computed tomography (CT) scan of the chest, abdomen and pelvis was also performed to rule out malignancy with a paraneoplastic syndrome. These investigations remained normal with the exception of mildly raised β2-microglobulin anti-cardiolipin antibody titre. The rheumatological opinion remained that there was no evidence of rheumatoid or other inflammatory arthritis.

In contrast, repeat rheumatological review post-resection demonstrated strong biochemical evidence of RA. Anti-CCP antibodies were strongly positive at >300 U/ml and rheumatoid factor had also become positive at 626.6IU/ml (both were last measured 3.5 years previously). There was also now clinical evidence of RA: her DAS28 disease activity score at a second postoperative review 4 months later was 6.87, with 18 tender and 16 clinically swollen joints.

An aggressive phenotype of RA is suspected. In the weeks following the procedure the patient complained of a new pain in her foot. Subsequent MRI demonstrated aggressive tenosynovitis presumed to be similar to the lesion excised from her wrist involving the tendon of tibialis anterior, as well as a soft tissue lesion in the heel consistent with a rheumatoid nodule (Figure 3).

Discussion

Space-occupying lesions (SOLs) such as in this case are a recognised cause of CTS, representing 3% of surgically-managed CTS in one case series [10]. It seems unclear from the literature how commonly there is a palpable mass when CTS is secondary to SOL: in two case series this figure varied widely, between 35 and 70% [10-11]. In the former series all patients with underlying tenosynovitis had diffuse swelling, tenderness, erythema and raised C-reactive protein (CRP). However in our case there was only a slightly raised erythrocyte sedimentation rate (ESR).

A case with a number of similarities to ours is described by Iyengar *et el*. [12] The patient presented with bilateral CTS, flexor tenosynovitis on ultrasonography, and normal laboratory values other than raised ESR. They performed tenosynovectomy and epineurolysis, though in contrast to our case noted a number of rice bodies amongst the flexor tendons. The histology was similar, with areas of fibrinoid necrosis and pallisading histiocytes.

The majority of carpal tunnel decompressions are uneventful and can be performed as day cases under local anaesthetic, with a huge potential benefit to efficiency and cost-effectiveness [13]. However, we believe that our case demonstrates that that experienced anaesthetic cover should be available as intraoperative conversion to GA can become indicated. In addition, in the case of general practitioners or more junior surgeons performing decompressions, an experienced surgeon comfortable with operating within the hand and wrist should be available. In our case, without intraoperative conversion to GA and extensive dissection including a number of flaps across the palm, extensive tenosynovectomy and median epineurolysis, then there may have been potentially severe post-operative ramifications with bleeding from this unexpected, invasive, and vascular tenosynovitis. Additionally, without prompt recognition and repair of the two ruptured flexor tendons, there may have been functional impairment and indication for re-operation.

In addition, this case demonstrated an unusually aggressive presentation of rheumatoid arthritis. Although RA is a common aetiology of CTS, which is in turn one of its most prevalent extra-articular features, [14] in this instance carpal tunnel syndrome heralded the onset of an aggressive phenotype of RA with strongly positive serum markers and rapid progression to multiple joint involvement and further foci of invasive tenosynovitis. This occurred in the context of a patient who had been extensively investigated by rheumatology previously with neither clinical nor biochemical suspicion of active rheumatological disease. This case demonstrates that RA should remain a differential diagnosis in these circumstances.

Learning Points

Previous negative investigations do not preclude evolution of rheumatoid arthritis and this fact should be borne in mind when there is significant suspicion, with appropriate re-referrals made and serological tests repeated.

Experienced anaesthetic and surgical cover should be available if necessary for all carpal tunnel lists. Though the majority are uneventful, unexpected and difficult-to-manage situations do occur.

Carpal tunnel syndrome due to space-occupying lesions within the tunnel can be a presenting feature of rheumatoid arthritis.

