Skeletal Radiology
OPEN QUIZ—SOLUTION

Skeletal Radiol (1990) 19:131–133

Case report 578

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Imaging studies

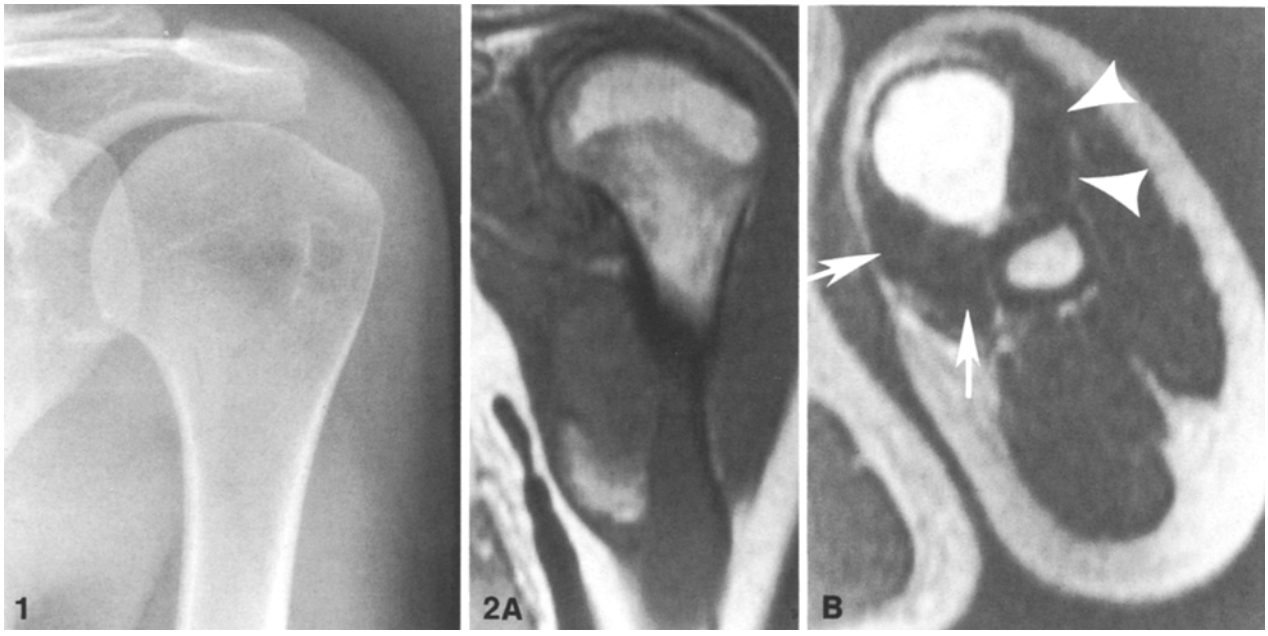

Fig. 1. A plain radiograph shows a small non-specific lytic lesion with sclerotic margins in the humeral head between the tuberosities

Fig. 2. A a Coronal T1-weighted MR scan (TR:700 ms; TE 25 ms) shows a mass of slightly increased signal intensity with an area of very high signal inferiorly. **B** an Axial T2-weighted MR scan (TR:2000 ms; TE 80 ms) shows a discrete mass of very high signal intensity in the anterior compartment of the arm, displacing the long (*arrows*) and short heads (*arrowheads*) of biceps muscle

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Clinical information

This 18-year-old woman was referred for evaluation of a painful, enlarging mass on the inner aspect of her left upper arm. On examination, a 5×7 cm mobile mass on the medial aspect of the left upper arm was noted to extend into the axilla.

Plain radiographs showed a lucent defect in the head of the humerus (Fig. 1). Magnetic resonance (MR) imaging was performed in the coronal and axial planes on a Signa (General Electric, Milwaukee) 1.5-T unit, using a surface coil. A $3 \times 3 \times 8$ cm mass was demonstrated, extending distally from the coracoid process, between the heads of the biceps muscle into the anterior aspect of the upper arm. On T1-weighted images, the signal intensity of the mass was slightly higher than that of surrounding muscle with an area of very high signal inferiorly (Fig. 2A). On T2-weighted images, a high signal throughout the lesion was present with the distal portion again slightly brighter (Fig. 2B). No areas of signal dropout were identified.

The mass was resected.

Diagnosis: Pigmented villonodular synovitis of the shoulder

Pigmented villonodular synovitis (PVS) must be distinguished from synovial sarcoma. While the clinical presentation may be similar, synovial sarcoma is frequently located outside the joint space and may contain areas of irregular calcification [1,3,10]. Rheumatoid and tuberculous arthritis should be considered when cystic bone changes are present [1, 3]. The clinical history and polyarticular location will often help differentiate these lesions from PVS. Other diseases in the differential diagnosis include calcium pyrophosphate deposition disease, angiomas of osseous origin, fibrous dysplasia, and multiple enchondromatosis [1, 3].

On pathological examination, PVS is characterized by pigmentation, villous transformation of the synovium, and the presence of foam cells. On gross examination, the lesion is rusty brown to golden yellow, reflecting the amount of hemosiderin and lipid. Nodular hypertrophy is also often found and is due to proliferation of synovium. Microscopically the appearance varies. Foam cells with ingested hemosiderin, multinucleate giant cells, or fibrotic changes may be seen, depending on the activity of the disease [2, 3].

