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Case Report

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Management of Recurrent Diffuse Pigmented Villonodular Synovitis: A Case Report

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Abstract

Introduction: Pigmented villonodular synovitis is a rare disorder that is a benign, proliferative type of lesion of synovial tissue as well as bursal and tendon sheath tissue. There are two types: localised and diffuse, of which the diffuse variety has a higher recurrence rate. The sheer size and recurrent nature of the lesion in our case presented a challenge to effective treatment. Various treatment strategies have been tried to completely and effectively excise the mass and prevent recurrence.

Case: A 38-year-old male patient, who had previously undergone arthroscopic debridement of the left knee, presented with pain and swelling in the ipsilateral knee. Relevant investigations were done, and he was diagnosed with recurrent diffuse pigmented villonodular sinus.

Surgery was planned, anteriorly a medial parapatellar approach was employed and posteriorly an inverted L-shaped Burks and Schaffer approach was utilised and the mass of proliferated synovium excised. Histopathological examination revealed a diffuse proliferation of mononuclear cells with interspersed giant cells, confirming the diagnosis of a tenosynovial cell giant cell tumour. Once the surgical incisions had healed, the patient underwent radiotherapy using the 3DCRT technique. At a year and a half post-surgery, the patient is symptom-free.

Conclusion: Recurrent diffuse tenosynovial giant cell tumour is an uncommon entity and needs to be treated aggressively to prevent recurrence while maintaining the functionality of the knee joint. A dual open approach and postoperative adjuvant radiotherapy is a safe and effective way to execute this.

Keywords: Pigmented villonodular sinus; Recurrent; Diffuse; Tenosynovial giant cell tumour

Introduction

Pigmented villonodular synovitis is a rare disorder that is a benign, proliferative type of lesion of synovial tissue as well as bursal and tendon sheath tissue [1]. There are two types: localised and diffuse. The diffuse variety has a higher recurrence rate. Current treatment options include arthroscopic or open debridement. External beam irradiation or internal irradiation is an adjuvant treatment to decrease the rate of local recurrence.

Case Report

A 38-year-old male presented to the orthopaedics outpatient department with a gradually progressive swelling in the left knee for three years, which extended like a horseshoe shape above the knee. There was no prior history

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of trauma. The patient had undergone an arthroscopic synovial debridement 2 years before the current scenario and was symptom-free for 15 months after that. He had begun experiencing increasing pain for 2 weeks and a recurrence of the diffuse swelling of the knee. Examination revealed a diffuse swelling of the knee, more prominent in the suprapatellar and parapatellar fossa. There was a fullness adjacent to the patella tendon. Posteriorly, there was a diffuse bulge in the popliteal fossa. The swelling was doughy in consistency. There was tenderness most pronounced in the suprapatellar region. There was no redness or engorged veins. The range of motion of the involved knee was decreased- 100° compared to the normal knee -140°. Neurological examination did not reveal motor or sensory deficits. All peripheral pulses were well palpated. The radiographs showed no significant changes except soft tissue swelling [Figure 1]. MRI of the knee showed extensive synovial thickening, hypointense areas and septations within the knee joint, which was suggestive of synovitis, likely pigmented villonodular sinus [Figure 2].

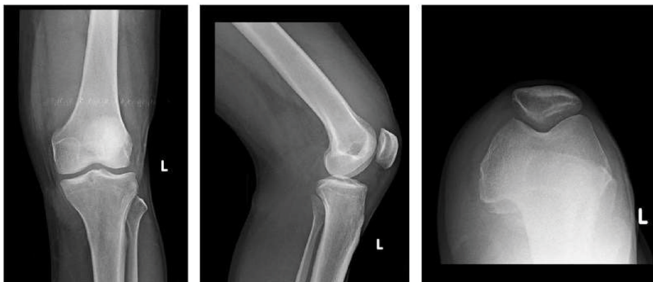

Figure 1: Pre-operative radiographs showing soft tissue swelling primarily in the supra- patellar region, most evident on the lateral projection (centre).

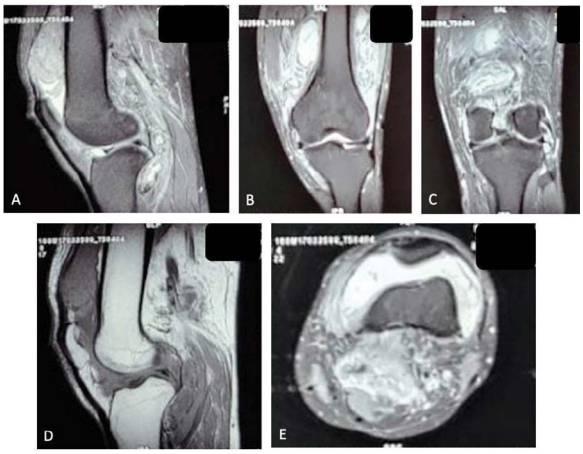

Figure 2: MRI Images showing extensive synovial thickening, hypointense areas and septations within the knee joint suggesting synovitis. A, D – sagittal T2 weighted and T1 weighted MRI respectively, showing diffuse involvement of synovium extending into the prepatellar area. B,C – Coronal T2 weighted images showing diffuse involvement on both sides of the femur and supracondylar area. E- Axial T2 weighted MRI showing a nearly circumferential involvement around the femur.

