

Highly Erosive Tenosynovial Giant Cell Tumor of the Hip Treated with Arthroscopic Synovectomy

ABSTRACT

We present the case of a 28 year old man who presented with chronic hip and groin pain found to be caused by a symptomatic tenosynovial giant cell tumor (TGCT), formerly known as pigmented villonodular synovitis (PVNS), which had eroded into his entire hip joint. Initial radiograph imaging showed rounded lucencies in the acetabulum, femoral head, and femoral neck, followed by an MRI revealing lytic lesions infiltrating the capsule and joint space. The tumor bulk had infiltrated the entire joint, including the central and peripheral compartments. An open approach with total joint replacement was offered, but the patient declined in order to salvage the native joint. Following arthroscopic tumor removal and synovectomy, the patient regained a near-baseline level of function. Importantly, there has been no evidence of tumor recurrence at 36 months follow-up. In conclusion, this case highlights the importance of vigilance for atypical conditions with common presentations. While the patient's TGCT was aggressive and erosive, the initial impression resembled standard intraarticular conditions causing a delay in appropriate diagnosis and treatment.

Tenosynovial giant cell tumor (TGCT), formerly known as pigmented villonodular synovitis (PVNS) is a neoplasm of the synovial lining that is typically seen in the knee; however, the hip is the second most commonly affected joint occurring in as many as 15% of cases.¹⁻⁹ It is classified as either a localized or diffuse type depending on the extent of intra-articular involvement. The localized subtype refers to TGCT that is significant for a mass affecting only a defined aspect of the joint, such as the tendon, while the diffuse subtype affects the entire joint and has a tendency to be more destructive. The latter is of particular concern; it presents with concurrent joint destruction. Lytic enzymes, secondary to the inflamed synovium, begin degrading the articular cartilage and bony structure resulting in a weakened, unstable joint in approximately 90% of cases.¹⁰ Presentation is often non-

Shivam Upadhyaya, BA^{1,2}
 Kyle Alpaugh, MA^{1,2}
 Scott D. Martin, MD¹

AUTHOR AFFILIATIONS

¹Department of Orthopaedics, Brigham & Women's Hospital, Harvard Medical School, Boston, MA

²Boston University School of Medicine, Boston, MA

CORRESPONDING AUTHOR

Shivam Upadhyaya, BA
 Department of Orthopaedic Surgery
 Brigham and Women's Hospital
 75 Francis Street
 Boston, MA 02115
 Phone: 954-829-4330
 Fax: 617-732-9263
supadhyaya@partners.org

The authors report no conflict of interest related to this work.

©2015 by The Orthopaedic Journal at Harvard Medical School

specific and varies depending on subtype with swelling, pain, and mechanical dysfunction as the predominant presenting symptoms.

Grossly, diffuse TGCT resembles an erosive synovial mass with thick strands of inflammation and a dark red to brown mottled coloring, due to hemosiderin.¹⁰ TGCT also exhibits various macroscopic phenotypes with villous, nodular, or combination villonodular morphology.¹⁰

The radiographic appearance of TGCT can also be non-specific including joint effusion, the presence of a soft tissue mass and preservation of joint space early in the disease process that can progress to joint obliteration with extrinsic bone erosion and extension into the femoral head and/or acetabulum. The classic magnetic resonance imaging (MRI) appearance is that of a tumor extending from the synovial lining and demonstrates low-signal (dark appearance) on all sequences. In certain cases, a “blooming artifact” will be seen due to hemosiderin deposition within the tumor.¹⁰ Another pathognomonic finding for the diffuse subtype is extrinsic bone erosion on both sides with a particular predominance in joints that exhibit less space, such as the hip.¹⁰

The current standard of care involves open synovectomy with adjunct radiotherapy. Arthroscopic therapy as management for TGCT and other synovial growth disorders has been a proven modality. A series performed by Byrd et al. showed significant improvement in pain and functionality with low levels of recurrence after a minimum of two years follow up.¹² The technical aspects for the complete survey and removal of TGCT tumor bulks via arthroscopy have been outlined and verified.¹³

