SONOGRAPHY

CASE SERIES

Pigmented villonodular synovitis

Quanson Sirlyn

Capital Radiology, Vermont South, Victoria, Australia

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Introduction

Pigmented villonodular synovitis (PVNS) is an uncommon benign proliferative disorder of the synovium that results in formation of villous and nodular protrusions.[1] There are two forms, categorised by the extent of involvement: diffuse and nodular. The diffuse form commonly involves the large joints, and the nodular form typically occurs around the hands.[2] PVNS is further categorised by its location: intra-articular or extra-articular. When PVNS extends beyond the intra-articular synovium, it may occur extra-articularly in a bursa or tendon sheath, which is known as pigmented villonodular bursitis (PVNB) and pigmented villonodular tenosynovitis (PVNTS), respectively.[3] The aim of this case report is to discuss two forms of PVNS: localised extra-articular that occurs in the ring finger and diffuse intra-articular that occurs in the ankle. Discussion will be made of the clinical presentation, the ethiopathogeny of the disease and the ultrasound characteristic features, and will also briefly address magnetic resonance imaging (MRI) and radiographic appearances.

Case one description – Localised extra-articular disease (PVNTS)

A 22-year-old woman presented with a slow growing, superficial lump on her left ring finger. The lump had been growing over a period of 2 years.

It was located superficially on the volar surface of the proximal interphalangeal joint. Her doctor requested an X-ray and ultrasound examination of the affected finger. A Logiq E9 (GE Medical Systems, Zipf, Austria) ultrasound system, with an 8–18 MHz linear-array transducer was used. The plain X-ray indicated soft-tissue swelling with no underlying bony lesion or arthropathy (Figure 1). Panoramic ultrasound imaging in the longitudinal plane demonstrated a predominantly hypoechoic, heterogeneous solid mass with multiple lobulations surrounding the flexor tendon sheath. The mass extended just distal to the metacarpo-phalangeal (MCP) joint to just proximal to the distal interphalangeal (DIP) joint (Figure 2). Transverse imaging over the palpable lump demonstrated a thickened hypoechoic area (Figure 3). The lesion showed no hyperechoic, cystic or calcifications regions. Colour Doppler imaging was used and demonstrated an increase in internal vascularity (Figure 4). Dynamic scanning demonstrated that the mass did not move with the tendon when the affected finger was flexed or extended. The

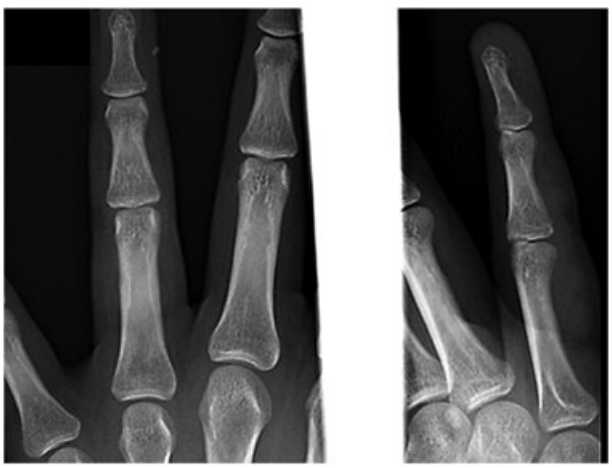

Figure 1 Ring finger soft-tissue swelling with no underlying bony lesion.

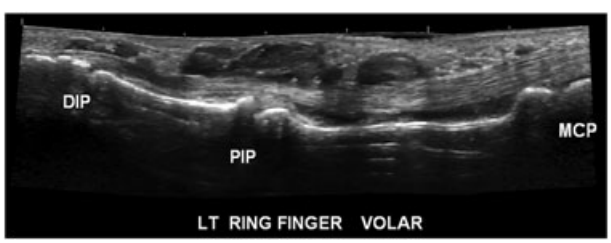

Figure 2 Panoramic ultrasound imaging. Hypoechoic, heterogeneous solid mass with multiple lobulations surrounding the flexor tendon sheath.

