

A Rare Manifestation of Pigmented Villonodular Synovitis of the Elbow in a Child

Hsiu Su, M.D., Elaine Gould, M.D., James Penna, M.D., and Hong Meng, M.D.

There are only a few reported cases in the literature of pigmented villonodular synovitis (PVNS) involving the elbow. Even more rare is its occurrence in the pediatric population as this condition mainly affects young adults. We report a unique case of an 8 year old girl presenting with diffuse form of PVNS of the elbow. The diagnosis is often not considered by clinical history and plain films as both are nonspecific in suggesting PVNS. MRI demonstrates the characteristic findings of PVNS, and therefore, is the imaging modality of choice for the evaluation of PVNS. When evaluating a pediatric patient with elbow pain, it is important to be aware of PVNS as part of the differential diagnosis if imaging features are suggestive and other etiologies have been excluded by history or imaging.

Introduction

Pigmented villonodular synovitis is an uncommon benign proliferative disorder of the synovium of the joint, tendon or bursa of uncertain nature. Traumatic, inflammatory and neoplastic pathogeneses have been

suggested for this condition [1]. It may appear as a localized or diffuse form. The diffuse type tends to recur in one-quarter of cases. The incidence of PVNS is approximately 2 patients per million population. It tends to occur in young adults, with no preponderance in sex. PVNS is more commonly a monoarticular process, most commonly affecting the knee joint, but may also affect other major joints, even the intervertebral and temporomandibular joints. In decreasing order of frequency, PVNS most commonly affects the knee, hip, ankle, shoulder, and elbow [2]. The clinical symptoms of PVNS are usually nonspecific. Patients often present with pain and swelling that is often mild and intermittent. The duration often ranges from months to several years.

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Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging; PVNS, pigmented villonodular synovitis

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Case Report

Patient is an 8-year-old girl who initially presented

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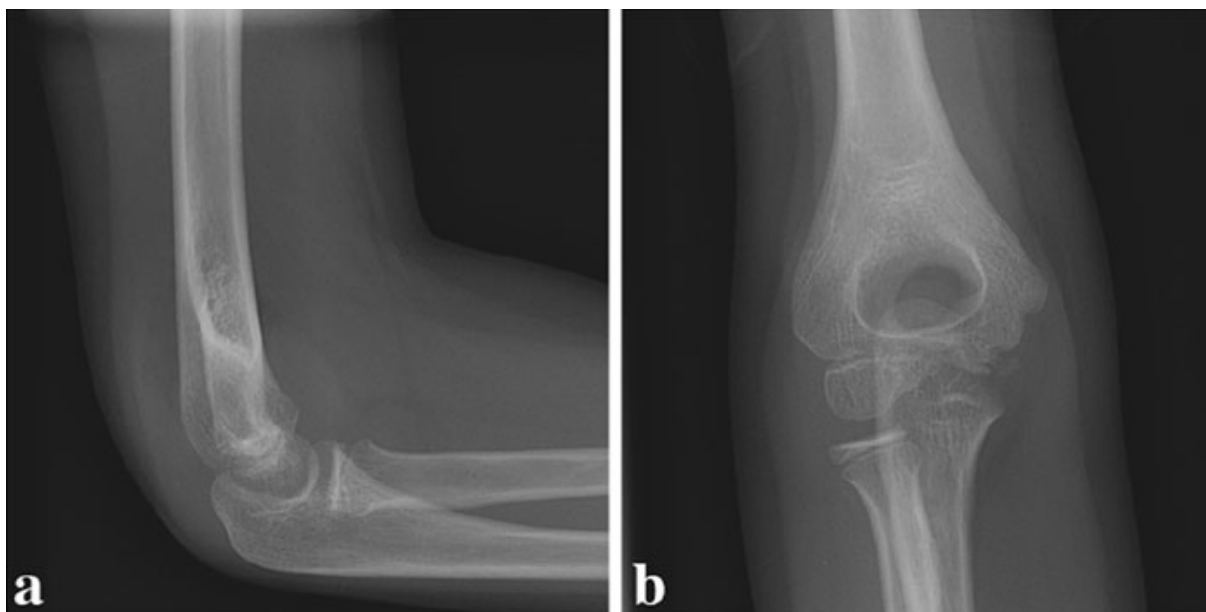


