CASE REPORT

PIGMENTED VILLONODULAR SYNOVITIS OF KNEE WITH INTRA AND EXTRA-ARTICULAR PRESENTATION – A CASE REPORT

Muraliswar Rao J¹, Rakesh Kumar Nanna², Geetha Vani. P³, Sravanthi V⁴, Swetha A⁵

HOW TO CITE THIS ARTICLE:

Muraliswar Rao J, Rakesh Kumar Nanna, Geetha Vani. P, Sravanthi V, Swetha A. "Pigmented Villonodular Synovitis of knee with intra and extra-articular presentation – A Case Report". Journal of Evolution of Medical and Dental Sciences 2013; Vol. 2, Issue 52, December 30; Page: 10320-10327.

ABSTRACT: Pigmented villonodular synovitis (PVNS) is a benign proliferative disorder primarily occurring in the large joints of the appendicular skeleton such as the knee and hip joints. PVNS is a slow-growing lesion of uncertain aetiology arising from the synovial membrane. It is characterized by villous and nodular overgrowths of the synovial membrane of the bursa or the tendon sheath. Though magnetic resonance imaging (MRI) is considered to be most sensitive in revealing findings consistent with PVNS, these findings are to be correlated with clinical and histopathological findings for precise diagnosis. Here we present a case with painless, non-traumatic swelling of left knee since 2 years, diagnosed as PVNS on plain radiograph, CT and MRI, underwent total synovectomy and was later on confirmed histopathologically.

KEYWORDS: Pigmented villonodular synovitis, Synovial proliferation, Tenosynovitis.

INTRODUCTION: Pigmented villonodular synovitis was first described by Chassaignac¹ in 1852 as a nodular lesion of the synovial membrane that affects the flexor tendons of the fingers. PVNS is seen to affect a wide age range of patients, with no definite sex predilection² and affects young adults in their third or fourth decades.³⁻⁵ It has a high recurrence post-surgery and recurrence rate is between 8- 56%.⁶

CASE REPORT: A 55 year old male patient presented with swelling of left knee since 2 years, not associated with pain and there is no history of trauma. On examination there is a large diffuse soft to firm swelling with nodular surface around the left knee which is non-tender on palpation. There is minimal local rise of temperature noted.

The patient was referred to radiology department where he was subjected to plain radiograph of left knee AP and lateral views which showed diffuse homogenous opacity around the knee predominantly in the supra patellar region anteriorly and in the popliteal fossa posteriorly. Then CT was done which revealed a large lobulated heterodense lesion predominantly isodense to adjacent muscles around knee with intra-articular extension and erosion of tibial plateau. MRI revealed a relatively well defined heterointense lesion in anterior compartment of the lower third of the thigh and posterior compartment of the upper third of the leg, popliteal region and with intra-articular extension. There is evidence of multiple focal cystic and ill-defined areas of hypointensity on all sequences. There is evidence of erosions of the tibial plateau with significant joint effusion.

He was operated and total synovectomy was done. Specimen was sent for histopathological examination. Grossly the specimen appears as multiple nodular grey yellow synovial masses. On microscopy there were multiple nodules, composed of mononuclear cells

CASE REPORT

mixed with foamy macrophages, few multinucleate giant cells and hemosiderin laden macrophages and final histopathological diagnosis given was pigmented villonodular synovitis.

DISCUSSION: Pigmented villonodular synovitis (PVNS) represents an uncommon benign neoplastic process that may involve the synovium of the joint diffusely or focally or that may occur extra-articularly in a bursa (pigmented villonodular bursitis [PVNB]) or tendon sheath (pigmented villonodular tenosynovitis [PVNTS]). The exact etiology of PVNS is ill understood. Inflammatory reaction, neoplasia, hyperplasia, metabolic derangement, and recurrent hemorrhages due to trauma have all been considered possible etiologies.⁷⁻⁹ The current description of this entity was applied since 1941 by Jaffe and colleagues¹⁰, who used the designations pigmented villonodular synovitis (PVNS), pigmented villonodular bursitis (PVNB), and pigmented villonodular tenosynovitis (PVNTS). Previous terminology for PVNS included synovial xanthoma, synovial fibroendothelioma or endothelioma, benign fibrous histiocytoma, xanthomatous giant cell tumor of the tendon sheath, myeloplaxoma, chronic hemorrhagic villous synovitis, giant cell fibrohemangioma, fibrohemosideric sarcoma, sarcoma fusiforme, benign or malignant polymorphous cellular tumor of the synovial membrane, and fibrous xanthoma of the synovial membrane.¹¹⁻¹⁶

The imaging findings of PVNS include:

PLAIN RADIOGRAPH: Radiographic features are nonspecific, and radiographs may appear normal. The diffuse intra-articular form of PVNS often demonstrates a joint effusion and extrinsic erosion of bone on both sides of the joint, but the joint space is unaffected. The localized forms of disease usually reveal only a soft-tissue mass. In 15% of cases there may be some bony changes like an increased density of the synovium or typical radiolucent cystic defects.^{4, 5}

