Multifocal pigmented villonodular synovitis in a child

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ABSTRACT

Pigmented villonodular synovitis is a rare benign disorder and usually affects young and middle-aged adults. It occurs either as a localised pedunculated form or more common diffuse form, and is almost always unifocal. Only few cases of multifocal involvement have been reported, all of them in children. Multifocal pigmented villonodular synovitis occurring in a 5-year-old girl is presented. Both her knees and left elbow were involved. All three lesions were of localised pedunculated type and were completely excised. She is believed to be one of the youngest patients reported.

Keywords: juvenile joint disease, multifocal pigmented villonodular synovitis, pigmented villonodular synovitis, synovitis

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INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a rare benign disorder of uncertain nature. Both inflammatory and neoplastic pathogeneses have been suggested for this condition[1]. Its incidence is said to be 1.8 patients per million population. It tends to occur in young adults, with a slight female predominance. PVNS almost always involves only one articulation, most commonly the knee joint, but also affects other major joints, even the intervertebral and temporomandibular joints. Extra-articular forms also exist. The minority of PVNS occurs as a localised pedunculated brown nodule, and a majority as a diffuse type involving the entire synovial surface. The diffuse type tends to recur in one-quarter of cases.

Only few cases of multifocal involvement of PVNS have been reported, all of them in children[2-9]. Bone erosion and malignant transformation have been reported[10,11]. Findings of trisomy 7 and some other chromosomal abnormalities suggest a clonal nature[12]. Open or arthroscopic simple excision for local forms and total synovectomy sometimes followed by cryotherapy or radiation for diffuse forms are current treatment options[13,14]. Early surgical therapy is the only recommended curative intervention. The decision regarding the surgical approach, arthroscopic versus open, depends on the form of PVNS, the extent of the disease and secondary changes of the joint[14]. We present a case that is unusual because of the patient's young age and involvement of both knees and one elbow.

CASE REPORT

A 5-year-old girl presented with pain in both knees. She had experienced increasing pain for the past five months. Her pain had not responded to analgesics prescribed in outpatient setting in another clinic. Her family history was unremarkable. On physical examination, both joints were mildly swollen with some limitation in extension. Circumference of both knees and thighs was equal. No scar was present. A swollen left elbow was also noted. There was mild warmth of overlying skin but no redness or fever was detected. Fluctuation test for patella and tests for muscular power, joint stability and gait were normal. No congenital anomaly was detected on thorough physical examination.

Radiographs showed a mild soft tissue swelling and slight articular space widening on both knees. The elbow and chest radiographs appeared normal. Her leukocyte count was 8.7 x 10^9/L (69% neutrophils, 26% lymphocytes, 5% monocytes,), haemoglobin level was 13.1 g/dL, mean cell volume was 89dL, and erythrocyte sedimentation rate was 12 mm/hr. Rheumatoid factor was negative, and prothrombin time (PT), partial thromboplastin time (aPTT) and clotting time were normal. The fluid obtained by needle aspiration of knee joints was bloody and microscopical examination showed erythrocytes. Her tuberculin test induration was only 4mm. After two days of bed rest, her severity of pain did not change and there was still no fever or skin redness.

The right knee was explored and a focal pedunculated 0.7 x 0.4 x 0.2 cm nodule was found suspended from the roof of the articular space. This was easily excised. The joint fluid was red-brown. With this finding, the other joints were opened and a similar but larger nodule was found in suprapatellar

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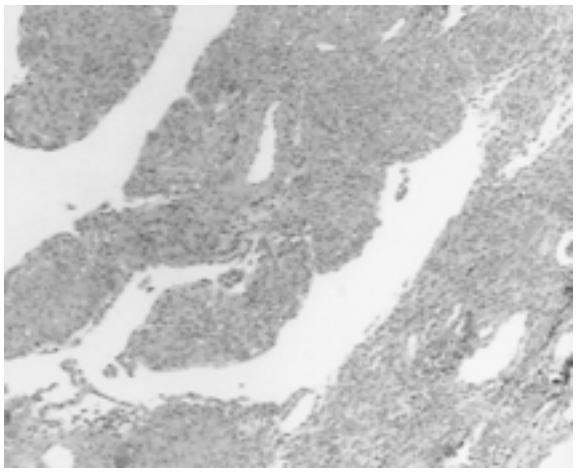

Fig. 1 Low power photomicrograph shows villous structure of the lesion (haematoxylin & eosin, x100).