The patient was taken up for surgery. A pneumatic tourniquet was used high on the thigh, and anteriorly, a medial parapatellar approach was used and posteriorly, a Burks and Schaffer inverted L-shape approach was used with the longitudinal limb over the medial area overlying the medial head of the gastrocnemius below the popliteal crease. The typical 'Crankcase oil' synovial fluid was immediately evident. An exuberant mass of deep brown, lobulated synovial tissue was noted extending into the medial and lateral parapatellar recess and superiorly into the popliteal fossa [Figure 3].

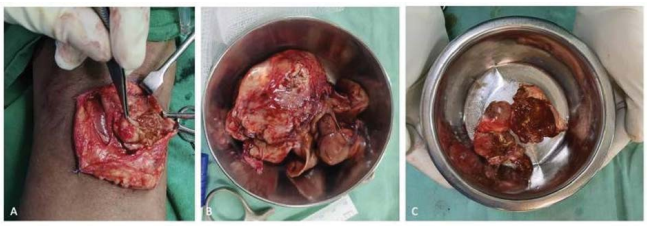

Figure 3: Deep brown pigmented synovium A- Thickened synovium seen through posterior approach. B- Specimen removed from anterior approach. C- Specimen removed from posterior approach.

The histopathological specimen showed tissue fragments lined by synovial epithelium with underlying diffuse proliferation of uniform mononuclear cells with moderate eosinophilic cytoplasm. A few osteoclast-type giant cells were noted. Focal stromal hyalinisation is noted. Admixed are sheets of foamy histiocytes, thin-walled capillaries, pigmented macrophages and mild lymphocytic infiltrate. These changes were suggestive of a diffuse type- tenosynovial giant cell tumour (villonodular synovitis) [Figure 4].

Postoperatively, the patient underwent radiotherapy after the surgical site wounds healed. He received adjuvant radiotherapy of 45 Gray in 25 fractions using the 3DCRT technique. There were no postoperative wound complications. Initially, the patient had some stiffness at a terminal range of motion. Physiotherapy was continued with range of motion exercises and continuous passive motion. At a one-and-a-

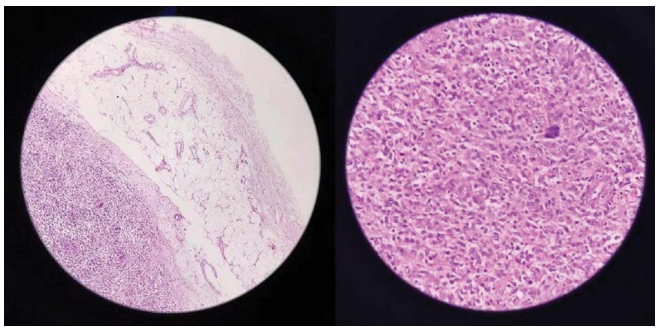

Figure 4: Histopathological specimen showed tissue fragments lined by synovial epithelium with underlying diffuse proliferation of uniform mononuclear cells with moderate eosinophilic cytoplasm. A few osteoclast-type giant cells were noted. (Left – 10x magnification, Right – 40x magnification)

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half-year follow-up, the patient was symptom-free, regained full range of motion and was able to squat on the floor. [Figure 5].

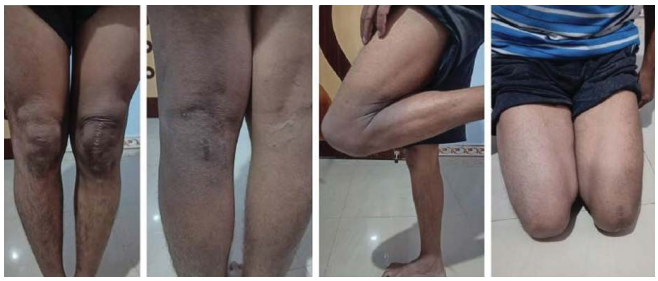

Figure 5: At a one-and-a-half-year follow-up, the patient could completely flex the knee and squat.

