

## Extraarticular pigmented villonodular synovitis of the hip

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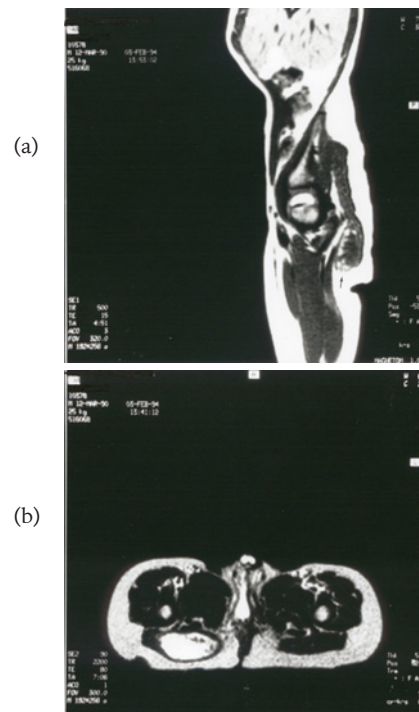
Pigmented villonodular synovitis of any joint in the extraarticular region is very rare. Its clinical and radiological differential diagnosis is difficult due to exhibiting findings of any soft tissue tumor. Here we report an extraarticular pigmented villonodular synovitis case of the hip of a five-year-old boy, with its radiological and histopathological aspects. The mass was completely extraarticular and was identified histologically and radiologically as pigmented villonodular synovitis. Six months after marginal excision, the lesion recurred. The physicians can face such cases of pigmented villonodular synovitis presenting with unusual extraarticular location, and the preferred excision should be wide to avoid possible recurrences.

**Key words:** pigmented villonodular synovitis, hip.

Pigmented villonodular synovitis (PVNS) was first described by Jaffe et al.<sup>1</sup> as a benign proliferative process of the synovial membrane with a potential for local recurrence<sup>2,3</sup>. Since then, many cases of this condition involving different joints, mostly knee, either intraarticularly or extraarticularly, have been reported<sup>2-5</sup>. The purpose of this paper was to represent an extraarticular case of PVNS occurring in the hip of a five-year-old boy.

### Case Report

A five-year-old boy presented with a four-month history of intermittent right hip pain in the posterior region, without any trauma. He was unable to bear full weight on his right leg. The motion of his right hip was decreased minimally and the pain was exacerbated by forced flexion. Roentgenographic and laboratory examinations were within normal limits. Bone scintigraphy showed a marked increase in uptake of Tc-99m in the entire right hip region. On magnetic resonance images, there was an extraarticular mass touching the right gluteus maximus. The mass was mainly hypointense on T1-weighted image (Fig. 1a) and hyperintense on T2-weighted image (Fig. 1b), and well marginated but slightly heterogeneous.



**Fig. 1a, b.** Sagittal T1-(a) and axial T2-weighted (b) spin-echo magnetic resonance (MR) images show a mass touching the inferior portion of the gluteus maximus (arrow). The mass, which is mainly hyperintense on T2-weighted image and hypointense on T1-weighted image, is well marginated and slightly heterogeneous.

An open marginal excision was performed through a posterior incision on right lateral aspect of the sacrum. The surgical findings and the gross macroscopical evaluation of the encapsulated cystic mass, containing a reddish brown fluid, was in favor of PVNS. The mass was 4.5x3.5x2 cm in size, firm and brown. Microscopically, there were marked villous proliferations infiltrated by lymphocytes, plasma cells and giant cells (Fig. 2). Abundance of hemosiderin pigment and some calcifications were noticed in the villous lesion.

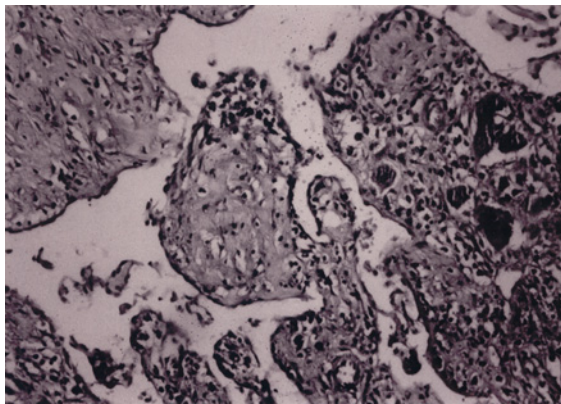


Fig. 2. The lymphocytes, plasma cells and giant cells (inset) infiltrating the villous proliferations (H&E X 200).