Correspondence: Quanson Sirlyn,
E-mail: q.sirlyn@capitalradiology.com.au
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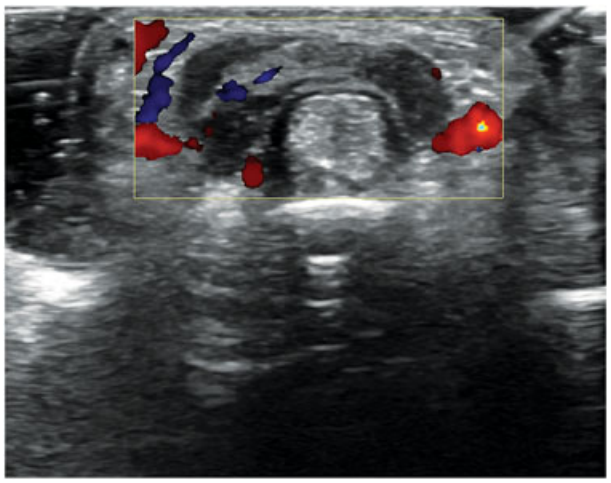

Figure 3 Transverse B-mode and Colour Doppler images shows the hypoechoic, heterogeneous, solid mass encasing the flexor tendons.

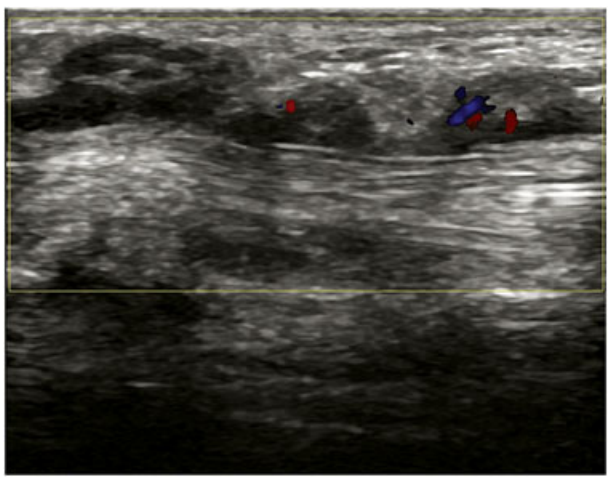

Figure 4 Colour Doppler imaging demonstrated an increase in internal vascularity of the solid mass.

ultrasound differential diagnosis included PVNTS, sarcoma or possibly a vascular malformation. MRI was recommended as further investigation of the lesion. The MRI images were generated using a Magnetom Avanto 1.5 T (Siemens, Erlangen, Germany) system. MRI demonstrated a soft-tissue mass surrounding the flexor tendon sheath. It extended from just distal to the MCP joint to just proximal to the DIP joint. It was heterogeneous in appearance in T2 signal with areas of both bright and low T2 signal suggestive of haemosiderin staining. It showed mild diffuse enhancement (Figure 5). The findings were reported as most compatible with extensive PVNTS.

Following this, the patient underwent local surgical resection, and a nodular, brownish soft-tissue mass was removed. The histology report of the mass was consistent with PVNTS. Recurrence of PVNTS has been reported as being rare following resection of extra-articular nodular synovitis.[3]

Case two description – Diffuse intra-articular disease (PVNS)

A 60-year-old man presented to our department for a left ankle ultrasound, with chronic swelling around the Achilles tendon. The patient indicated that the mass had gradually enlarged over the last 2 years. The referring doctor requested an ultrasound examination of the posterior ankle. The scan was performed using a Logiq E9 (GE Medical Systems, Zipf, Austria) E9 ultrasound system with a multifrequency linear-array transducer of 6-15 MHz. Colour Doppler, with a low pulse repetition frequency (PRF) setting, was used to demonstrate the internal vascularity of the mass. Ultrasound revealed a grossly abnormal hypoechoic, heterogeneous mass inferior but separate to the Achilles tendon. The lobulated mass appeared to arise from the subtalar joint (Figure 6). Colour Doppler shows some internal vascularity (Figure 7).

