

Localized Pigmented Villonodular Synovitis of the Shoulder: A Difficult Diagnosis of a Rare Disorder

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Abstract

Introduction: Localized pigmented villonodular synovitis (LPVNS) (LPVNS) of the shoulder joint is an extremely rare disorder. It is most often associated with a nonspecific clinical presentation resulting in both delays in diagnosis and in treatment. The growth characteristics and natural history of LPVNS are poorly understood.

Case report: This article describes an unusual case of a 53 -year-old woman whose treatment delay allowed us to more closely study the natural history of LPVNS. Our patient first presented with poorly localized posterior shoulder pain. Her symptoms slowly progressed. An initial MRI study more than 2 years after the onset of symptoms demonstrated a soft tissue tumor in a subscapularis recess location. Treatment with corticosteroid injections and physical therapy failed. A second MRI study, nearly 2 years later, found no change in signal characteristics, location, and size measurements of the soft tissue tumor, all important distinctions. Arthroscopic resection produced a definitive diagnosis of an intra-articular localized pigmented villonodular synovitis of the shoulder. At her final 18 mos. follow-up the patient demonstrated pain relief and no clinical recurrence of disease.

Conclusion: To our knowledge, this is the first case report of a nodular appearing LPVNS arising from an intra-articular shoulder location in a patient with no prior shoulder surgery or trauma. It is also unique in that the tumor originated in a subscapularis recess location. This case documents for the first time a LPVNS with limited growth potential and emphasizes the importance of careful direct study and clinical correlation of MRI findings to avoid delays in treatment.

Introduction

A patient presenting with nonspecific shoulder complaints related to a rare disorder creates a diagnostic challenge. The additional lack of physical and radiographic findings often leads to further evaluation with magnetic resonance imaging (MRI). MRI findings can be non-diagnostic and at other times can be overlooked. The failure to recognize and treat a soft tissue tumor can have serious consequences.

The most serious concerns about time delays are in the treatment of soft tissue sarcomas where early diagnosis is critical. Tumor size at the time of discovery is an important prognostic indicator [1]. However, benign soft tissue tumor progression about the shoulder also has consequences. The permanent erosion of bone, joint and soft tissue structures, as well as, progressive rotator cuff tears and neurovascular invasion have been documented [2-11]. The result of this destruction leads to increasing disability.

We present a patient with an unusual source of shoulder pain treated non-operatively for over 2 years despite initial MRI evidence of a soft tissue tumor. The eventual treatment of the condition produced a definitive diagnosis of an intra-articular localized pigmented villonodular synovitis of the shoulder. The delay in treatment provided a unique opportunity to further define the spectrum of clinical presentation and to more closely study the natural history of this rare condition.

Case Report

A 53-year-old right-hand dominate African-American woman was first evaluated in orthopedic consultation in March of 2008 with complaints of shoulder pain. The pain was poorly localized to the posterior aspect of her right shoulder and had progressed slowly over a 1-year period of time. Pain, measuring a score of 3 out of 10 on a Visual Analog Scale (VAS), had been present for over 6 months. The

pain was worsened by daily work activities of typing on a computer and was associated with stiffness. The initial radiographic findings of her right shoulder were reported as “unremarkable”. The patient underwent cortisone injections into the right shoulder on two different occasions with no relief and she failed to improve with a course of physical therapy. She was then offered surgery on her “rotator cuff” but declined.

The patient’s pain progressed and was nearly constant even without activity, averaging a VAS score of 7. A second orthopedic opinion, 20 months after the initial examination, recommended an MRI scan of her right shoulder for further evaluation. An MRI study of January 2010 on a 1.5 Tesla magnet suggested signs of mild impingement. A curvilinear soft tissue mass between the subscapularis tendon and the glenohumeral joint in the location of the subscapularis recess was reported by the radiologist as the dominant finding (Figure 1). Her treating physician at that time overlooked this finding and performed a cortisone injection into the subacromial bursa. This third injection provided no relief. Referral to a shoulder specialist was then recommended.

