Pigmented villonodular synovitis of the elbow in a 16-year-old student

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Abstract

We present a 16-year-old student with right elbow pigmentary villonodular synovitis. Clinical diagnosis, radiological features, MRI findings, biopsy results, management and follow-up are discussed.

Key words: pigmented, villonodular, synovitis, elbow, rare.

Case report

A 16-year-old right-handed twelfth standard student came to our hospital with progressively worsening right elbow pain of 3 years' duration. There had been no precipitating accident or injury. There was no history of fever, waxing or waning of the symptoms and constitutional symptoms (weight loss, loss of appetite). On examination the right elbow had diffuse swelling. The range of motion of the elbow and forearm was 0° to 150° flexion, 90° supination, and 90° pronation. Radiographs showed no obvious soft tissue mass or any erosive or cystic changes (Figure 1). Complete blood count examination showed a total leukocyte count of 11,200 cu/mm. Haemoglobin and other parameters were normal. Erythrocyte sedimentation rate was 24 mm/h (Normal < 15).

Magnetic resonance imaging was done at this point to distinguish between an intra-articular process and a juxta-articular soft tissue mass. Muscle iso- to mild hyperintense lesion on T1-weighted images becoming heterointense on T2-weighted images noted in the posterior aspect of the elbow from the posterior fat pad region extending superiorly deep to the triceps muscle tendon unit. Insertion of triceps tendon was normal. On T2-weighted sequences intensively hypointense foci suggestive of haemosiderin were seen scattered in between intermediate and high signal intensities (Figures 2-5). The lesion was enhanced heterogeneously by the administration of Gd-DTPA contrast (Figure 6). No evidence of bony marrow involvement, erosion, cyst formation, and joint effusion, extracapsular extension of the lesion or signal intensity change in bone was seen. Olecranon and trochlea were normal. The anterior group of muscles was normal. Medial and lateral collateral ligaments were normal. Based on the MRI features, including the intra-articular location of the mass and the areas of low signal on both the T1- weighted and T2-weighted sequences, a clinico-radiographic presumptive diagnosis of PVNS was made.

Needle biopsy was not performed due to the lower likelihood of obtaining a definitive diagnosis from the smaller amount of tissue obtained by needle

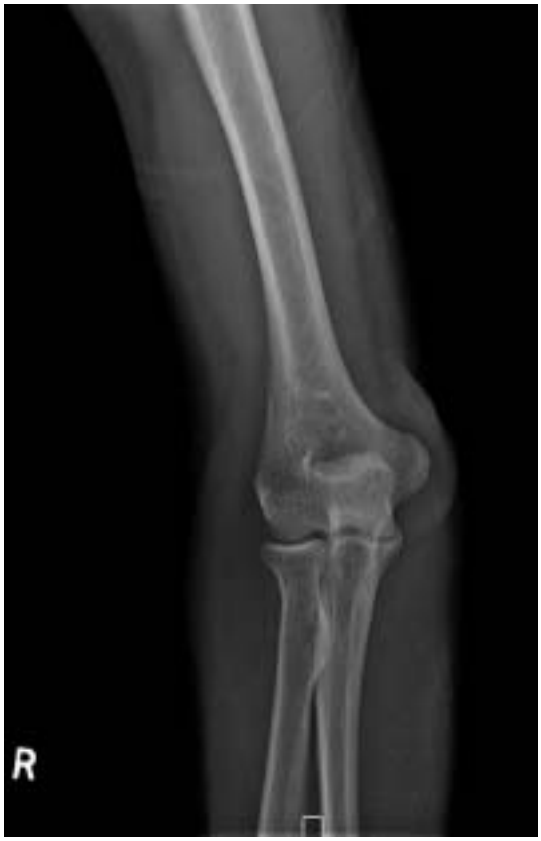

Figure 1. Shows the radiograph of the right elbow, which was normal. There was no bony erosion, and cyst formation seen

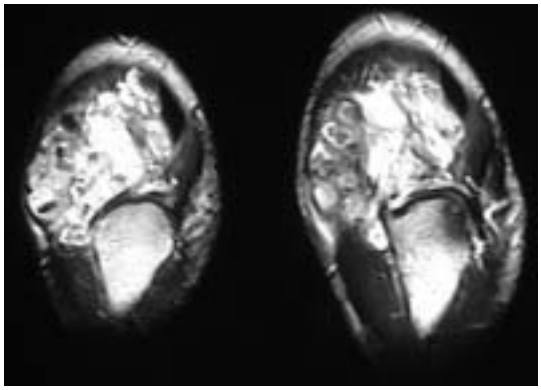

Figure 3. Axial T2-weighted fat-suppressed fast spin echo sequence demonstrating foci of low signal intensity within the predominately intermediate signal mass corresponding to similar areas on the T1-weighted image

biopsy. Through a posterolateral approach for the elbow, nearly total synovectomy was performed (Figure 7). A moderate amount of hypertrophic yellowish-gold synovium consistent with PVNS was seen. The joint surfaces showed no destruction. Histological evaluation showed haemosiderin-laden macrophages, neovascularisation, inflammatory cells, multinucleated giant cells, and foam cells, which confirmed the diagnosis of PVNS (Figure 8).