The preliminary ultrasound differential diagnosis was PVNS, and the report was forwarded to the referring doctor, with a recommendation of further evaluation using MRI. The patient presented for MRI of the symptomatic ankle and foot one week later. The MRI demonstrates a heterogeneous, multilobulated mass arising from the posterior aspect of the subtalar joint. It was predominantly centred within Kager's fat pad and caused smooth

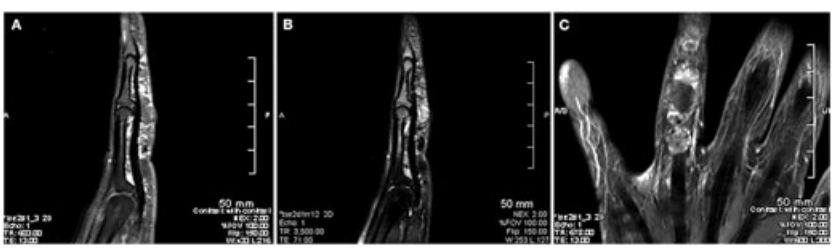

Figure 5 Magnetic resonance imaging images show heterogeneous in T2 signal with areas of both bright and low T2 signal suggestive of haemosiderin staining. It showed mild diffuse enhancement.

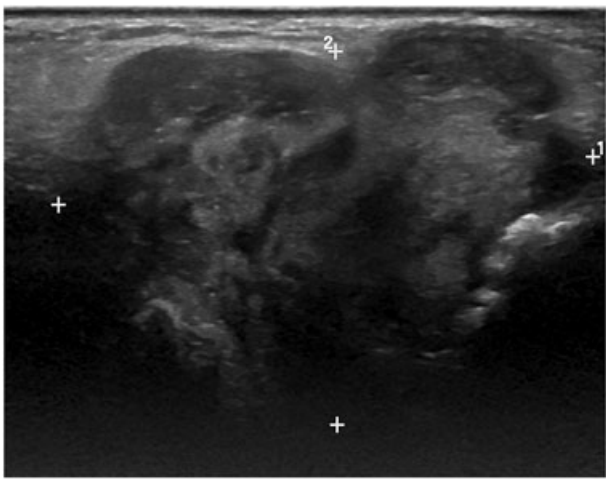

Figure 6 B-mode ultrasound demonstrates a solid, lobulated, heterogeneous mass arising from the subtalar joint.

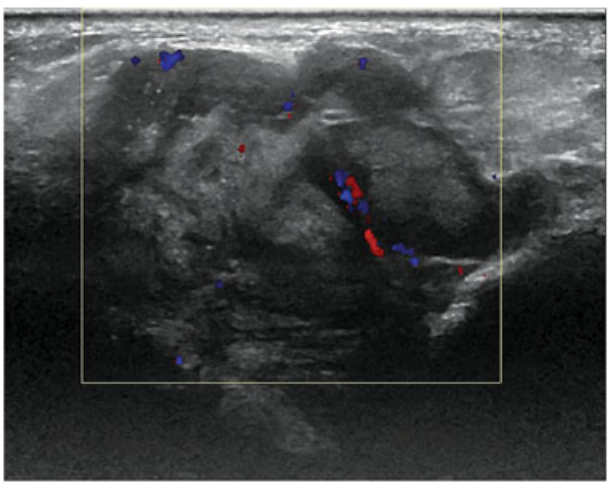

Figure 7 Colour Doppler demonstrates the lobulated mass contains internal vascularity.

remodelling of the posterosuperior surface of the calcaneum. Figures 8 and 9 demonstrate the sagittal MRI images of the lesion in T1-weighted inversion recovery and proton density, respectively. Figure 10 represents the axial scan of the lesion in T2-weighted fat saturation, showing an extensive area of haemosiderin within the lesion. There are some T2-weighted bright areas that demonstrate enhancement after contrast. The Achilles tendon is demonstrated as normal throughout its length. The retro Achilles and retrocalcaneal bursa are also normal in appearance. These findings were most compatible with extensive diffuse intra-articular PVNS.

Following imaging, the patient was referred to an orthopaedic surgeon for further evaluation and management.