Figures

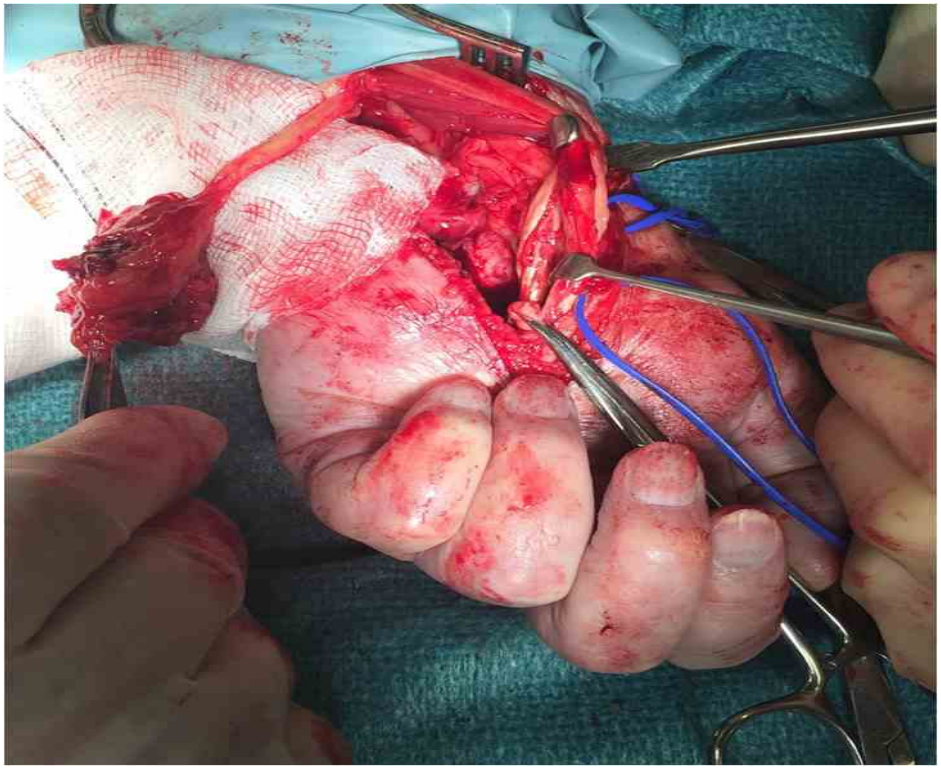

Figure 1: A large portion of the mass was found to be tethered to the flexor digitorum profundus slip to the index finger, which was noted to have ruptured as a result.

