Singapore Med J 2003 Vol 44(3) : 152-154

Extraarticular Pigmented Villonodular Synovitis of the Distal Forearm: A Case Report

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ABSTRACT

Extraarticular pigmented villonodular synovitis (PVNS) is very rare in the distal forearm[1]. There has only been one previous case report of this disease in the extensor tendons of a child[2]. We report a case of PVNS of the distal forearm that presented as two nodules over the radial aspect and a separate nodule on the ulnar aspect beneath the flexor carpi ulnaris tendon. Surgical exploration revealed an extensive extraarticular PVNS over the first and second dorsal compartment extensor tendons. On the anterior aspect it extended in the deep plane between the flexor tendons and the pronator quadratus and encased the radial artery completely. Complete excision of the tumour with the radial artery was done.

Keywords: Giant Cell Tumour of tendon sheath

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CASE REPORT

A 43-year-old Chinese woman presented with three lumps over the right distal forearm for four months duration. They were painless and had grown in size. A Chinese physician had pierced the radial lump with a heat sterilised sewing needle which resulted in a superficial abscess that drained spontaneously. Healing of the wound had left her with a transverse scar measuring 1 cm over the radial lesions.

Physical examination revealed three lumps at the distal forearm. There were two 1 x 1 cm discrete lumps over the radial aspect, one on each side of the first dorsal compartment of the hand. There was another 1 x 1 cm lump over the ulnar aspect emerging deep to the flexor carpi ulnaris tendon. The lumps were non-tender, well circumscribed and soft in consistency. The preoperative diagnoses were ganglions for the radial lumps and a lipoma for the ulnar lump. X-rays did not reveal any bony erosions of the radius or ulna.

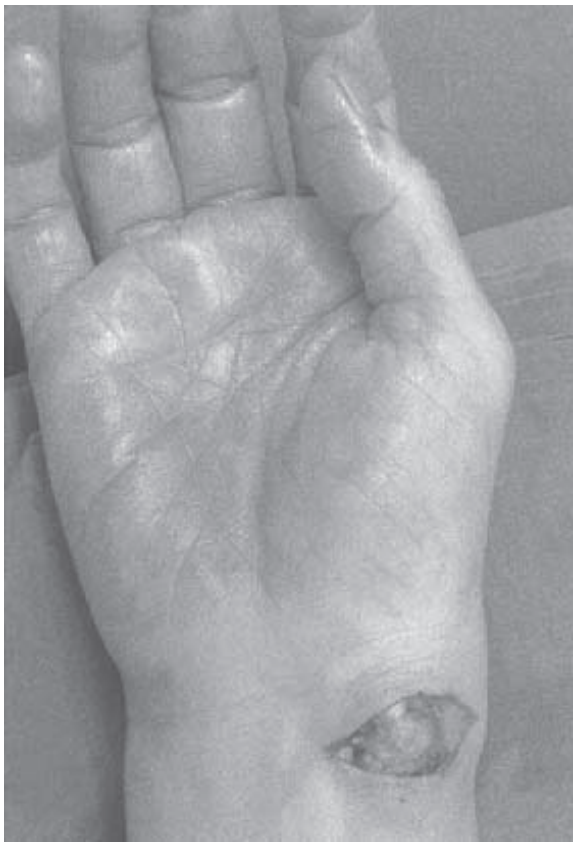

Fig. 1 Intraoperative photograph of the radial view of the distal forearm.

The exposure was via an incision over the radial scar (Figs. 1 and 2) and an ulnar incision along the flexor carpi ulnaris. The intraoperative diagnosis of PVNS was made when the lesion was found to be lobulated and yellowish in colour (Fig. 3). The lesion was found to extend dorsally across the first and second extensor compartment and anteriorly in the plane between the flexor digitorum profunda tendons and the pronator quadratus muscle. The lesion was also found to encase the radial artery and was inseparable from it.

Treatment involved complete excision of the tumour including the encased segment of the radial artery. Assessment of adequacy of circulation to the hand was performed intraoperatively using the Allen test.

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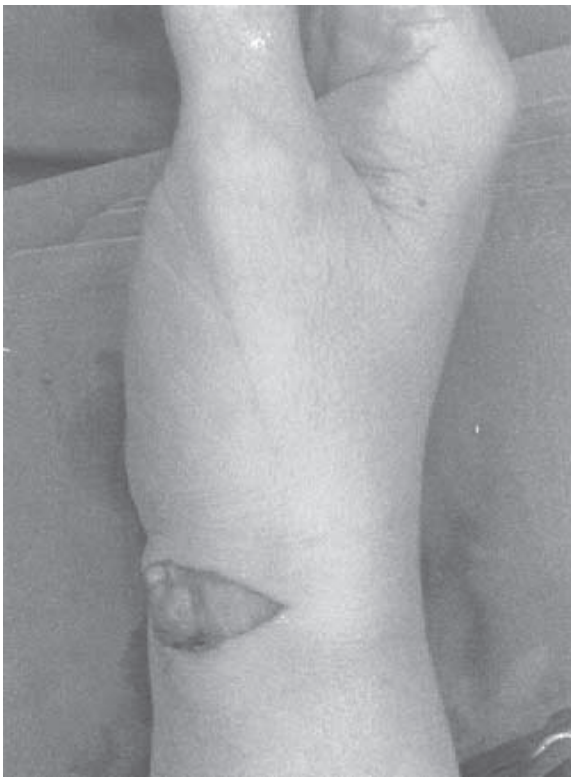

Fig. 2 Intraoperative photograph of the volar view of the distal forearm.