CASE REPORT

A 28-year old Indian male presented with one-year history of left hip pain that restricted his activities of daily living. He had been seen one year prior at an outside institution for the same complaint and diagnosed with hip flexor tendonitis. He subsequently left the country for travel and did not receive typical follow-up until his return. At the time of presentation to our clinic, he had undergone one year of physical therapy with no improvement. Initial radiographs taken at his first visit to our clinic showed rounded lucencies in the acetabulum, femoral head, and femoral neck, as well as increased density in the same region most likely representing “bone islands” (Figures 1 and 2). MRI showed

FIGURE 1 Plain hip radiograph of affected (left) hip. Rounded lucencies are seen within the acetabulum, femoral head & femoral neck

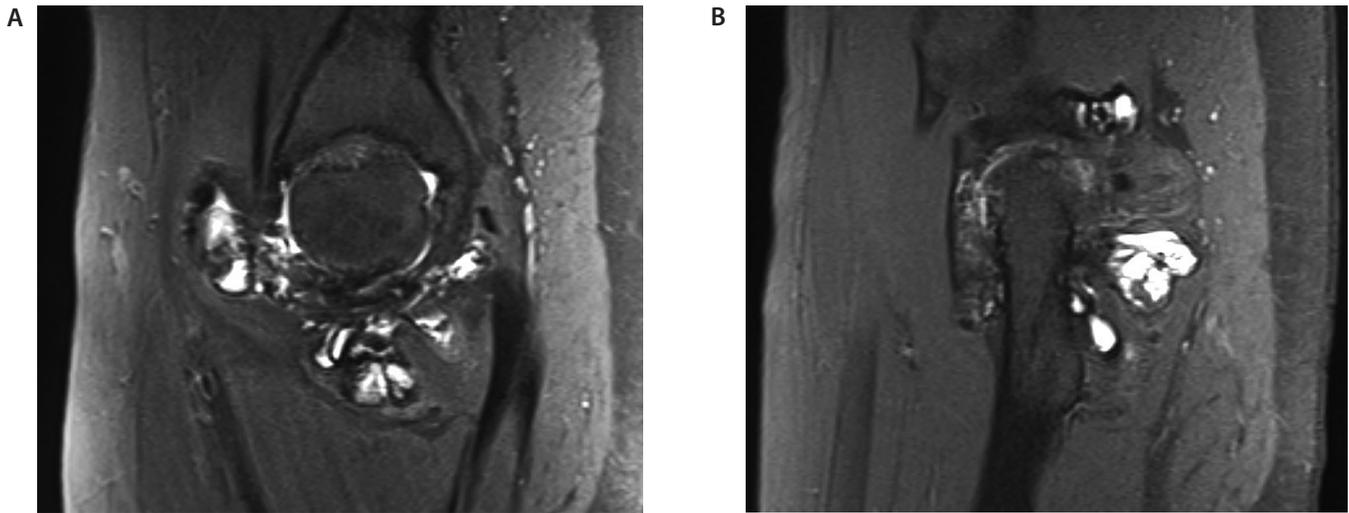


FIGURE 2 Plain pelvic radiograph comparing affected (left) hip to unaffected hip (right)



Obliteration of the joint space on affected side. Proliferation of lytic lesions, especially in the femoral head