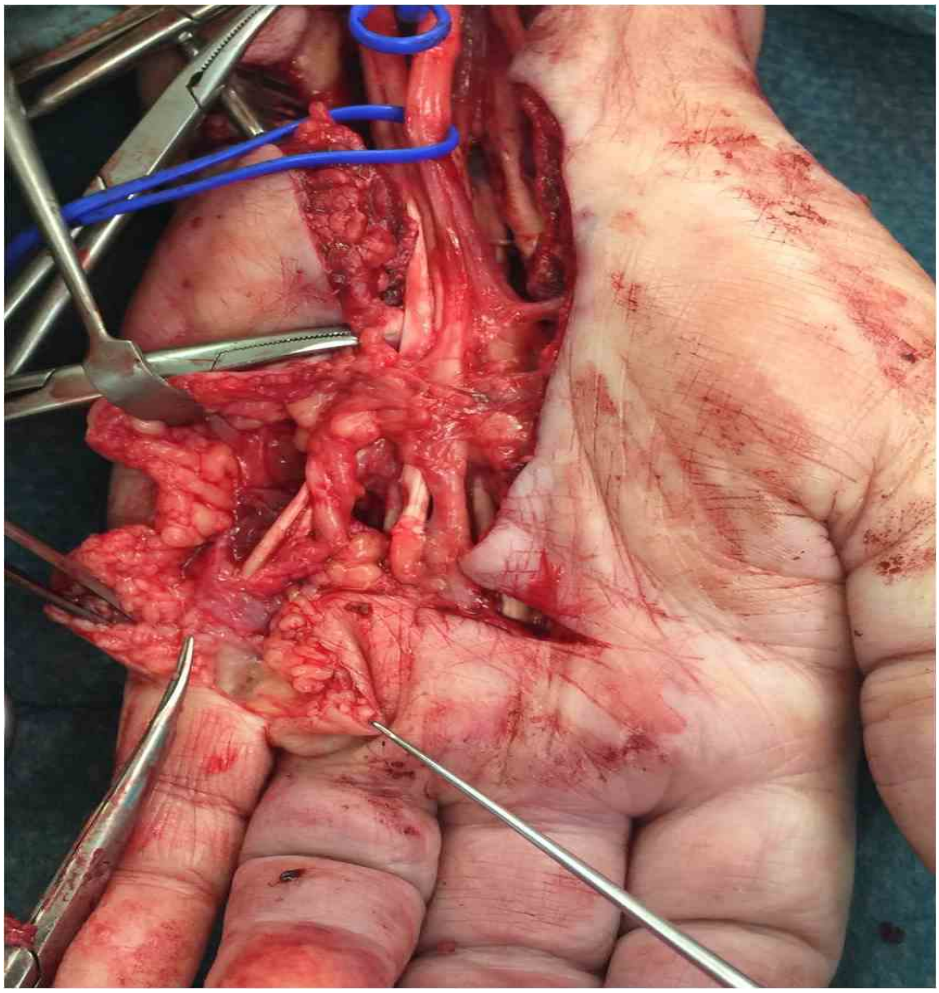

Figure 2: Appearances of the palm dissection after a second nidus of the mass was removed from close to the fifth finger.

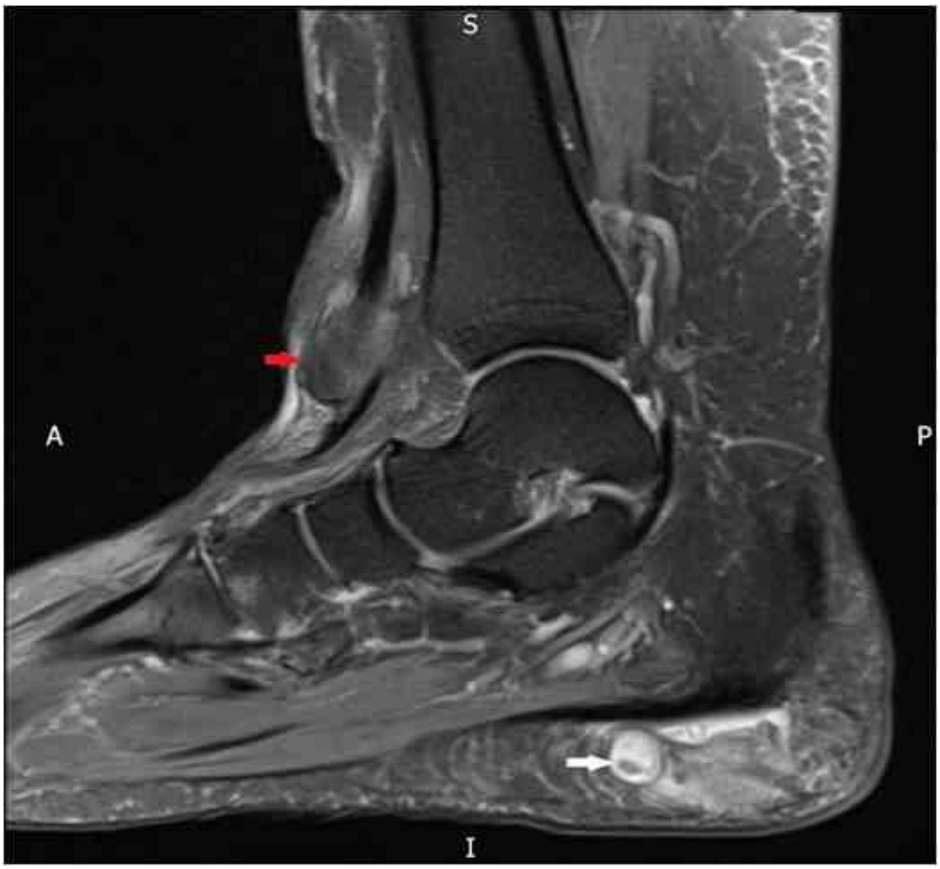

Figure 3: Sagital section from a contrast MRI demonstrating an area of abnormal thickening about the tibialis anterior (red arrow), presumed similar in character to the tenosynovitis encountered at the carpal tunnel. A second lesion consistent with a rheumatoid nodule is also noted adjacent to the plantar fascia (white arrow).

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Manuscript Information: Received: March 14, 2016; Accepted: May 26, 2016; Published: May 29, 2016

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Citation: Borton Z, Vratchovski V. Aggressive rheumatoid arthritis presenting as an unusual carpal tunnel syndrome. Open J Clin Med Case Rep. 2016; 1120

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