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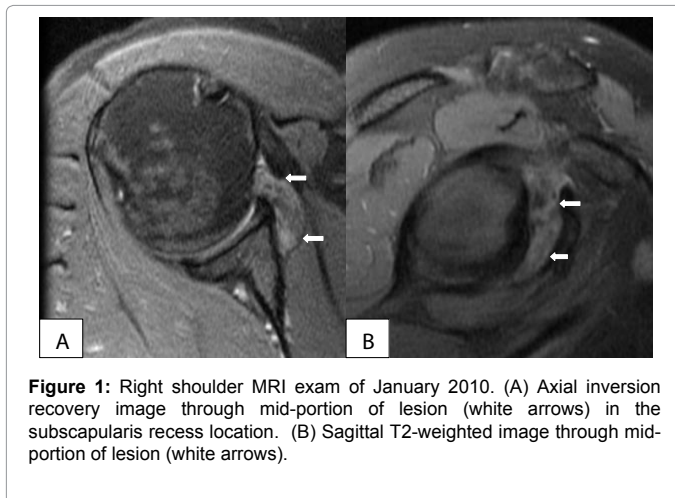


Figure 1: Right shoulder MRI exam of January 2010. (A) Axial inversion recovery image through mid-portion of lesion (white arrows) in the subscapularis recess location. (B) Sagittal T2-weighted image through mid-portion of lesion (white arrows).

Twenty-two months following the initial MRI scan, the patient was evaluated in our clinic. Her symptoms had not progressed during this time. She continued with a deep nonspecific posterior shoulder pain measuring 7 in intensity. She had no history of trauma or prior surgery involving the right shoulder. Her physical examination demonstrated full active range of motion and full strength of all muscle groups of the right shoulder. Impingement signs and instability tests were negative. Slight discomfort to palpation over a subtle fullness in the anterior shoulder region near the lesser tuberosity was noted. There was no posterior shoulder tenderness. Full cervical spine range of motion without tenderness was recorded. Repeat plain radiographs of the right shoulder showed no joint or soft tissue abnormalities, specifically no soft tissue calcifications. Review of her initial MRI confirmed the findings of a soft tissue mass in a subscapularis recess location, but without a specific diagnosis (Figure 1). A second MRI was requested. Her medical history was significant for long standing hypertension and chronic renal failure. She was therefore considered not to be a candidate for intravenous gadolinium enhancement.

This second MRI study confirmed the findings of the original study, again without a specific diagnosis (Figure 2). The MRI details of both studies were compared and demonstrated no measurable interval change. On both exams, a well circumscribed soft tissue mass measured 0.8×2.5×3.4 cm (AP×ML×CC) and demonstrated heterogeneous signal intensity on multiple sequences. All sequences on both exams demonstrated a few punctate foci of low signal intensity suggesting hemosiderin deposition. The findings on both studies were suggestive of pigmented villonodular synovitis.

The patient underwent right shoulder arthroscopy for both diagnostic and therapeutic purposes. The patient was placed in the left lateral decubitus position and then the right shoulder aspirated of nearly 1 cc of normal appearing clear synovial fluid. The viewing camera was placed through a standard posterior portal. An anterior-superior operating portal was established. Joint irrigation was maintained with a gravity system. An extensive examination of the glenohumeral joint was performed. Arthroscopic findings included a soft, apparently encapsulated, pale yellow nodular mass localized to the anterior subscapularis region arising between the middle and the inferior glenohumeral ligaments through a foramen of Rouviere (Figure 3A) [12]. No other arthroscopic abnormalities were noted. The mass was mobilized with a small elevator and clean planes identified. The mass was incised exposing numerous yellow and reddish-brown finger-like projections (Figure 3B).

The patient underwent arthroscopic partial synovectomy and complete mass excision using a motorized arthroscopic resector. All the involved areas were easily accessible and a clean bed of the subscapularis muscle was left. The 6.3×4.2×1 cm aggregate of pink-brown spongy and focally pale yellow rubbery tissue was entirely submitted for histopathological evaluation. The histology of the neoplasm consisted of a mixed proliferation of mononuclear epithelioid and multinucleated osteoclast-like giant cells, foamy (xanthomatous) and focally hemosiderin pigment containing macrophages in variable proportions with a background of focally collagenized stroma (Figure 4). There was no frank hemorrhage or necrosis. The overall gross and histological features were consistent with localized pigmented villonodular synovitis.

In the immediate postoperative period, following a 3 week course of physical therapy, pain relief was excellent. At the 6-month follow-up, the patient felt 95% improved showing a full range of shoulder motion and reported a VAS score of 2. At final follow-up (18 months postoperatively) physical examination showed no evidence of recurrence.