Figure 1. 8-year-old girl with PVNS. Lateral (a) and AP (b) elbow radiographs demonstrate large elbow joint effusion with preserved joint spaces.

with right elbow pain with decreased range of motion. There was no history of trauma. The patient did not have signs or symptoms of infection. Initial radiographs (Fig. 1) of the elbow demonstrated a large elbow joint effusion displacing the anterior and posterior fat pads. No fracture or bony destruction was identified and the joint spaces were preserved. Of incidental note was a septal aperture in the intercondylar region.

Six months following initial radiographs, the patient returned to the emergency room with complaints of increasing episodes of elbow pain and continued decreased range of motion. MRI was subsequently obtained. MRI demonstrated a large area of synovial nodular soft tissue masses within anterior and posterior joint space low signal on T1-weighted and proton density pulse sequences (Fig. 2), which represented hemosiderin deposits. No marrow abnormalities were noted. There was a small joint effusion. Gradient echo pulse sequence demonstrated more pronounced low signal due to increased susceptibility artifact from hemosiderin (Fig. 2C).

The patient subsequently underwent successful four portal elbow arthroscopy, and a complete arthroscopic

synovectomy was performed (Fig. 3). A synovial sample was obtained during the arthroscopic procedure and pathologic evaluation confirmed the diagnosis of PVNS. Following recovery, patient regained full range of motion and remained pain free. To this date, patient remains asymptomatic. H & E stain of synovial sample (Fig. 4) demonstrated hemosiderin laden within cytoplasm of multiple multinucleated macrophages characteristic of pigmented villonodular synovitis.

Discussion

Pigmented villonodular synovitis is a rare benign proliferative disorder of the synovium of joint, tendon or bursa. Etiology is unknown and has been attributed to inflammation, neoplasm, and trauma. It usually affects young adults in the third and fourth decades. There are two types: diffuse and localized. The diffuse form involves the entire synovium within a joint and makes up 15-25% of PVNS. The localized form is usually extra-articular along the tendon sheath with the majority (80%) involving the volar surface of the wrist and hands. The

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Figure 2. A, T1 Sagittal MRI (TR 700, TE 9); B, Proton Density Sagittal MRI (TR 3000, TE 32); C, Gradient Echo Sagittal MRI (TR 750, TE 17, FA 30); D, Axial T1 MRI (TR 700, TE 9); E, Coronal T1 MRI (TR 700, TE9); demonstrate nodular low signal intensity mass on all sequences in the anterior and posterior elbow joint spaces consistent with hemosiderin deposits characteristically seen with PVNS.

localized form makes up 75-85% of PVNS. Clinically patients present with pain, swelling, and decreased range of motion. The condition is predominately monoarticular; the knee is the most common joint involved. PVNS rarely involves the elbow joint in adults [3-4]. We present a unique manifestation of diffuse PVNS in a pediatric patient affecting the elbow where there is a rare body of literature on this subject in children [5].

Radiographs of PVNS can be normal or show an intraarticular effusion, which can be dense. They can also demonstrate para/intra-articular soft tissue dense mass and in long-standing disease, juxtaarticular bone erosions. Computed tomography may show evidence of hemosiderin and fat deposits, and can detect bone erosions not evident on the radiographs.

On MRI, the appearance of PVNS depends on the relative proportions of lipid, hemosiderin, fibrous stroma, pannus, fluid, and cellular elements. The most characteristic finding is nodular intraarticular masses of low signal intensity on T1-, T2-, and proton density-weighted sequences: the low signal intensity is a result of the presence of hemosiderin deposition. Gradient echo pulse sequence correspondingly shows increased susceptibility artifact from hemosiderin, known as blooming phenomenon. PVNS lesions characteristically shows prominent contrast enhancement of synovium with the administration of gadolinium.