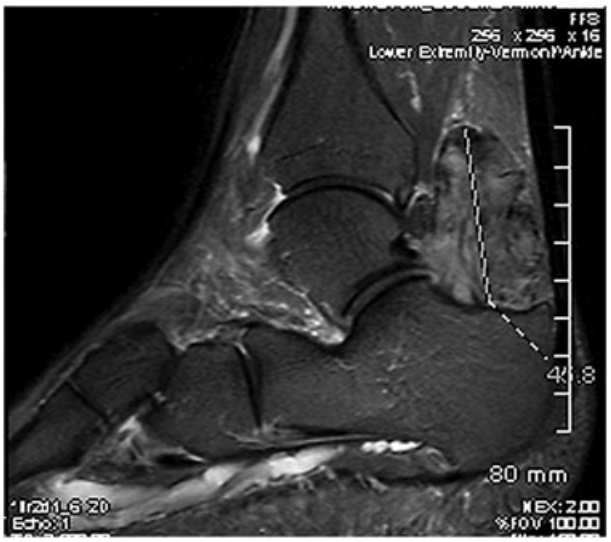

Figure 8 Sagittal magnetic resonance imaging images of the lesion in T1-weighted inversion recovery.

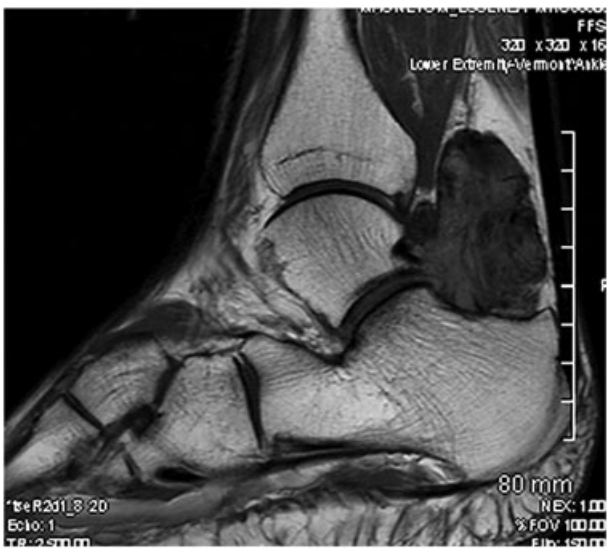

Figure 9 Sagittal magnetic resonance imaging images of the lesion in proton density.

Subsequently, the patient underwent local excision to remove the soft-tissue mass, and there were no complications. Based on the histological finding, PVNS was confirmed.

Discussion

PVNS can potentially affect any joint. It is a rare benign condition of the synovium that results in formation of villous and nodular protrusions.[1] The incidence of PVNS has been reported as 9.2 and 1.8 cases per year,

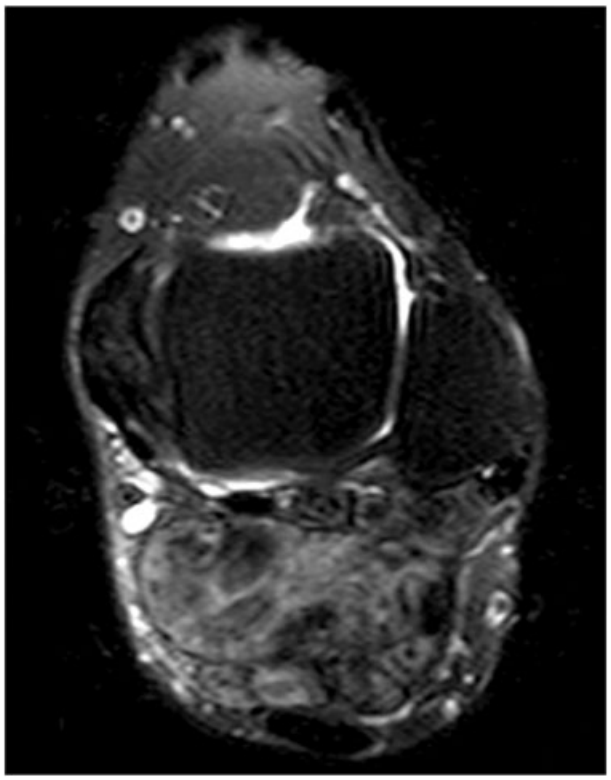

Figure 10 Axial scan of the lesion in T2-weighted fat saturation, showing an extensive area of haemosiderin within the lesion.