At surgery in the case presented here, a white, shiny, encapsulated cystic lesion was found. It extended distally from the coracoid process of the scapula, lying between the short and long heads of the biceps, and contained thick brown fluid. The resected specimen was a pinkish tan mass which was hemorrhagic, friable and necrotic. Histological studies showed hemosiderin laden macrophages. A diagnosis of PVS was made.

Synovectomy was performed approximately two months later. At that time a large quantity of synovium histologically typical of PVS was removed from around the biceps tendon and the shoulder joint (Fig. 3). A small focus of intra-osseous extension was resected, corresponding to the bone lesion in the humeral head (Fig. 1).

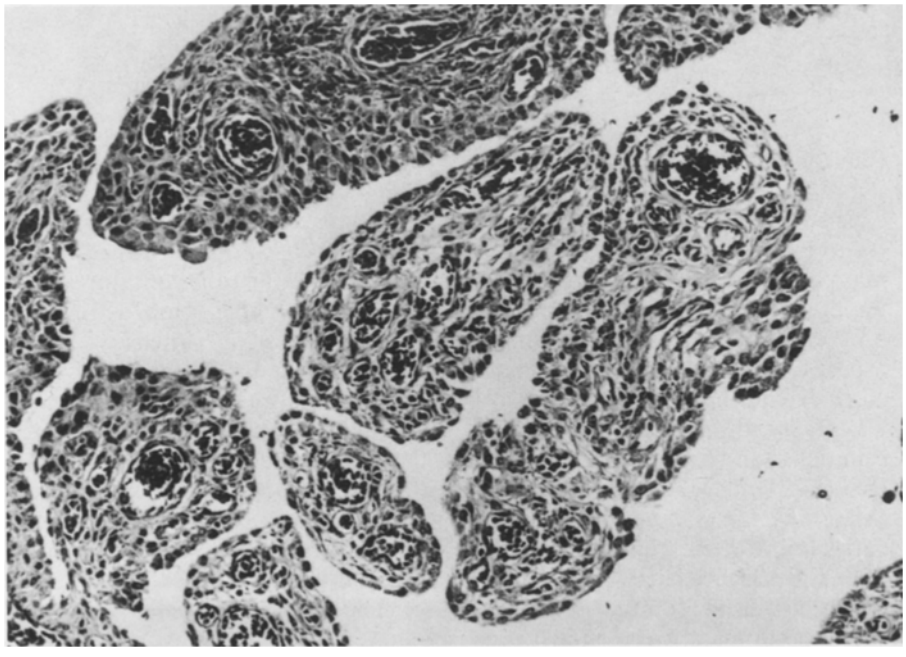
Discussion

Pigmented villonodular synovitis (PVS) is uncommon, with an annual incidence in the United States of 1.8 individuals affected per million [7]. It is a disease of young adults with a marked predilection for the lower extremity [1, 3, 10]. The knee is the site of involvement in up to 80% of cases and monarticular involvement is the rule. Other joints involved in decreasing order of frequency include the hip, ankle, small joints of the hands and feet, and rarely, the shoulder. To our knowledge, shoulder involvement has been described in only nine patients [2]. The clinical history is typically one of intermittent pain and swelling with associated joint stiffness and locking [1–3, 10].

The etiology of PVS remains unknown. Originally described as inflammatory by Jaffe et al. [5], various authors have postulated neoplasia, repeated trauma, a disorder or lipid metabolism, and overgrowth of synovial and perisynovial cells as possible causes [1, 10].

Traditionally, PVS has been evaluated by plain film radiography and occasionally by arthrography. Soft tissue changes or effusions are not uncommon [1, 3, 8] and localized osseous changes are found in approximately 50% of patients [1, 7]. CT of PVS may demonstrate a high attenuation area due to deposition of hemosiderin in the abnormal synovium [9].

MRI has recently been used to evaluate PVS in the knee [6, 11] and hip [11]. Lesions in both locations demonstrated intermediate signal intensity on T1-weighted images, and areas of high signal intensity inter-



Pathological study

Fig. 3. In this illustration there is a villous proliferation with centrally located blood vessels, reactive fibroblastic cells and reactive synovial membrane typical of pigmented villonodular synovitis

spersed with signal dropout on T2-weighted images. The areas of high signal corresponded to inflamed synovium and fluid collections. The areas of signal dropout were attributed to the presence of hemosiderin [6, 11]. In the brain, the mechanism of signal dropout at the periphery of intracranial hematomas has been described by Gomori et al. [4] as enhancement of proton relaxation due to the paramagnetic effect of hemosiderin. This results in shortening of the T2 relaxation time of surrounding water with associated signal dropout [11].

In our case, no areas of signal dropout were seen, despite the pathologically proven presence of hemosiderin. The high signal on the T2-weighted images could be explained by the presence of edema, inflammation, fat, or chronic hematoma, all of which could contribute to mask the paramagnetic effect of hemosiderin on the overall T2-weighted image. The absence of signal dropout could also reflect the relatively small amount of hemosiderin present.

Multiplanar imaging allows accurate preoperative localization of the lesion.

In *summary*, PVS is an uncommon disease of uncertain etiology, which rarely affects the shoulder. Pathologically, hemosiderin deposition is characteristic. Recent reports have demonstrated MRI findings of signal dropout on T2-weighted images attributed to the presence of hemosiderin. Our case reiterates the value of coronal scanning in certain patients and suggests that signal dropout on T2-weighted images is not an essential finding in PVS.

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