Discussion

Pigmented villonodular synovitis (PVNS) is a benign, locally invasive synovial proliferation of unknown etiology affecting joints, bursae, and tendon sheaths [13]. Each case of PVNS is usually characterized by its site of origin and by its form of growth. Various

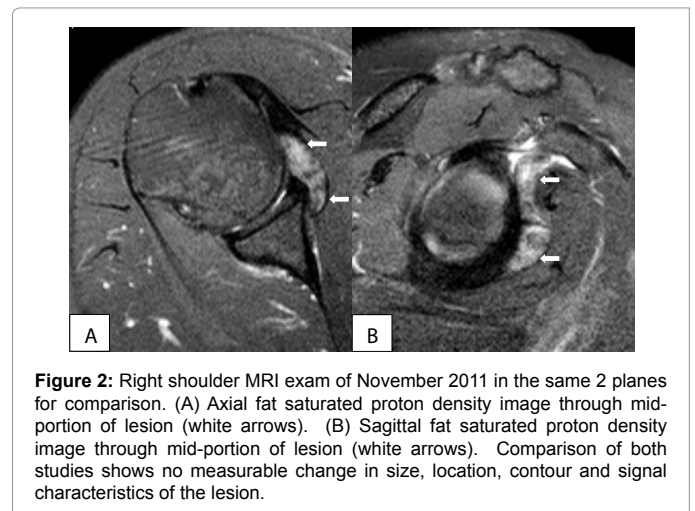


Figure 2: Right shoulder MRI exam of November 2011 in the same 2 planes for comparison. (A) Axial fat saturated proton density image through mid-portion of lesion (white arrows). (B) Sagittal fat saturated proton density image through mid-portion of lesion (white arrows). Comparison of both studies shows no measurable change in size, location, contour and signal characteristics of the lesion.

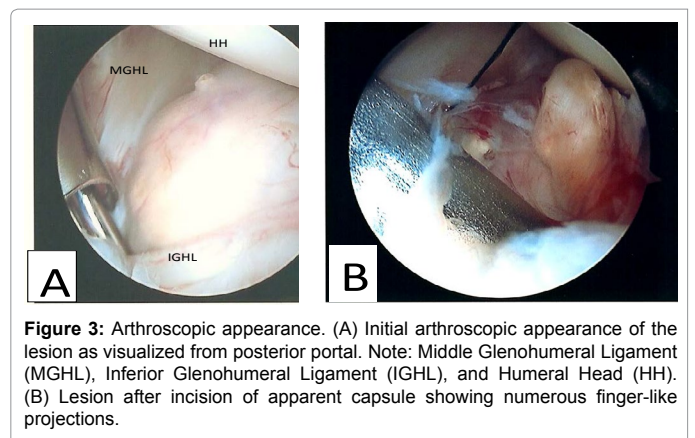
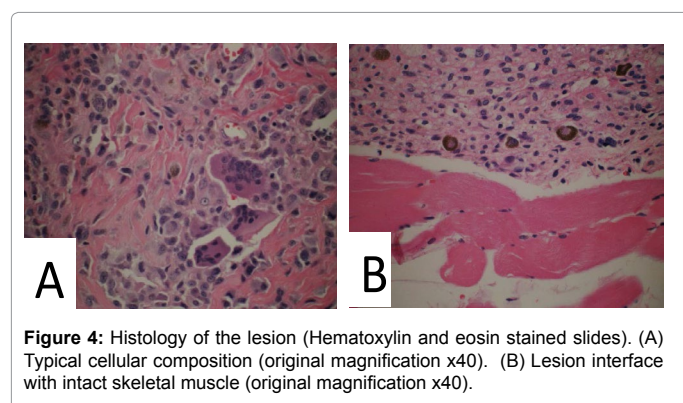


Figure 3: Arthroscopic appearance. (A) Initial arthroscopic appearance of the lesion as visualized from posterior portal. Note: Middle Glenohumeral Ligament (MGHL), Inferior Glenohumeral Ligament (IGHL), and Humeral Head (HH). (B) Lesion after incision of apparent capsule showing numerous finger-like projections.



intra-articular and extra-articular sites of origin including specific bursa locations have been identified. The knee is the most commonly affected joint, but rare cases of shoulder joint involvement have been reported [2-5,7,9-11].

Two primary growth forms of PVNS have been described [14]. The two forms vary significantly in clinical presentation, prognosis, and response to treatment. Consequently, distinguishing between the two different forms is important. A diffuse form (DPVNS) that affects the entire synovial lining is more common and is usually considered more aggressive. A rare focal, or localized, form (LPVNS) frequently presents with a slow progression of symptoms which often adds to delays in diagnosis. Any given lesion of PVNS can be further characterized by its gross appearance. A particular case may show a predominantly villous or a predominantly nodular appearance [15].