per million individuals for the extra-articular and intra-articular forms of the disease, respectively.[3,4] It often occurs in young adults.[5] There is comparative incidence of male to female gender seen in the diffuse intra-articular form of the disease, whereas the ratio for male to female gender in the localised extra-articular form is 1:1.5–2. In a study of 166 cases of PVNS, 77% of cases were localised disease (whether extra-articular or intra-articular).[3] The remaining 23% were found to be the diffuse intra-articular form.[6] The current description of PVNS was first proposed by Jaffe et al. in 1941[7] and described the benign proliferative inflammatory nature of these manifestations. The term PVNS was further subclassified in 1976 to PVNB or PVNTS[4]; PVNTS is frequently referred to as giant cell tumours of the tendon sheath.[4] PVNTS frequently involves the hand or wrist, specifically in the long and index fingers.[8–10] It occurs twice as frequently in the volar than the dorsal aspect of the hand.[8,11,12] It is the most common soft-tissue mass of the hand and wrist secondary to ganglia.[8,13] PVNS and PVNB are predominantly found in the knee, with 80% of PVNS cases involving the knee.[3,4] It may occur in other large joints, such as the hip, ankle and shoulder, but multiple joint involvement is extremely rare.[14] PVNS may extend beyond intra-articular synovium, occurring extra-articularly in a bursa or tendon sheath.[3]

Depending on the location of the lesion (intra-articular or extra-articular), the clinical symptoms can vary greatly.[3,15] Intra-articular PVNS frequently manifests clinically with pain (79–90%) and is often associated with swelling in the region (72–79%). Joint dysfunction (26–28%) and the presence of a soft-tissue mass (6–19%) are also reported.[16,17] With extra-articular PVNTS or PVNB, soft-tissue masses occur commonly (83–99%) and pain variably (22–71%). Joint dysfunction and swelling are less frequent symptoms (0–4%).[12,18] PVNS is a chronic disease, with the duration of the symptoms ranging from months to years.[15]

The ethiopathogeny of this disease remains uncertain, but studies have shown that it is possibly associated with repeated hemarthrosis and possibly due to the affect of multiple mild trauma, or a disorder of lipid metabolism.[3,4] Other authors describe PVNS as a result of haemorragic synovial disorders.[5] As discussed previously, PVNS may be diffuse or localised. In the diffuse form, the entire synovial lining membrane becomes thick and overgrown, accumulating a brownish soft-tissue pigment known as haemosiderin. All forms of PVNS usually contain haemosiderin, although the extent of haemosiderin deposition is found to be more prominent in the diffuse intra-articular form.[3,15] In the localised form, a focal area of synovial lining membrane becomes thickened and overgrown, resulting in a discrete nodule. This nodular tissue remains attached to the synovial lining by a stalk. Although this disease proliferation results in abnormal tissue growth, and at times can be quite aggressive, the condition has not been known to metastasise, and is uniformly benign.[19] In the two cases discussed, the patients did not complain of pain, but symptoms of PVNS have been known to be progressive. In the localised form, the diagnosis is quite difficult, especially in the early stage of the disease. The lump does not become noticeable until the tissue nodule reaches a size that causes impingement and swelling. As in the first case discussed, the patient noticed that the finger lump enlarged and experienced difficulty in flexing her ring finger. The symptoms worsened over the year. Lesions typically range from 0.5 to 4 cm in greatest dimension.[3]

If the localised form of PVNS occurs in a region of a bigger joint, the nodule could act as intra-articular-free bodies, causing internal joint impingement.[3] The patient might feel some mechanical problems, like snapping or joint locking, simulating a torn cartilage. There may be complaints of abnormal mobility.[4] The diffuse form of PVNS is more problematic, because the abnormal mass extensively infiltrates and causes thickening of the entire synovial lining.[3]