Six months later, the patient had no complaint, but there was a mass at the same localization on the ultrasonographic examination that revealed recurrence.

### Discussion

Pigmented villonodular synovitis mostly affects people in the third or fourth decade of life. However, it can also be seen in younger and older individuals, as in the present case. In most series, the hip is reported as the second most common joint affected by PVNS<sup>5-6</sup>.

According to Boyd and Sledge<sup>4</sup>, the specific criteria for PVNS are monarticular involvement, normal bone density, synovial swelling, capsular expansion, juxtaarticular cysts with sclerotic margins on both sides of the joint and slow progression. The diagnosis can be difficult, and delay in diagnosis is common<sup>7</sup>. In a study of Chung et al.<sup>8</sup>, the delay in diagnosis ranged from 2.5 to 11 years after the onset of symptoms.

When diagnosed at the hip, PVNS has been found to be localized mostly intraarticularly, just like the other joints, causing invasion of the

femoral head and neck and acetabulum<sup>2-5,7,8</sup>. Nevertheless, most of the above findings are true for intraarticularly localized mass. De Visser et al.<sup>5</sup> reported that only two cases were extraarticular out of 38 PVNS cases found over 36 years, one being in the inguinal region and the other in the gluteal region. Extraarticular appearance is also very rare in the other joints<sup>9</sup>. Thus, the incidence of extraarticular cases is very rare and cases do not exhibit the findings seen in intraarticular location.

Differential diagnosis of the clinical and radiological features of PVNS includes synovial proliferative disorders such as hemosiderotic synovitis, synovial chondromatosis, crystal synovitis, synovial hemangioma, synovial sarcoma, rheumatoid arthritis, osteoarthritis, infections, gout, and familial Mediterranean fever<sup>2-4,6,10</sup>. In extraarticular location, the whole spectrum of soft tissue tumors must be considered in differential diagnosis.

If calcifications or ossified nodules are visible radiographically, synovial chondromatosis should be considered<sup>2,10</sup>. Positive family history and laboratory findings are helpful to distinguish rheumatoid arthritis, infections, gout and familial Mediterranean fever from PVNS. Nonetheless, as extraarticular PVNS presents with soft tissue mass and swelling only, just like in the present case, clinical differentiation of this form from the other soft tissue tumors is difficult.

Magnetic resonance imaging (MRI) is useful for preoperative, non-invasive diagnosis of PVNS cases<sup>5,10</sup>. The most characteristic MRI finding in PVNS is nodular intraarticular masses of low signal intensity on T1-, T2-, and proton density-weighted sequences; the low signal intensity is due to hemosiderin deposition<sup>10</sup>. Nonetheless, despite hemosiderin, cases without signal drop have been described. The absence of signal drop in these cases could be due to fat, inflammation and edema masking the paramagnetic effect of hemosiderin on all sequences<sup>11</sup>. Presence of nodular hypointense areas on MR images could be highly suggestive of PVNS, but in cases like hemophilic arthropathy, amyloid arthropathy, synovial chondromatosis and gout, low signal intensity areas could also be seen. The signal drops in these cases are mostly due to intraarticular hemorrhage, calcification and synovial depositions. The differentiation of

PVNS from such diseases on the basis of MR findings only may not be possible unless clinical, laboratory and roentgenographic findings are evaluated. Synovial sarcoma, which originates in similar localizations with PVNS, often appears as a heterogeneous multilocular mass with internal septations. The mass is relatively well circumscribed and looks encapsulated. The heterogeneous pattern is due to the presence of hemorrhagic, cystic and necrotic areas<sup>12</sup>. They usually do not contain serial nodular hypointensities, which we observed in our case.

As confirmed with histopathological examination in our case, the lesion is characterized by a fibrous stroma, deposition of hemosiderin, nodules of proliferating collagen-producing polyhedral epithelioid cells, and multinucleated giant cells in the synovial membrane<sup>6,13,14</sup>.

Due to its extraarticular localization and size, we performed open surgery in this case. Because of its proximity to sacral and pudendal nerves, we preferred marginal excision. Avoiding wide excision is likely the reason for the recurrence after six months. Unfortunately, the parents refused a second surgery due to problems with health insurance, so the follow-up period was very short and treatment was not completed.

In conclusion, we wanted to share our clinical and radiological experience with this case. The important message is that the physicians can face such cases of PVNS presenting with unusual extraarticular location. In these cases, histologic and radiologic examinations stand as the major objective criteria in reaching the diagnosis.

In treatment of such cases, the preferred excision should be wide to avoid possible recurrences.

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