The differential diagnosis of heterogeneous joint effusion with intra articular low signal are PVNS, chronic effusion containing hemorrhage and hemosiderin

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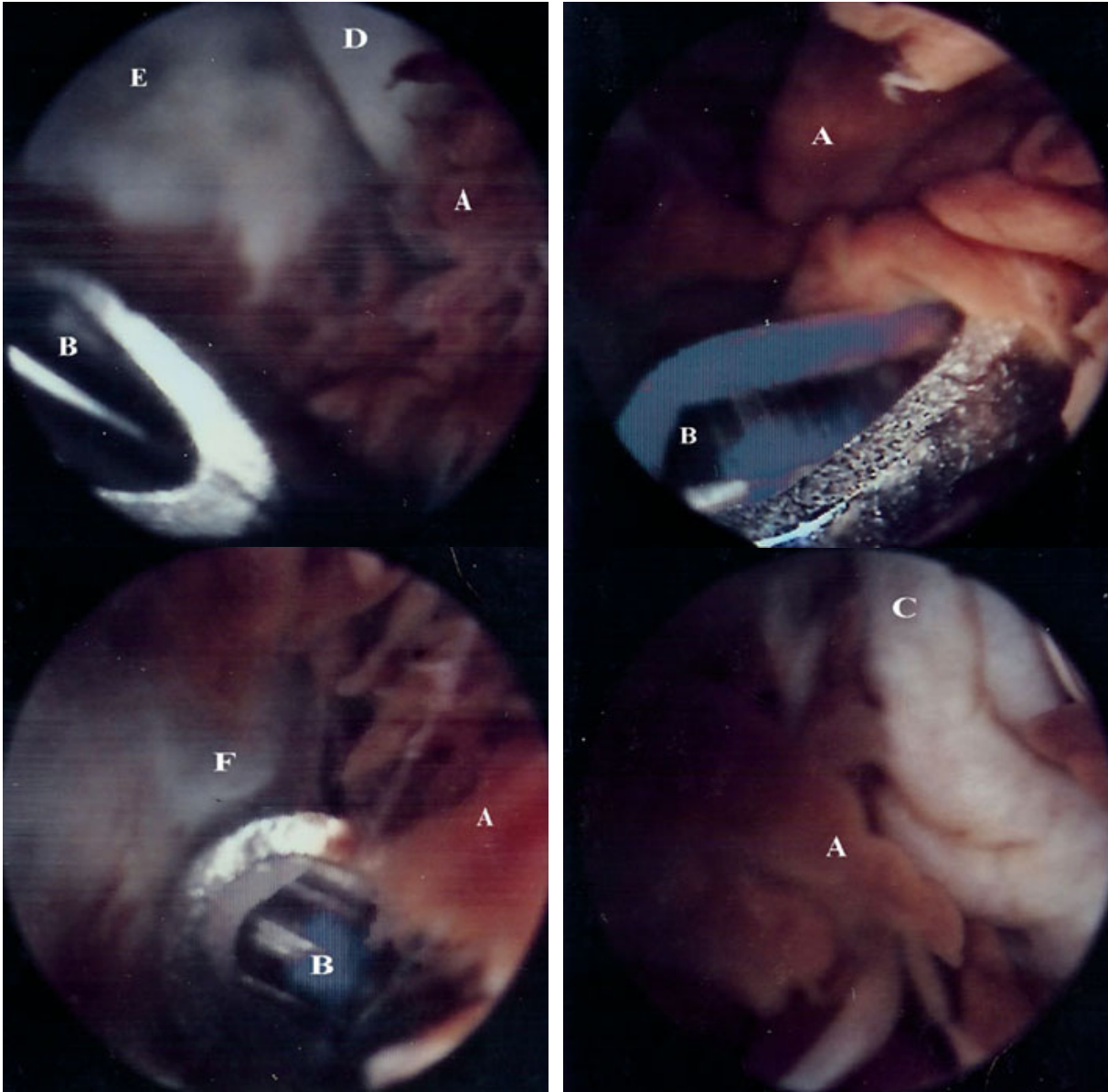


Figure 3. Images from arthroscopic synovectomy. Labels: A = Reddish-brown synovial fronds; B = 3.5mm mechanical shaver; C = Non-pigmented synovial hypertrophy; D = Proximal portion of the capitellum; E = Uninvolved joint capsule/synovium laterally; F = Medial joint capsule/synovium.