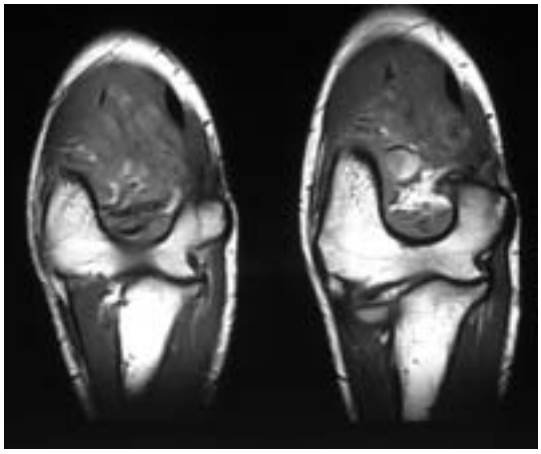

Figure 2. Axial T1-weighted fast spin echo image through the right elbow demonstrating a heterogeneous mass with low and intermediate signal areas within the joint

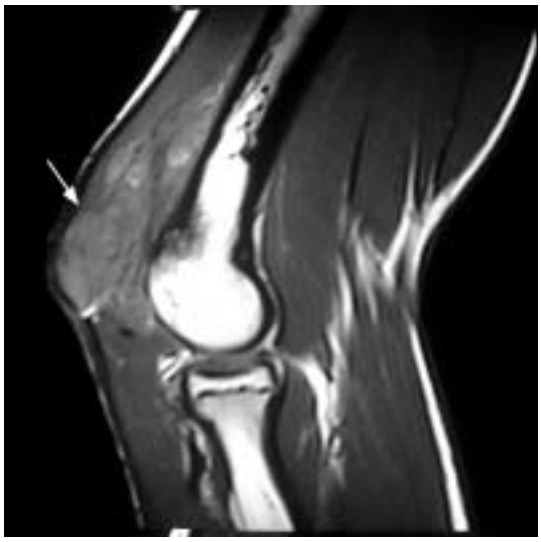

Figure 4. Magnetic resonance images show synovial proliferation (indicative of signal attenuation by the haemosiderin) as an isointense to mild hyperintense signal with the surrounding muscle in T1-weighted image and as high signal intensity in a T2-weighted image

The symptoms resolved and the patient returned to school. The elbow's range of movement was 130° in flexion, with 20° loss of extension. Radiographs and magnetic resonance images showed no recurrence or any degenerative changes. At the latest follow-up examination (48 months) following the initial surgery he remained asymptomatic and is under continuing follow-up surveillance.

Discussion

Pigmented villonodular synovitis (PVNS) was defined as a benign but locally progressive synovial proliferative lesion in the joint, bursae, and tendon sheath [1, 2]. The knee and hand are the most

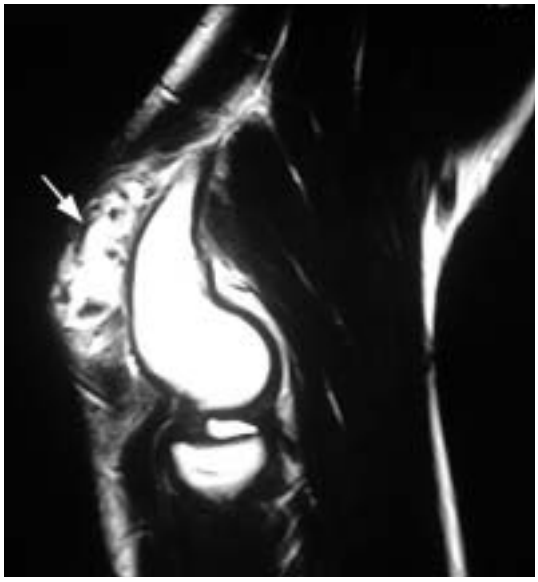

Figure 5. Magnetic resonance images show synovial proliferation (indicative of signal attenuation by the haemosiderin) as an isointense to mild hyperintense signal with the surrounding muscle in T1-weighted image and as high signal intensity in a T2-weighted image