CT: Computed tomography (CT) depicts these lesions as either diffuse thickening of the tissue about the joint (PVNS) or as a localized soft-tissue mass (PVNB or PVNTS). In diffuse PVNS, the attenuation of the lesion may be somewhat increased relative to that of muscle owing to the presence of hemosiderin. Extrinsic erosion on both sides of the joint is well demonstrated by CT and is more frequent in lesions in less capacious joints such as the hip. CT is optimal for demonstrating extrinsic erosion of bone on both sides of the joint, and it may also reveal subchondral cyst formation. Localized PVNS manifests as a nonspecific well-defined soft-tissue mass with attenuation similar to that of adjacent muscle. Mildly increased attenuation is uncommon, a characteristic that likely reflects the more variable amount of hemosiderin in these lesions, compared with diffuse intra-articular PVNS.^{13, 17}

MRI: MR imaging reveals characteristic features of a heterogeneous, diffuse, synovial based, plaque-like thickening that often is associated with nodularity. Associated joint effusion is common, particularly in large joints such as the knee, but the effusion is generally surrounded by thickened synovial rinds of hemosiderin-laden tissue. The signal intensity of the synovial thickening is intermediate to low on T1-weighted images. Low signal intensity also predominates on T2-weighted MR images. This signal feature correlates with the varying amounts of hemosiderin in the lesion^{5, 18-20}, owing to the preferential shortening of T2 relaxation time caused by hemosiderin. Use of gradient-echo pulse sequences allows

CASE REPORT

confirmation of the presence of hemosiderin, which appears as a prominent “blooming” of low signal intensity due to magnetic susceptibility artifact. In cases in which hemosiderin deposition is minimal, the characteristic low signal may not be apparent.^{19, 21} If less hemosiderin is present, signal intensity is more likely to be intermediate between that of diffuse PVNS and that of skeletal muscle.⁴ MRI is particularly useful because it depicts the complex intra-articular relationships of the joint and delineates the nature and extent of soft tissue lesions.²² Detection of disease location and extent are important both for diagnosis and to guide treatment.

CONCLUSION: Pigmented villonodular synovitis (PVNS) represents an uncommon benign neoplastic process of the synovium. It may also involve the bursa, joint and the tendon sheath. Synovial involvement can be diffuse or focal. The knee, followed by the hip, is the most common location for PVNS. The hypertrophic synovium is typically villous, nodular, or villonodular and contains variable amounts of hemosiderin. Hemosiderin deposition occurs in the majority of cases.

IMAGES:

Image 1

Radiograph of left knee AP and lateral view showing diffuse homogenous opacity around the knee, predominantly in the supra patellar region anteriorly and posteriorly in the popliteal fossa.

Images 2 and 3

CT axial images showing a large lobulated heterodense lesion predominantly isodense to adjacent muscles around knee, also seen is the thinned out lower third of vastus medialis muscle (★).

CASE REPORT

Image 4

MRI T1WI coronal image of left knee showing homogenous lesion hypointense to the adjacent muscle predominantly on the medial side of distal third of thigh abutting the femur and displacing the muscles peripherally.

Image 5

Image 5: MRI STIR coronal image at the same section showing the lesion is hyperintense to the adjacent muscle.

Images 6 and 7

MRI T2WI axial and sagittal sections showing a large heterointense lesion predominantly hyperintense to the adjacent muscle noted in the suprapatellar region and popliteal fossa. Few nodular hypointense areas noted within the lesion suggestive of hemosiderin deposits (★)

CASE REPORT

Image 8

MRI proton density sagittal image at the same sections showing the heterointense lesion predominantly hyperintense to the adjacent muscle.

Images 9 and 10

MRI gradient coronal and axial images showing a large heterointense lesion predominantly isointense to the adjacent muscle noted on the medial and posterior aspects of distal third of thigh. Few nodular hypointense areas noted within the lesion, suggestive of hemosiderin deposits (★)

Images 11 and 12

Intra-operative pictures showing thickened synovium with nodular surface and golden-brown in colour in anterior aspect and in popliteal fossa respectively.

CASE REPORT

Image 13

Specimen showing total excised synovium.

Image 14

Showing comparative radiographs of knee before and after surgery.

Images 15 and 16

Histopathological slide showing nodules composed of mononuclear cells mixed with foamy macrophages, few multinucleate giant cells and hemosiderin laden macrophages.