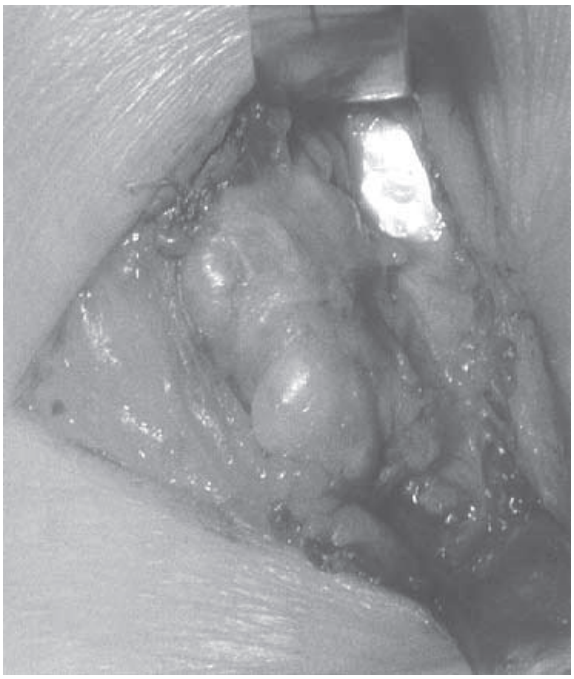

Fig. 3 Intraoperative photograph of the close-up of the lesion.

Table I. X-ray findings in extraarticular pigmented villonodular synovitis.

I	Normal findings
II	Minimal changes: synovitis, an expanded synovial sac, superficial osseus erosion and cysts
III	Extensive involvement: diffuse non-calcifying synovial tumefactions of lobular outline and varying density

The lesion measured 4 x 3 x 2 cm in size and the histology was consistent with that of PVNS.

DISCUSSION

Pigmented villonodular synovitis is classified into three prevailing types by Jaffe[3], namely pigmented villonodular synovitis (PVNS), pigmented villonodular bursitis (PVB), and pigmented villonodular tenosynovitis (PVTS). Granowitz[4] further subclassified this into two distinct clinical forms identified by the prefix: L for a sharply localised or pedunculated lesion and D for diffuse involvement of the synovial membrane.

The pathogenesis of PVNS is controversial[1]. The strongest correlation is with chronic trauma and repeated hemarthrosis. However, the most widely accepted cause is that of a reactive or regenerative hyperplasia associated with an inflammatory process[3]. Other causes include a benign neoplastic process[1] and a localised disturbance in the metabolism of lipids[5].

Macroscopically, the tumour is multilobulated, fairly well-circumscribed and subcutaneous. If it is large, there may be satellite lesions extending into the tendon sheath or synovium[6]. The colour of the tumour varies from yellow to red-brown in colour and is due to the proportion of haemosiderin, collagen and quantity of histiocytes within the stroma. Involvement of the joint may also cause xanthochromic pigmentation of the synovial fluid

Histologically the lesion is characterised by a fibrous stroma, deposition of haemosiderin, histiocytic infiltrate, and giant cells occurring in the synovial membrane of tendon sheaths and large joints.

Clinically, the majority of patients present with a painless palpable mass which gradually increases in size. Signs and symptoms of the tumour are mainly due to pressure and mass effect. Involvement of the nerve may give rise to pain and numbness, and involvement of the flexor tendons of the hand may cause triggering of fingers.

Various methods of investigation have been described. X-ray findings are categorised into three classes (Table I). Magnetic Resonance Imaging (MRI) is most useful because it can reveal the size and involvement of the tumour. Fine Needle Aspiration Cytology (FNAC) has been found to correlate well with the final diagnosis[7,8].

This lesion must be treated by complete resection to prevent tumour spillage and seeding of surrounding tissues. In addition, we advocate careful exposure of the tumour, working more on adjacent tissues than on the lesion itself. The entire lesion must be traced to detect satellite nodules, which must be removed together with the main lesion,

to prevent recurrence[5]. The synovial origins of these tumours must be appreciated and partial excision of the sheath or joint capsule may be required to ensure complete removal of the lesion.

The mean interval to recurrence is $4^1/_2$ years[9]. Rates of recurrence range from 9% to 22%[10,11] and is caused chiefly by inadequate surgery. Other risk factors include increased immature cells and mitotic activity on histology[1], presence of adjacent degenerative joint disease, location at the distal interphalangeal joint of the finger or interphalangeal joint of the thumb, and radiographic presence of an osseous pressure erosion[12]. Treatment of recurrent disease consists of total synovectomy, arthroplasty, arthrodesis or a course of external beam radiotherapy for unresectable lesions[13].

CONCLUSION

Extraarticular pigmented villonodular synovitis is a rare tumour that should be treated by complete excision with special care to prevent tumour seeding.

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