FIGURE 3 Sagittal MRI views of the affected hip

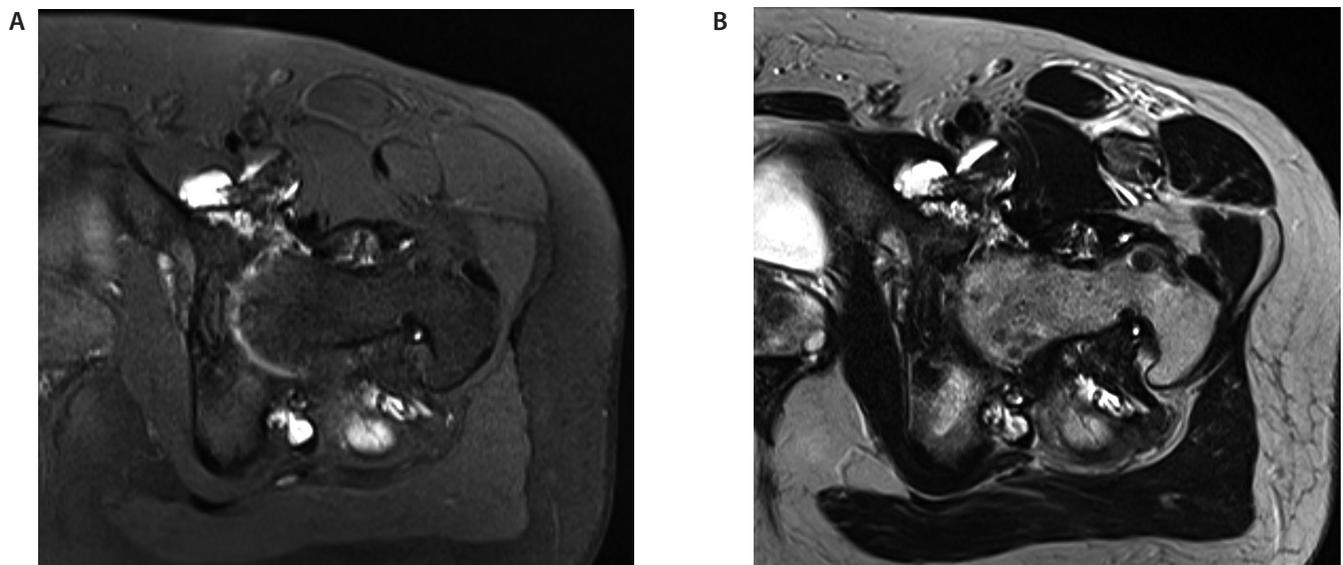


(A) Lytic lesions penetrating the joint capsule and infiltrating entirety of the hip joint including the anterior and posterior aspects of the central and peripheral compartments (B) Demonstrating the extent of posterior infiltration into the peripheral compartment

lytic lesions and erosion of the femoral head extending to the femoral neck with invasion of the joint capsule anteriorly and posteriorly (Figures 3 and 4). Subse-

quently, three diagnostic core biopsy specimens were obtained via computed tomographic (CT) guidance and revealed “diffuse type giant cell tumor, pigmented

FIGURE 4 (A) Axial T1 flare MRI of affected hip shows similar findings as Figure 3 (B) Axial T2 flare MRI of affected hip