CASE REPORT

REFERENCES:

1. Chassaignac M. Cancer de la gaine des tendons. *Gas HospCivMilit* 1852;47:185–190.
2. Giannini C, Sheithauer BW, Wenger DE, Unni KK. Pigmented villonodularsynovitis of the spine: a clinical, radiological and morphological study of 12 cases. *J Neurosurg* 1996; 84:592–597.
3. Visser ED, Pruszczynski M, Wobbles T, van de Putte LBA. Diffused and localized pigmented villonodularsynovitis: evaluation of treatment of 38 patients. *Arch Orthop Trauma Surg* 1999; 119:401-4.
4. Bojanic I, Ivkovic A, Dotlic S, Ivkovic M, Manojlovic S. Localized pigmented villonodularsynovitis of the knee: diagnostic challenge and arthroscopic treatment: a report of three cases. *Knee Surg Sports TraumatolArthrosc* 2001;9:350-4.
5. Choi N. Localized pigmented villonodularsynovitis involving the fat pad of the knee. *Am J Knee Surg* 2000;13:117-9.
6. Chin KR, Barr SJ, WinalskiC, Zurakowski D, Brick GW. Treatment of advanced primary and recurrent diffuse pigmented villonodularsynovitis of the knee. *J Bone Joint Surg Am* 2002; 84:2192– 2202.
7. Gezen F, Akaz KM, Aksu AY, et al. Spinal pigmented villonodularsynovitis: a case report. *Spine* 1996;21:642–645.
8. Weidner N, Challa VR, Bonsib SM, et al. Giant cell tumour of the synovium (pigmented villonodularsynovitis) involving the vertebral column. *Cancer* 1986;57:2030–2036.
9. Granowitz SP, D’Antonio J, Mankin HL. The pathogenesis and long term results of pigmented villonodularsynovitis. *ClinOrthop* 1976;114:335–351.
10. Jaffe HL, Lichtenstein L, Sutro CJ. Pigmented villonodularsynovitis, bursitis and tenosynovitis. *Arch Pathol* 1941;31:731–765.
11. Weiss SW, Goldblum JR. Benign tumors and tumor-like lesions of synovial tissue. In: Enzinger& Weiss’s soft tissue tumors. Philadelphia, Pa: Mosby Elsevier, 2008; 769–788.
12. Kempson RL, Fletcher CDM, Evans HL, Hendrickson MR, Sibley RK. Synovial tumors. In: Atlas of tumor pathology: tumors of the soft tissues. 3rd series. Washington, DC: Armed Forces. Institute of Pathology, 2001;138:387–394.
13. Kransdorf MJ, Murphey MD. Synovial tumors. In: Imaging of soft tissue tumors. Philadelphia, Pa: Lippincott Williams & Wilkins, 2006; 381–436.
14. Hughes TH, Sartoris DJ, Schweitzer ME, Resnick DL. Pigmented villonodularsynovitis: MRI characteristics. *Skeletal Radiol* 1995;24:7–12.
15. Dorfman HD, Czerniak B. Synovial lesions. In: Bone tumors. St Louis, Mo: Mosby, 1998; 1061–1071.
16. Flandry F, Hughston JC. Pigmented villonodularsynovitis. *J Bone Joint Surg Am* 1987;69:942–949.
17. Jelinek JS, Kransdorf MJ, Utz JA, et al. Imaging of pigmented villonodularsynovitis with emphasis on MR imaging. *AJR Am J Roentgenol* 1989;152: 337–342.
18. Farrokh D, Annaert JM, Fabeck L, Theunis A, Delince P. Localized pigmented villonodularsynovitis of the knee with bone involvement mimicking a benign bone tumor: CT and MR findings. *JBR-BTR* 2001;84:253-5.

CASE REPORT

19. Frassica FJ, Khanna JA, McCarthy EF. The role of MR imaging in soft tissue tumor evaluation: Perspective of the orthopaedic oncologist and musculoskeletal pathologist. MagnReson Imaging Clin N Am 2000;8:915-27.
20. Durr HR, Stäbler A, Maier M, Refoir HJ. Pigmented villonodular synovitis. Review of 20 cases. J Rheumatol 2001;28:1620-30.
21. Bhimani MA, Wenz JF, Frassica FJ. Pigmented villonodular synovitis: keys to early diagnosis. Clin Orthop Rel Res 2001;386:197-202.
22. Hogan CJ, Diduch DR. Patellar instability as a result of localized pigmented villonodular synovitis. Am J Knee Surg 2001;14:259-63.

AUTHORS:

1. Muraliswar Rao J.
2. Rakesh Kumar Nanna
3. Geetha Vani P.
4. Sravanthi V.
5. Swetha A.

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Radiodiagnosis, ASRAM Medical College.
2. Post Graduate, Department of Radiodiagnosis, ASRAM Medical College.
3. Post Graduate, Department of Radiodiagnosis, ASRAM Medical College.
4. Post Graduate, Department of Radiodiagnosis, ASRAM Medical College.

5. Post Graduate, Department of Radiodiagnosis, ASRAM Medical College.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Muraliswar Rao J.,
Associate Professor,
Department of Radiodiagnosis,
ASRAM Medical College,
Eluru, West Godavari (DT),
Andhra Pradesh, India.
Email-muraliradiology@gmail.com

Date of Submission: 28/11/2013.
Date of Peer Review: 29/11/2013.
Date of Acceptance: 10/12/2013.
Date of Publishing: 27/12/2013