With localised intra-articular disease, once the lesion has undergone total resection, in open operative or arthroscopic techniques, the success rate is 100%. With the extra-articular disease, the recurrence rate following surgical resection ranges from 0% to 44%.[3] On the other hand, the diffuse intra-articular PVNS recurrences rate, following initial treatment, ranges from 8% to 56%. As a whole, the recurrence rate for diffuse intra-articular form is higher than the localised form.[3] An average of 24 months from the initial treatment has been reported for a lesion to recur.[20] For patients with recurrent diffuse intra-articular PVNS, the treatment options are a combination of therapy and/or joint replacement.[3]

There is no known imaging technique that provides an absolute characteristic feature in the diagnosis of PVNS. Also, there are no conclusive clinical presentations definitive of the condition. PVNS can only be diagnosed based on the histological examination. The histological findings show that there is a presence of intracellular and pigments of subsynovial haemosiderin, the presence of macrophage multinucleate giant cells, foamy histiocytes and inflammatory cells.[4]

Imaging provides no absolute characteristic features. Initial complementary imaging examinations are necessary, and would include plain radiography, ultrasound and/or MRI. As in the left ring finger case discussed in this article, general radiography indicated soft-tissue swelling with no underlying bony lesion or arthropathy. With the left ankle case, the X-ray demonstrated a large soft-tissue mass in Kager's fat pad, without lesional fat density, with no evidence of arthropathy. General radiography can provide crucial information, because there may be adjacent cortical erosion with a periosteal reaction underlying the PVNS, in regions without loading.[4]

Ultrasound is particularly useful in the diagnosis of PVNTS. It is able to demonstrate the relationship between the nodules and the adjacent tendon. It allows dynamic assessment, showing whether the lesion is adherent to the tendon sheath, because PVNTS lesions arise from the tendon sheath[3] it moves independently to the tendon during dynamic assessment.[3] The characteristic features of the masses are typically homogeneously, lobulated, hypoechoic with internal vascularity, although in some cases, the masses may appear heterogeneous. The lesion is known to have close contact with a tendon, 6 mm being the average length of tendon contact and the average circumferential tendon involvement being 140 degrees.[21]

The left ring finger case in this article is the extra-articular localised form of PVNTS. Longitudinal ultrasound imaging demonstrated a predominantly heterogeneous, hypoechoic solid mass with multiple lobulations contacting the flexor tendon. Internal vascularity was demonstrated with colour Doppler. The transverse B-mode image demonstrates the solid, homogeneous, hypoechoic mass almost completely encasing the flexor tendons. The colour Doppler image demonstrates the detectable peripheral and central vascularity in the tumour. These tumours are usually superficial, and it is necessary to use high-frequency transducers to optimise the images. In the left ankle case where there was diffuse intra-articular localised PVNS, it is more difficult to delineate the lesion with ultrasound. Ultrasound features of intra-articular PVNS are nonspecific, and include markedly thickened hypoechoic synovium, and heterogeneous echogenic masses.[3] Nevertheless, ultrasound can be used to describe the extent of the tumours. As in the case reported herein, the ultrasound shows a large hypoechoic, heterogeneous mass, appearing to arise from the subtalar joint. The mass is lobulated and irregular, and colour Doppler shows increased perfusion. The Achilles tendon is not involved as the lesion is located within the Kager's fat pad.

Conclusion

The PVNS is a rare benign condition of the synovium that results in formation of villous and nodular protrusions.[1] Imaging provides no absolute characteristic features in the diagnosis of PVNS. Nevertheless, initial complementary imaging examinations are necessary. Ultrasound is particularly useful in the diagnosis of PVNTS because of the nature of the disease having a close intimate relationship to the tendon. The finding of PVNTS should initially be sought via an ultrasound scan and confirmed with histological findings. The diagnosis of the intra-articular localised form and the diffuse form of PVNS should be relied on by utilising MRI. MRI is able to identify the presence of haemosiderin precipitates within the nodules with the disease confirmed by histological examination. Good knowledge of the various types of PVNS and being able to recognise the typical appearance, which reflect the pathologic characteristics, is important for optimal patient management.