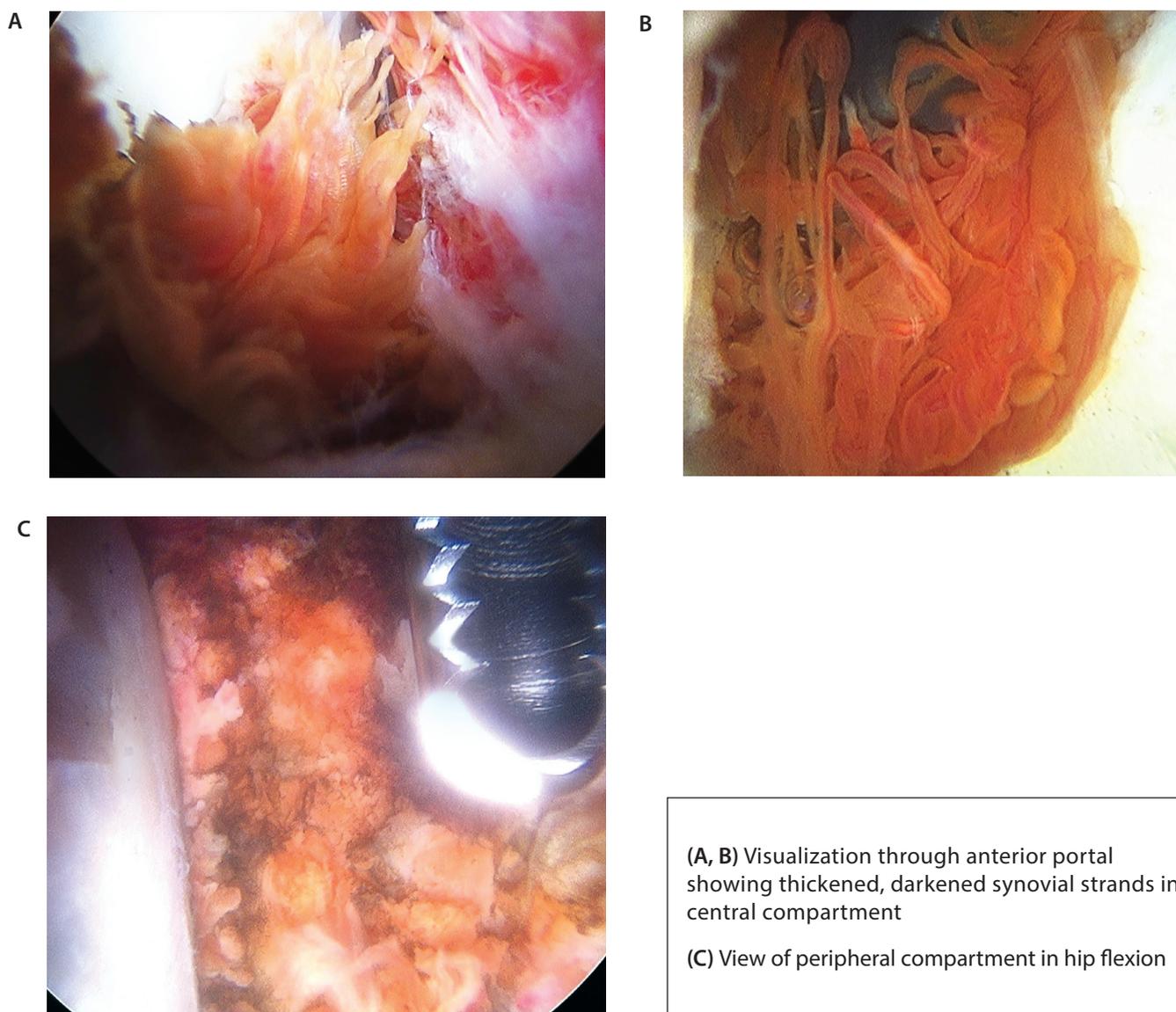
(A) Complete proliferation of the lytic lesions throughout the capsule, central and peripheral compartments (B) Structural damage to the femoral head and acetabulum as demonstrated by the absence of bone due to lytic lesions

villonodular synovitis". We initially recommended the patient undergo total hip arthroplasty with en bloc resection of the neoplasm as a definitive treatment secondary to the severity of joint destruction and erosive quality of the tumor. However, the patient was reluctant to pursue this treatment option due to his young age. The risks and benefits of arthroscopic tumor removal and complete synovectomy were discussed with and understood by the patient, including the risk of tumor recurrence given the severity of pre-existing disease.

Technique

Visualization began with an anterolateral portal, and then under direct fluoroscopic guidance, the following portals were established: mid anterior portal, anterior portal, and proximal mid anterior portals. The patient had diffuse, complete, and abundant TGCT throughout the entire central compartment, including into the pulvinar and the area of the ischium, as well as into the capsular and inferior recesses (Figure 5). With this, the #4.5 shaver was inserted and compartmental areas and surrounding synovium were debrided of tumor followed

FIGURE 5 Intraoperative arthroscopic images of affected hip



(A, B) Visualization through anterior portal showing thickened, darkened synovial strands in central compartment