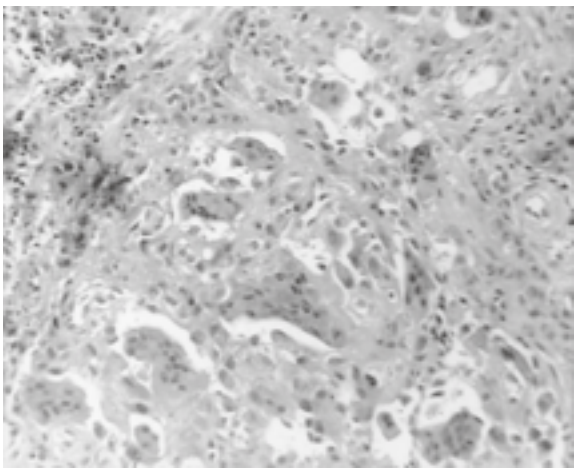

Fig. 2 Mid power photomicrograph reveals scattered multinucleated giant cells among histiocyte-like cells in a fibrous background (haematoxylin & eosin, x400).

pouch of the left knee. This measured 0.9 x 0.5 x 0.4 cm, and a 0.6 x 0.5 x 0.3 cm nodule was present in the elbow. All masses were completely excised, and had velvety surfaces with variegate pink-brown granular cut surface. On microscopical examination, there were villous structures lined by proliferated synoviocytes on a population of histiocyte-like cells, with oval coffee bean nuclei and haemosiderin-laden macrophages in a fibrous stroma (Figs. 1 & 2). Many foam cells and scattered giant cells were seen. Cells showed no atypia but rare mitotic figures were found. Diagnosis of multifocal pigmented villonodular synovitis was made.

DISCUSSION

Pigmented villonodular synovitis is a benign proliferative disease of the synovium that affects joints, bursae or tendon sheaths. Its aetiology is not completely understood, with reactive tissue growth, inflammatory origin and neoplastic pathogeneses being suspected. The clinical symptoms of PVNS are nonspecific. The patient usually suffers from restriction of joint movement, accompanied by intermittent pain, joint locking and swelling. Macroscopically, PVNS can present as a diffuse or nodular form. The histological features are a fibrous stroma covered by hyperplastic lining synoviocytes, presence of histiocytes containing haemosiderin, and multinucleated giant cells.

Radiographs can be normal or show an intra-articular effusion, para-articular soft tissue dense mass and in long-standing disease, juxtaarticular bone erosions. Computed tomography shows evidence of haemosiderin and fat deposits, and can detect bone erosions not evident on the radiographs. Magnetic resonance imaging demonstrates synovial effusion and hyperplastic synovitis containing scattered areas related to haemosiderin deposits that show low signal on both T1- and T2-weighted sequences. Since less haemosiderin is present in localised PVNS, signal intensity is more likely to be intermediate between that of diffuse PVNS and that of skeletal muscle. Although similar findings can be observed in hamophilia and rheumatoid arthritis, these features are considered to be highly suggestive of PVNS.

In a patient presenting with knee swelling, one should consider a varity of diagnoses, such as septic arthritis, haemarthrosis, rheumatoid arthritis, and intra-articular or juxta-articular masses. In our case, chronicity, lack of fever and normal PT and aPTT rule out septic arthritis as well as haemarthrosis as does a dry aspiration of the joint. Low erythrocyte sedimentation rate makes active rheumatoid diseases unlikely. Differential diagnosis of a focal expansile lesion at the anterior region of the knee joint includes the more frequently seen meniscal and cruciate ligament cysts, and the less common intra-articular chondroma, synovial haemangioma, lipoma arborescens and synovial sarcoma. The point worth highlighting in this report is the diagnosis of multifocal PVNS in a 5-year-old child. To the best of our knowledge, this is one of the youngest patients with multifocal PVNS reported in the literature.

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