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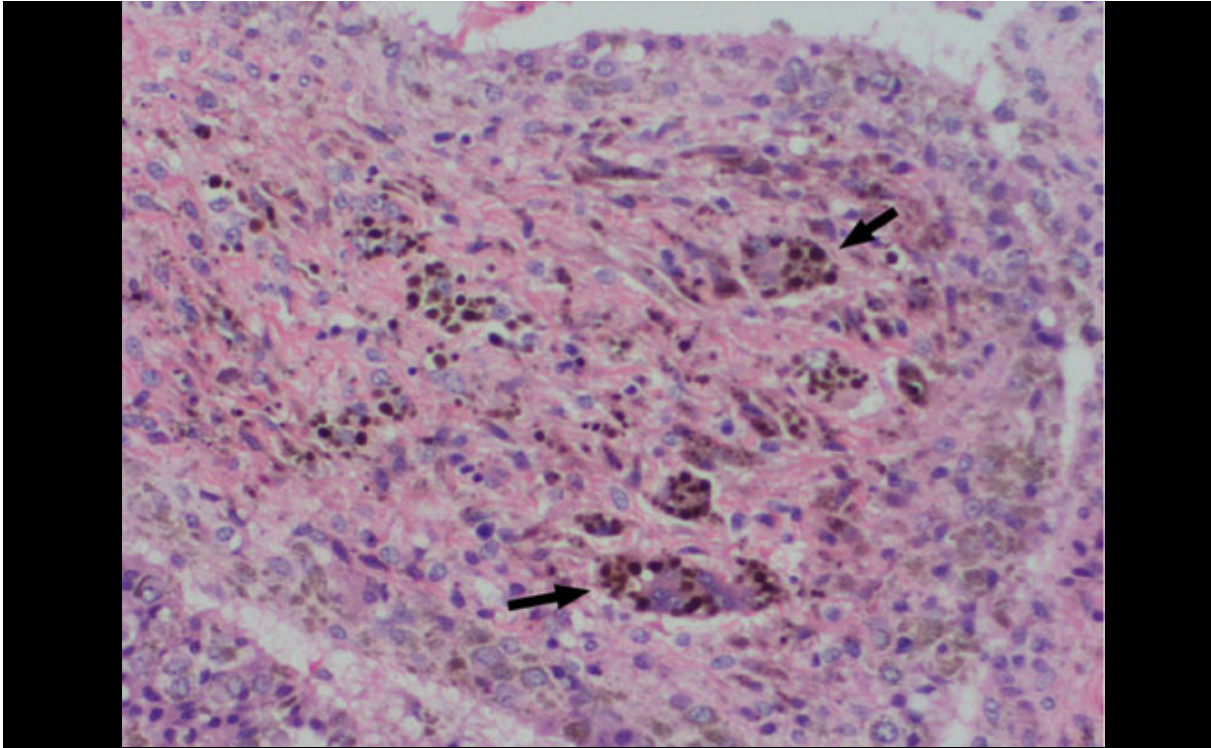


Figure 4. Photomicrograph of synovial tissue (H & E stain 400x magnification). Arrows point to two multinucleated giant cells containing brown pigmentation within the cytoplasm secondary to hemosiderin.

from entities such as hemophilia, trauma, intraarticular hemangiomatosis/hemangioma and inflammatory conditions with chronic effusions such as rheumatoid arthritis. Our case is typical of the diffuse form of PVNS. If discrete radiodense bodies present, synovial osteochondromatosis would be considered. Less likely, with the focal form of PVNS entities such as intraarticular chondroma could be included. In pediatric patients presenting with elbow pain, clinicians should be aware of PVNS and consider this condition in the differential diagnosis.

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