(C) View of peripheral compartment in hip flexion

by use of an ultrasonic radiofrequency ablator (Arthrex, Naples, Florida) with a Mitek VAPR (DePuy Synthes, Zuchwil, Switzerland) to cauterize. An approach in which separate compartments were engaged was done with complete and total synovectomy and removal of the tumor. The labrum, which was extremely frayed, was slightly debrided as well. A tumor measuring approximately 2.5-cm x 2.5-cm was found in the femoral head (Figure 6). The peripheral compartment was then entered and found to be completely obliterated by tumor. It was addressed in similar fashion, working from the central portion of the peripheral compartment working out into both the medial and lateral gutters, including the medial synovial fold, which was also completely obliterated by tumor. The tumor had also invaded the anterior aspect of the femur at the metaphyseal-diaphyseal junction and the zona orbicularis was completely enveloped with TGCT tumor. Any residual tumor that was found was removed accordingly and the surgery was concluded. He tolerated surgery well and was scheduled for periodic follow up (Figures 7 and 8).

During follow up, the patient had regular radiography that showed grade IV osteoarthritic changes. At 8 months post-op, he began experiencing symptoms, but experienced relief with an injection of

60 mg (1.5 cc) Depo Medrol. He continued physical therapy with no significant residual disease on MRI at 12 and 24 months post-op (Figure 7). At 30 months post-op, the patient continues to improve with physical therapy and subjectively reported he has returned to his original baseline function level, with no findings on plain radiograph at 36 months (Figure 8).

FIGURE 6 Intraoperative view of erosion of femur by the lytic lesions, described as “wormholes”

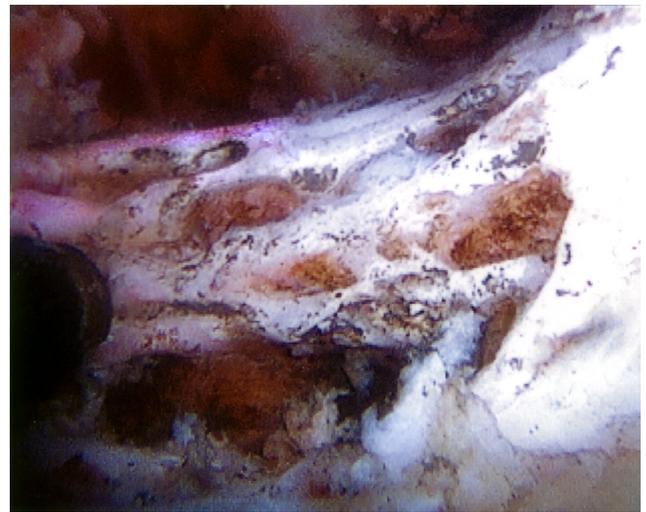
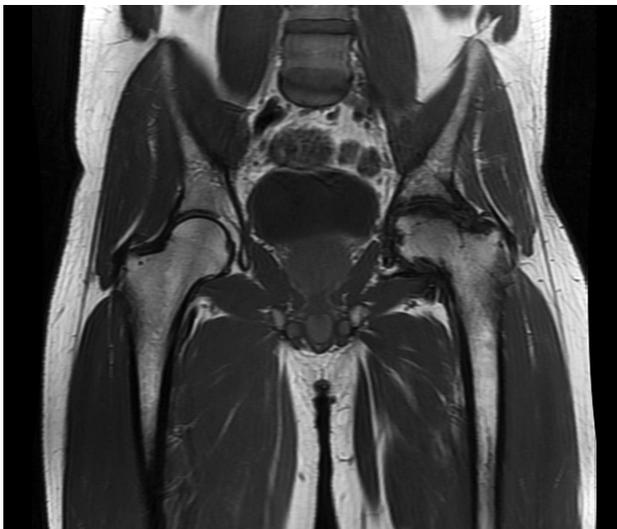


FIGURE 7 Two year follow up fat saturated coronal MRI



Extensive low signal and susceptibility consistent with post-surgical changes, severe loss of cartilage, subchondral sclerosis, cyst formation, femoral head deformity, and osteophyte formation