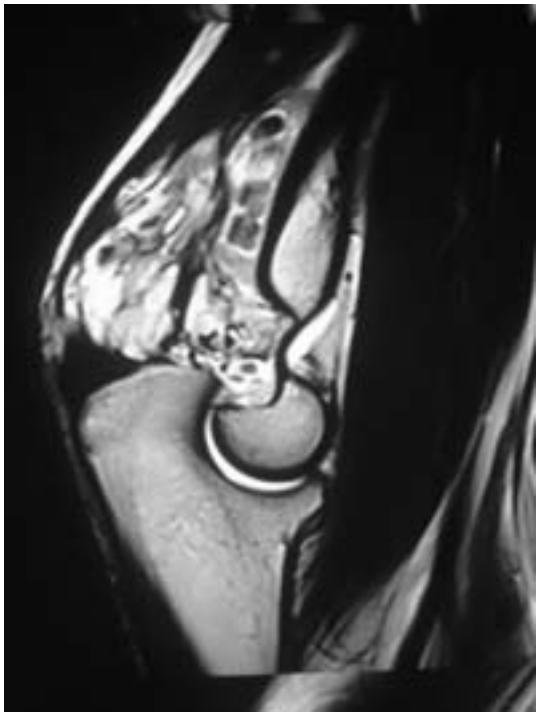

Figure 6. The lesion was enhanced heterogeneously by the administration of Gd-DTPA contrast

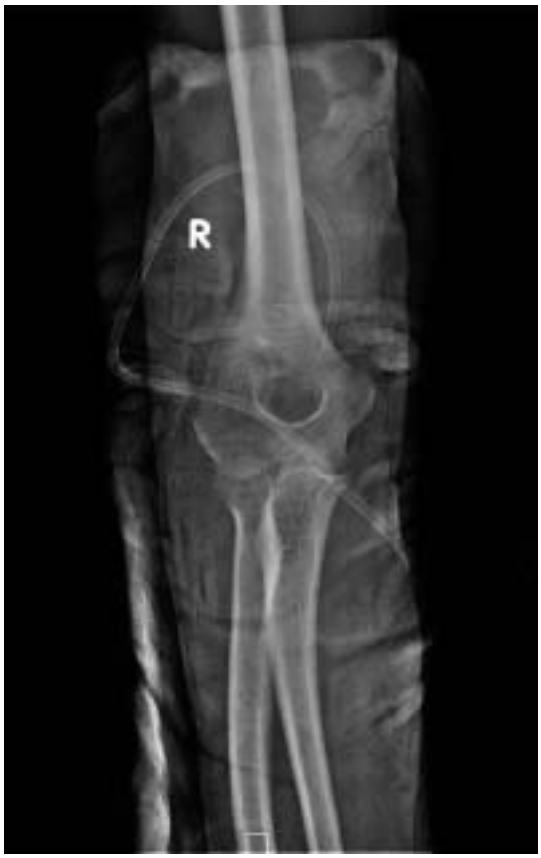

Figure 7. Post-operative radiograph of the right elbow after a total synovectomy

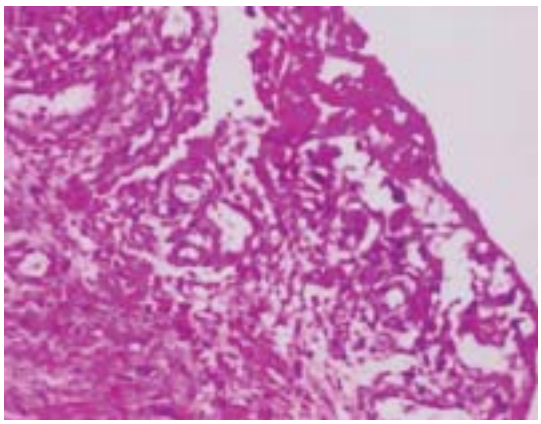

Figure 8. Hyperplastic synovium consists of solid and finger-like masses of fibrous stroma. Fibrous stroma contains foamy macrophages, lymphocytes, plasma cells, multinucleated giant cells, and golden brown haemosiderin within the cytoplasm of synovial lining cells and macrophages

frequently involved sites, followed by the hip, wrist, and ankle, joints of the hands and feet, and shoulder. Involvement of the elbow is rare. Only 21 cases of PVNS of the elbow have been reported [2, 3]. It can occur in all age groups; adults in their third or fourth decade commonly present with symptoms of pain and recurrent effusions of the joint [2]. All cases were monoarticular except for that reported by Lindenbaum and Hunt, which was polyarticular and involved both elbow joints [3, 4]. The duration of symptoms was from 1.5 to 6 years (mean 3.6 years). Swelling was the most

common presenting symptom. Pain, limited range of motion, ipsilateral hand numbness, minimal to mild joint swelling, weakness, heat and tenderness are other symptoms reported in various reports [3-6]. The disease is usually slowly progressive. Laboratory findings including blood count and sedimentation rate are normal, and joint fluid is most often serosanguineous.

