Localised Pigmented Villonodullar Synovitis; A Rare Cause of Carpal Tunnel Syndrome

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Citation

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Abstract

We report a case of localised pigmented villonodular synovitis that presented as a ganglion-like lesion on the volar aspect of the wrist causing symptoms of carpal tunnel syndrome.

INTRODUCTION

First described by Jaffe et al in 1941, PVNS is a relatively uncommon benign disorder of unknown etiology that is characterized by progressive synovial proliferation. Benign, but locally aggressive, it is found most commonly in the knee and other large joints. PVNS affecting the carpal tunnel is extremely rare, and in our opinion suitable for reporting.

CASE REPORT

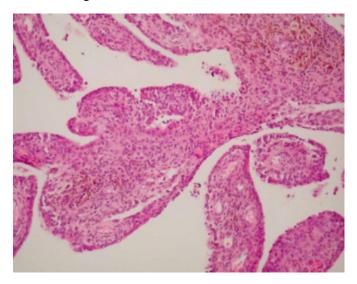
A 45-year old right-handed lady presented to the orthopaedic clinic with a swelling over the volar aspect of her left wrist associated with tingling and numbness in her thumb, index and middle fingers. The symptoms had been present for a year and were getting worse, and she had noticed weakness of grip and a tendency to drop objects from the left hand.

Examination revealed a 1cm x 1.5 cm firm non-tender swelling overlying the wrist just distal to the insertion of the Palmaris Longus tendon. Clinically, it had characteristic ganglion-like features. She also had evidence of reduced sensation in the median nerve distribution, and a positive Phalens test within 10 seconds. There was mild weakness of opposition. Nerve conduction studies confirmed the diagnosis of carpal tunnel syndrome of moderate severity.

She was posted for excision of the ganglion and carpal tunnel decompression. At surgery, the swelling was found to be thickened brown villous synovium arising from the tendons of the flexor digitorum superficialis. It extended into the carpal tunnel and was causing obvious compromise of the tunnel space, producing the symptoms of median nerve compression. The nerve was intact without any other obvious pathology. The synovium was excised completely to free the nerve, and the incision was extended proximally into the distal forearm to allow this. The entire specimen measured 10x5x3 cm. Histopathology confirmed the diagnosis of pigmented villonodular synovitis (Figure 1).

Figure 1

Figure 1: Histopathological appearance of pigmented villonodular synovitis, showing scattered osteoclast-like multinucleated giant cells and Large-sized mononuclear histiocyte-like cells containing a peripheral rim of hemosiderin granules



The wound healed without any complications and she recovered full function with physical therapy at five months after surgery. She remains asymptomatic and recurrence-free at a 2-year follow-up.

DISCUSSION

Pigmented villonodular synovitis is a benign proliferative disorder of uncertain etiology, despite the high risk of

recurrence ₁. Several reports of this condition have been published since Jaffe, Lichtenstein and Sutro first described it in 1941. It affects synovial lined joints, bursae, and tendon sheaths.

Clinically, there are two types of growth patterns: the localized or nodular type, which is a solitary nodule, and the diffuse type, which is a villous, pigmented process involving the synovial tissue and eradication can be more difficult in large joints than in digits in patients with diffuse disease rather than single nodules ₂ . In large joints, giant cell tumours can be more difficult to diagnose, as the symptoms are non-specific and the signs far and few. The joint most affected is the knee. In the digits, they are usually well-circumscribed multinodular subcutaneous masses which are often painless, but increase in size over short period of time and raise concerns.

Large joints affected include the ankle, knee, wrist and the elbow $_3$. The soft tissue mass grows and expands into areas of least resistance. X-rays are often unhelpful, but may show bone erosions or a soft tissue swelling occasionally.

Sir James Paget first reported median nerve compression at the wrist in 1854 following a distal radius fracture. Direct pressure or a space occupying lesion within the carpal canal can increase pressure on the median nerve and produce carpal tunnel syndrome. Patients with CTS averaged pressures of 32mmHg compared to 2mmHg in control subjects. The safe threshold beyond which median nerve dysfunction may occur has been identified as 40 to 50 mmHg₄. In the past, there has been only one case report of acute carpal tunnel occurring as a result of haemorrhage into the carpal tunnel from a pigmented villonodular synovitis. A second case of pseudo-carpal tunnel syndrome has been reported due to localised pigmented villonodular synovitis 7. Other uncommon causes include hamartomas, tuberculous synovitis, ganglia, arteriovenous forearm shunts and fractures of the hook of the hamate.

Preoperative MRI is an important diagnostic tool. MRI shows a typical image of the lesion because of hemosiderin deposition, which leads to low to intermediate signal intensity on T1-weighted and low signal intensity on T2weighted images $_8$. Preoperative magnetic resonance imaging results corresponded well with histological diagnoses and intraoperative findings⁹.

In our case, the diagnosis of PVNS was not considered in the first instance and the swelling was presumed to be a ganglion producing compressive neuropathy. However, intra-operative findings were found to be more extensive and typical of PVNS. The report highlights the fact that one must not forget the unusual causes of carpal tunnel syndrome. Although rare, these can often be overlooked. They must be considered in cases of carpal tunnel syndrome with an atypical presentation.

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