FIGURE 8 Three year follow up plain radiograph



Severe superior cartilage space narrowing with accompanying sclerosis, small subchondral cystlike changes, large marginal osteophytes, and scalloping of the medial acetabulum

DISCUSSION

We present the first case of diffuse subtype TGCT treated with arthroscopic synovectomy without recurrence at 36 months. TGCT is a neoplastic process with findings of karyotypic aberration and monoclonal lineage in cell lines that affects patients in their third to fifth decade of life.¹ The prevalence of TGCT was estimated at 1.8 million affected individuals by Myers and Masi.⁵ This case was chosen due to the highly erosive pattern it exhibited on presentation. The erosive aspect of this case has been found in a comparative case of TGCT of the shoulder.² Initial MRI and bone scan, combined with CT guided biopsy were sufficient in excluding other etiologies, such as infection. In our case, surgical synovectomy with subsequent tumor removal and debridement of the affected areas was deemed to be sufficient.

The role of complementary therapies, such as external radiotherapy and isotopic synoviorthesis (medical synovectomy), have been studied but their efficacy have not been confirmed.⁶ Radiation is generally reserved for localized, recurrent disease due to the propensity for complications, such as fibrosis and secondary malignancy formation. Medical treatment targeting the identifiers for the origin of the neoplasm has also been introduced with targeting of the macrophage infiltration process that is thought to cause the growth and sustenance of TGCT masses.¹¹ Medical treatment of TCGT has begun in clinical trials and is supported by theoretical modeling based on the universal overexpression of macrophage colony stimulating factor-1 (MCSF-1).¹¹ Tumor grafts were transplanted into rodent models and treated with anti-MCSF-1 monoclonal antibodies resulting in a decrease in macrophage infiltration and contribution to the tumor bulk.¹¹

With hip arthroscopy becoming a more popular modality in the treatment of hip disorders, there are several limitations that should be considered before its utilization. Intra-operative problems and complications such as improper portal placement, lead wire breakage, insufficient tumor bulk reduction/subtotal synovectomy, and lower extremity/pelvic nerve palsy are common issues faced when attempting arthroscopy. These may be secondary to lack of sufficient skill on the part of the surgeon or complicated 3D anatomy due to significant anatomic alteration caused by destruction of the joint unit.

In our case, the patient was able to regain the majority of his functionality after arthroscopic surgery with postoperative physical therapy. The main concern for a more conservative surgical approach was the higher risk of recurrence, which was shown to be approximately 23% in one retrospective study.⁴ Long term follow up (average time from last contact was 7.0-14.5 years) in a cohort of patients with both subtypes showed that the diffuse subtype of TGCT had similar levels of quality of life when compared to the localized subtype.⁸ There was a positive correlation with increased recurrence in patients over time, especially in the diffuse subtype, which showed a recurrence rate of up to 45% compared to the localized subtype's 15% recurrence rate.⁸

CONCLUSION

Due to the nonspecific presentation of this condition, it is imperative that clinicians be vigilant when evaluating a patient with hip pain and atypical image findings. This patient's course highlights the destructive nature of a benign neoplasm in a less commonly affected joint, the hip, with a positive outcome and lack of recurrence after 36 months of follow up.

REFERENCES

1. Chin KR, Brick GW. Extraarticular pigmented villonodular synovitis: a cause for failed knee arthroscopy. *Clin Orthop Relat Res.* 2002 Nov;(404):330-8.
2. Ji, Jong-Hun, Mohamed Shafi, Sang-Eun Park, and Weon-Yoo Kim. Subacromial bony erosion: a rare presentation of pigmented villonodular synovitis of the shoulder. *Knee Surg Sports Traumatol Arthrosc.* 2009 May;17(5):534-8. doi: 10.1007/s00167-009-0752-x. Epub 2009 Feb 28.
3. Kramer, Dennis, Frank Frassica, Deborah Frassica, and Andrew Cosgarea. Pigmented Villonodular Synovitis of the Knee. *J Knee Surg.* 2009 Jul;22(3):243-54.
4. Ma, Xiaomei, Guodong Shi, et al. Pigmented villonodular synovitis: a retrospective study of seventy five cases (eighty one joints). *Int Orthop.* 2013 Jun;37(6):1165-70. doi: 10.1007/s00264-013-1858-9. Epub 2013 Mar 19.
5. Myers BW, Masi AT. Pigmented villonodular synovitis and tenosynovitis: a clinical epidemiologic study of 166 cases and literature review. *Medicine (Baltimore).* 1980 May;59(3):223-38.