LPVNS originating in the shoulder joint appears to be extremely rare. To our knowledge, only 4 cases in the English literature have been reported [5,16-18]. The two earliest reports of these lesions arising in the shoulder provide little clinical information in their descriptions [16,17]. The two most recent case reports provide the best understanding of this rare form [5,18].

In 1997, Cheng et al. reported a case of a 20-year-old man with LPVNS discovered incidentally during surgery on a multiply operated shoulder [18]. Bioabsorbable tacks and nonabsorbable suture material had been used in prior stabilizing operations. Three nodular appearing localized PVNS masses were found in the anterior glenoid, anterior capsule, and axillary pouch locations 33 months after the initial surgery. Histological evaluation including polarized light microscopy showed the classic cytological appearance of PVNS and no evidence of foreign body debris. Despite the absence of foreign material on histology, questions remain about the relationship of foreign material in the pathogenesis of these 3 lesions.

The most recent case report and review of Mahieu et al. published in 2001 appears to be the most detailed in reporting a case of LPVNS originating in the shoulder joint of a 30-year-old woman [5]. Proliferative synovial lesions, grossly villous in appearance, were localized to the inferior and the posteroinferior part of the shoulder joint. All indications were that the tumor began *de novo* in a shoulder with no prior surgery or trauma.

The growth characteristics and the natural history of LPVNS are poorly understood. An analysis of this small group of 4 cases originating in the shoulder joint provides us with a limited overview of this particular form. This group appears demographically diverse, yet they share some important clinical characteristics attributable to

this form of PVNS in this location. This group contains both men and women with ages ranging from 20 to 55 years at presentation. The tumors were found in various locations within the joints. Nonspecific symptoms dominated patient complaints with a range of the onset of symptoms from 24 to 96 months prior to correct diagnosis. Despite demographic differences, all 4 patients had satisfactory outcomes and were tumor free following total resection of localized intra-articular disease by either open or arthroscopic methods [5].

In our case report, we have documented for the first time a nodular appearing LPVNS arising from an intra-articular shoulder location in a patient with no prior shoulder surgery or trauma. Other unique aspects of this case include: 1) a subscapularis recess site of origin of this tumor; and 2) a time delay from initial MRI study to a definitive diagnosis. Together these findings challenged our understanding of this rare disorder and stimulated further analysis.

This is the first report to distinguish a LPVNS originating in a subscapularis recess location. The subscapularis recess has often been referred to as the subscapularis bursa. It is, however, not truly a separate bursa. This unique recess is a synovial lined outpouching that serves to protect the subscapularis tendon as it passes over the neck of the scapula. The recess communicates with the glenohumeral joint through capsular openings that are variably present [12]. De Palma has classified this variable capsular anatomy into 6 different types [19]. Our patient appeared to have Type 2 capsular anatomy with an opening into the joint between the middle and the inferior glenohumeral ligaments known as the foramen of Rouviere.

In contrast, the subacromial bursa is a separate extra-articular structure and is a true bursa of the shoulder. There are only two cases of LPVNS found originating in and isolated to this bursa that have been reported [20,21]. Despite being isolated to the bursa, with no adjacent joint involvement, this tumor was considered to be locally invasive to the surrounding soft tissue and joint if left untreated. This tumor's true natural history is unknown. Regardless of the site of origin, both diffuse and localized forms of PVNS appear to originate from the same synovial cell type [22].

A second distinct aspect of our case is the time delay from initial MRI findings to treatment and a definitive diagnosis. Left untreated, LPVNS continues to cause pain and discomfort, thus limiting activity and function. No studies have examined the long-term outcomes of patients with LPVNS who are left untreated [23]. During a treatment delay of more than 2 years; we found no change in size, location, contour and signal characteristics of the lesion. Clinically, there were no corresponding changes in symptoms. We considered several possible explanations for this.

We propose that synovial cells of the subscapularis recess were stimulated to grow by a benign neoplastic or reactive process [24]. Our findings would strongly suggest that the tumor grew along a path of least resistance in the confines of the subscapularis recess anatomy and into the joint through the foramen of Rouviere. After reaching a certain size the tumor stopped growing. We found no evidence of vascular compromise or lack of nutritional support to account for this. Recent evidence suggests that under certain circumstances pressure from surrounding healthy tissue can inhibit tumor growth [25]. We speculate that the pressure from the surrounding local anatomy to be the major factor in limiting the growth of this tumor. Alternatively, the tumor itself may simply have had limited growth potential.