The radiographic findings in the active phase of PVNS correlate well with the pathology. Localized nodular variety of PVNS has normal radiograph study. Bone mineralization is also normal with nodular and diffuse varieties of PVNS. Involvement by the diffuse type will result in noncalcified periarticular (intracapsular) soft-tissue masses, juxtaarticular cysts, small erosions of the subchondral bone and arthrosis. Arthrography, radionuclide scintigraphy, computed tomography, angiography, and MRI were used in isolated cases in the literature as diagnostic modalities [1-5]. Magnetic resonance imaging strongly suggests the diagnosis by demonstration of low signal areas on T1- and T2-weighted images, indicative of signal attenuation by the haemosiderin, within a thickened synovium [6]. The presence of fatty signal within the synovium due to accumulation of lipid-laden macrophages is another classic feature sometimes observed within the synovium of PVNS. This feature was not observed in our case. Definitive diagnosis of PVNS is usually confirmed by histopathological examination of biopsy tissue specimens [5-8].

Grossly, the synovium has a mossy or nodular texture, spongy cut surfaces, and a rusty, red-brown, or yellow-brown colour. Microscopically, the synovium is composed of finger-like or rounded masses of fibrous stroma covered by hyperplastic lining cells. Large numbers of foamy macrophages in the stroma account for the yellow colouration, and the rusty or brown colour is due to haemosiderin deposits in the stroma and in the cytoplasm of macrophages and synovial lining cells. The aetiology of PVNS has been variously attributed to an inflammatory response to an unknown agent, repeated haemorrhage into the joint, neoplasia, disorder of lipid metabolism, repeated minor trauma, patient with abnormal cellular and humoral immunity and proliferation of histiocytes. No single explanation has been incontrovertibly proven, however [7-9]. Because of this lack of specificity, together with the rare occurrence of the disease, late diagnosis is not uncommon, making correct management an even greater challenge in the growing child. Genetic factors are suspected in childhood cases [9].

Synovial sarcoma must be differentiated from PVNS. The lesion entirely or partly outside the joint capsule, with scattered, irregular calcifications within the mass, and bone destruction, is synovial sarcoma. Rheumatoid arthritis, tuberculous arthritis, osteoarthritis, angiomas of osseous origin, amyloidosis, fibrous dysplasia, and multiple enchondromatosis are the differential diagnostic considerations when cystic bone changes are seen [7-9].

Treatment of pigmented villonodular synovitis depends primarily on the severity of joint destruction and secondarily on the age of the patient [1-5]. The treatment of choice in the early case (before significant cartilage loss or bony erosion) is a complete synovectomy as possible to reduce the likelihood of recurrence. Unfortunately, despite meticulous surgical treatment, the recurrence rate has been reported to be as high as 46%. In the cases involving the elbow joint, of duration ranging from 1 to 4 years, none of the cases showed a recurrence during the follow-up period [1-9]. When the disease is advanced (severe pain and limited joint motion due to destruction of joint cartilage and/or bone erosion), arthrodesis or arthroplasty is indicated. Irradiation cannot be recommended for younger patients, because of possible carcinogenic effects and because of the high incidence of induced joint stiffness. When PVNS occurs at the tendon sheath, it is relatively easy to remove the entire lesion. Nonetheless, because of the complexity of the joint structure in the knee and elbow joints, complete resection of the synovial tissue in these joints is often difficult [1-9]. For patients severely affected by bony destruction and arthroplasty use of radiation along with synovectomy has been reported.

Our patient came to us with progressive pain of the right elbow with a diffuse swelling. His elbow function, blood parameters and radiographs were normal. Magneric resonance imaging was done to differentiate between the juxta- and intra-articular involvement. It showed synovial proliferation depicted as having iso-intensity with the surrounding muscle in a T1-weighted image and high signal intensity suggestive of PVNS. Biopsy confirmed the diagnosis. Complete synovectomy was done. The patient regained his normal elbow activities at the end of 4 years' follow-up. The literature on PVNS is huge and refers, not specifically, to every joint. The fact that PVNS in an adult has not been extensively described does contribute a base for our observation.

In conclusion, pigmented villonodular synovitis should be included in the differential diagnosis of elbow monoarticular joint swelling in young and middle-aged adults. The MRI signal attenuation by haemosiderin results in low signal areas on both T1- and T2-weighted sequences, which again is the diagnostic modality of choice and assists the surgeon in making a proper incision. Correlation of all the clinical aspects and the histological features is usually required for definitive diagnosis. A complete surgical synovectomy provides the best chance for cure, but the goal of eradication of this

benign process must be balanced against the morbidity of more aggressive surgery.

Acknowledgments

Written consent for publication was obtained from the patient's father.

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