6. Park, Geumju, Young Seok Kim, et al. Low-dose external beam radiotherapy as a postoperative treatment for patients with diffuse pigmented villonodular synovitis of the knee. *Acta Orthop*. 2012 Jun;83(3):256-60. doi: 10.3109/17453674.2012.678803. Epub 2012 Apr 11.
7. Vastel, L. Surgical Treatment of Pigmented Villonodular Synovitis of the Hip. *J Bone Joint Surg Am*. 2005 May;87(5):1019-24.
8. Verspoor FG1, Zee AA2, Hannink G2, van der Geest IC2, Veth RP2, Schreuder HW2. Long-term follow-up results of primary and recurrent pigmented villonodular synovitis. *Rheumatology (Oxford)*. 2014 Nov;53(11):2063-70. doi: 10.1093/rheumatology/keu230. Epub 2014 Jun 10.
9. Yudd, Anthony P, and Michael G. Velchik. Pigmented Villonodular Synovitis of the Hip. *Clin Nucl Med*. 1985 Jun;10(6):441-2.
10. Murphey, M., Rhee, J., Lewis, R., Fanburg-Smith, J., Fleming, D., & Walker, E. Pigmented Villonodular Synovitis: Radiologic-Pathologic Correlation. *Radiographics*. 2008 Sep-Oct;28(5):1493-518. doi: 10.1148/rg.285085134.
11. Cheng, Hongwei, Paul W. et al. "Therapeutic Antibodies Targeting CSF1 Impede Macrophage Recruitment in a Xenograft Model of Tenosynovial Giant Cell Tumor." *Sarcoma*. 2010;2010:174528. doi: 10.1155/2010/174528. Epub 2010 Oct 13.
12. Byrd, J., Jones, K., & Maiers, G. (n.d.). Two to 10 Years' Follow-up of Arthroscopic Management of Pigmented Villonodular Synovitis in the Hip: A Case Series. *Arthroscopy*. 2013 Nov;29(11):1783-7. doi: 10.1016/j.arthro.2013.08.002.
13. Lee, S., Haro, M., Riff, A., Bush-Joseph, C., & Nho, S. Arthroscopic Technique for the Treatment of Pigmented Villonodular Synovitis of the Hip. *Arthroscopy Techniques*. 2015 Jan;4(1):e41 - e46. [cited 2015 May 5] Available from: [http://www.arthroscopytechniques.org/article/S2212-6287\(14\)00125-X/abstract](http://www.arthroscopytechniques.org/article/S2212-6287(14)00125-X/abstract)
14. Il-Kyu Kim, Hyun-Young Cho, Hyun-Woo Cho, Ji-Hoon Seo, Dong-Hwan Lee, and Wang Peng. Pigmented villonodular synovitis of the temporomandibular joint - computed tomography and magnetic resonance findings: a case report *J Korean Assoc Oral Maxillofac Surg*. 2014 Jun;40(3):140-6. doi: 10.5125/jkaoms.2014.40.3.140. Epub 2014 Jun 27.
15. Liu YK, Chan JY, Chang CJ, Huang JS. Pigmented villonodular synovitis of the temporomandibular joint presenting as a middle cranial fossa tumor. *J Oral Maxillofac Surg*. 2012 Feb;70(2):367-72. doi: 10.1016/j.joms.2011.02.031. Epub 2011 Jul 13.