Discussion

Pigmented villonodular synovitis (PVNS) is a benign, locally aggressive disorder characterised by the proliferation of synovial tissue and deposition of hemosiderin [1]. It is also known as a tenosynovial giant cell tumour and is associated with the chromosomal translocation t(1;2) (CSF-1;COL6A3) which leads to colony stimulating factor 1 (CSF 1) overexpression [2-5]. Although PVNS is a rare disorder that can affect any joint, it most commonly affects the knee. Within the knee, the most common region affected is the synovium overlying the medial meniscus anterior horn [2]. Repetitive trauma and haemarthrosis are likely etiological factors, although it is largely idiopathic. Histologically, macrophage-like characteristics in fibroblasts and synoviocytes are postulated to occur due to the presence of large amounts of iron [3]. Although benign, Wang et al reported on PVNS with lymph node involvement [6]. There are two variants: a localised (LPVNS) and a diffuse variant (DPVNS), of which the latter is the rarer [1]. DPVNS is more aggressive, can extend beyond the joint cavity and has a higher propensity for recurrence. The diffuse variety involves almost the entire knee's synovium [2]. Extraarticular spread occurs in certain patterns and most commonly involves the posterolateral area [7]. Extracapsular spread per se does not influence recurrence rates [8].

MRI of the knee will show a 'dark-on-dark' appearance on both T1 and T2 weighted images and may demonstrate 'blooming' on gradient echo sequences due to the presence of hemosiderin [2,3].

Irrespective of the type, treatment includes complete resection of the mass, and an incapacity to do so adequately will render the patient prone to recurrence [1]. The goal is to prevent joint destruction, reduce pain and maintain the functionality of the joint, thereby improving the patient's quality of life [2,8]. Once articular damage destruction occurs, arthroplasty may be required [8]. The rate of the requirement for total knee arthroplasty due to PVNS is said to be between two and ten percent [8]. Resection of the mass can be done as an open approach, arthroscopically using an anterolateral, anteromedial, posterolateral and posteromedial portal or a combined approach [1,9]. Song et al. described a technique in the arthroscopic treatment of PVNS which they called a total synovial peel [10]. Some may even consider a staged approach, rather than address both approaches in a single sitting [3]. An arthroscopic approach tends to cause less soft tissue destruction and quicker recovery, although it's known to be particularly difficult to access extraarticular spread and the posterior extent of the disease [1,8]. However, in our case with diffuse PVNS, a dual open approach was chosen for a more thorough excision to decrease the risk of recurrence. The current view is that recurrence is the result of an inability to completely excise the diseased synovium [10]. Although an open approach is known to cause knee stiffness and haemarthrosis, which translates to a longer hospital stay and rehabilitation period, we had no such complications in our case. Often, the extent of the disease is the primary indication for an open approach [1,8]. The problem with post-operative stiffness is subjecting the patient to the requirement of an additional procedure - manipulation under anaesthesia [2] Kerschner et al. showed that the functional outcome scores of an all-arthroscopic or combined approach were comparable, indicating that an open approach, although more invasive, does not necessarily hamper the knee's functionality [8]. Although an open approach may seem altogether more thorough, studies have shown up to an eight per cent recurrence in a dual open approach to DPVNS [2]. In addition to the necessity to address the symptoms themselves, another problem of recurrence is damage to the articular surface, which may push the patient closer to the requirement of a joint replacement procedure [8] Instability may require a Lemaire procedure to be done [11].

The posteromedial (Burks and Schaffer) approach is a safe and easy approach and has less morbidity [12]. It involves an inverted L-shaped incision with the longitudinal limb along the medial side of the transverse limb. The incision can also be extended superiorly along the lateral end of the transverse limb in an overall S-shaped manner [2].

Radiotherapy is a modality that can be used either in isolation or as an adjunct to surgery [8]. Some studies show that radiation therapy may not be a necessary adjunct to surgery for mere DPVNS [1]. However, studies have shown a decrease in recurrence rates when post-operative adjuvant radiation is used [2,13] Koutalos et al. used the 3DCRT technique after a staged dual open excision, however, they had encountered significant post- operative stiffness [3]. Radiation itself can sometimes cause skin erythema, skin fibrosis, hyperpigmentation, lymphedema, articular damage and arthrofibrosis [3,8]. Injection of yttrium 90 in particular has been associated with skin ulceration [14] Radiosynoviorthesis has also been described to treat PVNS

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[15]. Fang et al. described a case similar to ours, where PVNS had recurred initially, but was successfully treated with radiotherapy [16].

Imatinib mesylate is an inhibitor of tyrosine kinase that is being investigated for its role in preventing the recurrence of PVNS. Cairalizumab and Emactuzumab are monoclonal antibodies that are CSF-1 receptor inhibitors currently being studied for use in PVNS [8].

Conclusions

Recurrent diffuse pigmented villonodular synovitis is a rare disorder that can be safely and effectively treated with a dual open approach and postoperative adjuvant radiotherapy.

Statement of informed consent

The informed consent was taken from the patient for submission of the case details to this journal.

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