References

1 Mukhopadhyay K, Smith M, Hughes PM. Multifocal PVNS in a child – followed over 25 years. *Sketletal Radiol* 2006; **35**: 539–42.
2 Monu JUV, Chew FS. *Pigmented Villonodular Synovitis Imaging*. Medscape, Updated: May 2011; **18**: 2011.
3 Murphy MD, Rhee JH, Lewis RB, Fanburg-Smith JC, Flemming DJ, Walker EA. Pigmented villonodular synovitis: radiologic-pathologic correlation. *RadioGraphics* 2008; **28**: 1493–1518.
4 Godoy FADC, Faustino CAC, Meneses CS, Nishi ST, Goes CEG, Canto ALD. Localized pigmented villonodular synovitis: case report. *Rev bras Ortop* 2011; **46**: 468–71.

5 Barile A, Sabatini M, Iannessi F, Di Cesare E, Splendiani A, Calvisi V, Masciocchi C. Pigmented villonodular synovitis (PVNS) of the knee joint: magnetic resonance imaging (MRI) using standard and dynamic paramagnetic contrast media. Report of 52 cases surgically and histologically controlled. *Radiol Med* 2004; **107**: 356–66.
6 Myers BW, Masi AT. Pigmented villonodular synovitis and tenosynovitis: a clinical epidemiologic study of 166 cases and literature review. *Medicine* 1980; **59**: 223–38.
7 Jaffe HL. Lichtenstein L, Suro CI. Pigmented villonodular synovitis, bursitis and tenosynovitis. *Arch Pathol* 1941; **31**: 731–65.
8 Kransdorf MJ, Murphey MD. Synovial tumors. *Imaging of Soft Tissue Tumors*. Philadelphia: Lippincott Williams & Wilkins; 2006; 381–436.
9 Somerhausen NS, Fletcher CD. Diffuse-type giant cell tumor: clinicopathologic and immunohistochemical analysis of 50 cases with extraarticular disease. *Am J Surg Pathol* 2000; **24**: 479–92.
10 Rao AS, Vigorita VJ. Pigmented villonodular synovitis (giant-cell tumor of the tendon sheath and synovial membrane): a review of eighty-one cases. *J Bone Joint Surg Am* 1984: **66**: 76–94.
11 Dorfman HD, Czerniak B. Synovial lesions. *Bone Tumors*. St Louis: Mosby; 1998; 1061–71.
12 Enzinger FM, Weiss SW. *Soft Tissue Tumors*, 4th edn. St Louis: Mosby; 2008; 1054–6.
13 Hughes TH, Sartoris DJ, Schweitzer ME, Resnick DL. Pigmented villonodular synovitis: MRI characteristics. *Skeletal Radiol* 1996; **24**: 7.
14 Sheldon PJ, Forrester DM, Learch T. Imaging of intra-articular masses. *RadioGraphics* 2005; **25**: 105–19.
15 Bassetti E. Candreva R, Santucci E. Pigmented villonodular synovitis of the knee: a case report. *J Ultrasound* 2011; **14**: 167–9.
16 Sharma H, Jane MJ, Reid R. Pigmented villonodular synovitis: diagnosis pitfalls and management strategy. *Curr Orthop* 2005; **19**: 215–22.
17 Weiss SW, Goldblum JR. Enzinger & Weiss's soft tissue tumors. *Benign Tumor-like Lesions of Synovial Tissue*. Philadelphia: Mosby Elsevier; 2008; 769–88.
18 Kempson RL, Fletcher CDM, Evan HL, Hendrickson MR, Sibley RK. Synovial tumors. *Atlas of Tumor Pathology: Tumors of Soft Tissue*, 3rd series. Washington, DC: Armned Forces Institute of Pathology; 2001; 387–94.
19 Sepega A. Pigmented Villonodular Synovitis (PVNS). [cited 2014 Jul 9]. Available from URL: http://www.kneeandshoulder.md/pvns_01.html.
20 Ushijima M, Hashimoto H, Tsuneyoshi M, Enjoji M. Giant cell tumor of the tendon sheath (nodular tenosynovitis): a study of 207 cases to compare the large joint group with the common digit group. *Cancer* 1986; **57**: 875–84.
21 Middleton WD, Patel V, Teefey SA, Boyer MI. Giant cell tumors of the tendon sheath: analysis of sonographic findings. *Am J Roentgenol* 2004; **183**: 337–9.