Plain radiographs rarely reveal any abnormalities in cases of LPVNS. Normal radiographs can help rule out pathologies, such as

osteoarthritis, rheumatoid arthritis and gout. Occasionally, radiographs do reveal soft tissue shadows or bony erosions [4,5]. These findings are nonspecific and the differential diagnosis is extensive.

Shoulder ultrasonography appears to have limited value in the evaluation process. These exams detect gross structural findings including rotator cuff pathology and assist with localization of disease. They are not capable of detecting the presence of hemosiderin and do not offer specific diagnosis [24]. A recent case in which ultrasonography and repeat ultrasonography were used demonstrated findings of subacromial bursitis/synovial membrane proliferation, but appeared to only delay the more specific final diagnosis [21].

Magnetic resonance imaging is much more sensitive for diagnosing LPVNS. In general, the MRI characteristics of PVNS can vary related to the amounts of hemosiderin, fibrous tissue, protein, lipid, water and cellular material. However, the MRI findings of prominent low signal intensity (seen with T2-weighting) and “blooming” artifact from the hemosiderin (seen with gradient-echo sequences) are nearly pathognomonic of this diagnosis.

The characteristics of this particular lesion are suggestive of but not diagnostic of Localized pigmented villonodular synovitis (LPVNS). The differential diagnosis for this lesion also includes synovial hemangioma, synovial chondromatosis, siderotic synovitis and hemophilic arthropathy. Gout, rheumatoid arthritis, amyloid arthropathy and degenerative joint disease are less likely given the localized lesion, lack of extra-articular involvement and absence of bony erosions.

Despite presenting with a somewhat characteristic appearance of PVNS on MRI, a definitive diagnosis could not be made in this case prior to surgical intervention and review of the histopathology. However, the value and accuracy of two MRI studies in assisting with the location and characterization of this tumor should be emphasized. MRI is helpful in distinguishing between DPVNS and LPVNS, determining the presence of extra-articular involvement, as well as providing valuable information such as lesion size and location.

Independent of etiology, it appears that complete marginal excision is the treatment of choice for the localized form of PVNS [5,23]. We conclude arthroscopic synovectomy is the preferred surgical option for cases of intra-articular LPVNS of the shoulder. Arthroscopy allowed excellent visualization and extensive exploration of the glenohumeral joint. At the same time, it allowed full access for complete removal of the tumor without the increased morbidity of an open procedure.

Finally, in this case report we have uniquely identified pathology involving the subscapularis recess as the source of poorly localized posterior shoulder pain. Clinicians should keep this in mind, as well as a variety of neck, upper spine, and shoulder conditions, when challenged by a patient presenting with this type of pain.

Conclusions

Rare disorders of the shoulder girdle including neoplasms are often diagnosed with significant delay. This occurs most often in patients who present with a slow, chronic progression of symptoms. The nature and the location of the disorder may produce nonspecific symptoms and little if any physical findings. A high degree of clinical suspicion is most important in preventing delays in diagnosis. The early use of MRI in evaluating these types of patients can lead to a timely diagnosis and treatment. However, a process that includes careful direct study and clinical correlation of MRI findings is critical. Despite the difficulties and delays in diagnosis often associated with cases of LPVNS involving

the shoulder, it appears that patients treated surgically have consistently good to excellent results with no recurrences.

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References

1. Gustafson P (1994) Soft tissue sarcoma. Epidemiology and prognosis in 508 patients. *Acta Orthop Scand Suppl* 259: 1-31.
2. Dorwart RH, Genant HK, Johnston WH, Morris JM (1984) Pigmented villonodular synovitis of the shoulder: radiologic-pathologic assessment. *AJR Am J Roentgenol* 143: 886-888.
3. Sawmiller CJ, Turowki GA, Sterling AP, Dudrick SJ (1997) Extraarticular pigmented villonodular synovitis of the shoulder: a case report. *Clin Orthop Relat Res* 335:262-267.
4. Müller LP, Bitzer M, Degreif J, Rommens PM (1999) Pigmented villonodular synovitis of the shoulder: review and case report. *Knee Surg Sports Traumatol Arthrosc* 7: 249-256.
5. Mahieu X, Chaouat G, Blin JL, Frank A, Hardy P (2001) Arthroscopic treatment of pigmented villonodular synovitis of the shoulder. *Arthroscopy* 17: 81-87.
6. Huang TF, Wu JJ, Chen TS (2004) Bilateral shoulder bursal osteochondromatosis associated with complete rotator cuff tear. *J Shoulder Elbow Surg* 13: 108-111.
7. Ji JH, Shafi M, Park SE, Kim WY (2009) Subacromial bony erosion: a rare presentation of pigmented villonodular synovitis of the shoulder. *Knee Surg Sports Traumatol Arthrosc* 17: 534-538.
8. Trajkovski T, Mayne IP, Dehesi BM, Ferguson PC (2011) Synovial chondromatosis of the shoulder: open synovectomy and insertion of osteoarticular allograft with internal fixation to repair intraoperative glenohumeral joint instability. *Am J Orthop* 40: E154-E158.
9. Petsatodis G, Karataglis D, Kapoutsis DB, Papadopoulos P, Christodoulou AG (2011) Hemiarthroplasty for pigmented villonodular synovitis of the shoulder: a report of two cases. *J Orthop Surg (Hong Kong)* 19: 116-119.
10. Park JH, Park JW, Shin JS, Lee JM, Lee JI (2012) Hemiarthroplasty in a patient with pigmented villonodular synovitis of the shoulder. *Orthopedics* 35: e104-107.
11. Gumina S, Carbone S, Campagna V, Castagna A, Rocca CD, et al. (2013) Pigmented villonodular synovitis of the shoulder associated with massive rotator cuff tear treated by arthroscopic synovectomy and debridement. *Musculoskelet Surg* 97: 79-84.
12. Zlatkin MB, Hoffman CJ, Needell S (2003) Shoulder anatomy. In: Zlatkin MB, (eds) *MRI of the Shoulder*. (2nd edn.) Lippincott, Williams & Wilkins, Philadelphia, PA, USA pp- 96.
13. Jaffe HL, Lichtenstein L, Sutro CJ (1941) Pigmented villonodular synovitis, bursitis and tenosynovitis. *Arch Pathol* 31:731-765.
14. Granowitz SP, D'Antonio J, Mankin HL (1976) The pathogenesis and long-term end results of pigmented villonodular synovitis. *Clin Orthop Relat Res* : 335-351.
15. Byers PD, Cotton RE, Deacon OW, Lowy M, Newman PH, et al. (1968) The diagnosis and treatment of pigmented villonodular synovitis. *J Bone Joint Surg Br* 50: 290-305.
16. Johansson JE, Ajjoub S, Coughlin LP, Wener JA, Cruess RL (1982) Pigmented villonodular synovitis of joints. *Clin Orthop Relat Res* : 159-166.
17. Schwartz HS, Unni KK, Pritchard DJ (1989) Pigmented villonodular synovitis. A retrospective review of affected large joints. *Clin Orthop Relat Res* : 243-255.
18. Cheng JC, Wolf EM, Chapman JE, Johnston JO (1997) Pigmented villonodular synovitis of the shoulder after anterior capsulolabral reconstruction. *Arthroscopy* 13: 257-261.
19. DePalma AF, Callery G, Bennett GA (1949) Variational anatomy and degenerative lesions of the shoulder joint. *Instr Course Lect* 6: 255-281.
20. Cho CH, Sohn SW, Song KS, Kang CH, Min BW, et al. (2008) Extra-articular pigmented villonodular synovitis of the subacromial space. *Orthopedics* 31.
21. Madruga Dias J, Costa MM, Duarte A, Pereira da Silva JA (2013) Localized

- Pigmented Villonodular Synovitis of the shoulder: a rare presentation of an uncommon pathology. *Acta Med Port* 26: 459-462.
22. O'Connell JX, Fanburg JC, Rosenberg AE (1995) Giant cell tumor of tendon sheath and pigmented villonodular synovitis: immunophenotype suggests a synovial cell origin. *Hum Pathol* 26: 771-775.
23. Tyler WK, Vidal AF, Williams RJ, Healey JH (2006) Pigmented villonodular synovitis. *J Am Acad Orthop Surg* 14: 376-385.
24. Perka C, Labs K, Zippel H, Buttgerit F (2000) Localized pigmented villonodular synovitis of the knee joint: neoplasm or reactive granuloma? A review of 18 cases. *Rheumatology (Oxford)* 39: 172-178.
25. Montel F, Delarue M, Elgeti J, Malaquin L, Basan M, et al. (2011) Stress clamp experiments on multicellular tumor spheroids. *Phys Rev Lett* 107: 188102.