

Pigmented villonodular synovitis of knee joint: A case report

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World Journal of Advanced Research and Reviews, 2021, 09(03), 346–349

Publication history: Received on 16 February 2021; revised on 20 March 2021; accepted on 22 March 2021

Article DOI: <https://doi.org/10.30574/wjarr.2021.9.3.0112>

Abstract

Diffuse type tenosynovial giant cell tumor (D-TGCT) is a locally aggressive benign proliferation. Knee joint is the most commonly affected site. We present a case report with a 16 months follow up after recurrent surgery and postoperative radiotherapy.

Keywords: *Pigmented Villonodular Synovitis; proliferative disorder; radiotherapy; giant cell tumor*

1. Background

Pigmented villonodular synovitis (PVNS) is a proliferative disorder of unknown etiology and rare disease arising from synovial cells of joint capsules, bursae, and tendon sheaths that can cause limitation of the joint mobility [1]. Chassaignac in 1852 described it as a nodular lesion in the flexor tendon sheath of the fingers [2], and the term PVNS was introduced by Jaffe et al. in 1941 [3].

Depending on the location and extent of synovial involvement, two types can be differentiated: the localized type (L-PVNS) and the diffuse type (D-PVNS) [4], also-called diffuse type tenosynovial giant cell tumor (D-TGCT) for lesions originating from the tendon sheath [5].

The D-TGCT is generally benign, locally aggressive neoplastic proliferation. It arises intraarticular within large joints or extraarticular invasive tumors of tendon sheath, bursa or soft tissue origin [1]. The pathogenesis is due to balanced translocation involving 1p13 [CSF1] in most tumors [6]. Its annual incidence is 1.8 cases per million, with average age of 35 years, range from 1st to 7th decades [7]. There is a slight predominance in the female population [8].

The lesion is restricted to one joint in the majority of cases. The knee joint is the site most commonly affected (75-80%) followed by the hip (15%) then ankle, elbow, shoulder, temporomandibular joint and spine if intraarticular. In the extraarticular type also the knee region is the most affected site followed by foot, wrist, inguinal, elbow region and digits [9].

Usually the patient presents with painful mass with long duration and decreased range of motion.

The lesion is diagnosed by Magnetic Resonance Imaging (MRI) with low signal on T1 and T2, and shows contrast enhancement [10].

Surgical excision is the primary treatment in the form of synovectomy, which is rarely complete especially in large joints [11]. Despite its benign histological character, it has a high rate of recurrence in up to 55% of patients that mandate

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recurrent surgery [12]. Consequently adding external beam radiotherapy has been considered as an adjuvant treatment in extra-articular involvement, residual or recurrent disease or as a primary treatment for inoperable disease. Total dose in the range of 30-36 Gy is recommended [13]. Relapsing and uncontrolled tumor can be treated by TKI (tyrosine kinase inhibitor) e.g. CSFR1 inhibitor [14] or imatinib [15].

2. Case report

We present a thirty seven years old female patient presented on July 2019. Her condition started in 2015 with right knee pain; it was firstly diagnosed as inflammation. MRI right leg was done in March 2019 which revealed moderate joint effusion with suspected pigmented villonodular synovitis and septated popliteal cyst.

In April 2018 patient underwent right knee endoscopic surgery and the pathology revealed pigmented villonodular tenosynovitis.

She started Physiotherapy then after 4 months she started to feel increasing pain in the knee joint.

In July 2019 a new MRI right leg showed extensive periarticular intra synovial bodies of low T1 and T2 signal, the largest one seen anteriorly measuring 3.5x2.3cm, with associated synovial thickening and enhancement.

In August 2019 patient underwent second operation which pathology revealed pigmented villo-nodular tenosynovitis.

Patient experienced improvement regarding pain and limitation of movement. The decision was to give post-operative radiotherapy.

Patient was simulated in supine position with feet first. Serial CT cuts were taken with 3 mm thickness. CTV and PTV were contoured guided by the preoperative MRI, and the patient received 3D conformal radiotherapy technique with 6MV photon to a total dose of 30 Gy/ 15 fx ended in September 2019 and was well tolerated.

After 16 months follow up, patient is still free of disease.

3. Discussion

Several studies have reported good results after a combination of synovectomy and external beam radiotherapy with recurrence rates of less than 20% [13].

The German Cooperative Group on Radiotherapy in Benign Diseases (GCG-BD) [16] studied a pattern-of-care analysis of the results of adjuvant radiotherapy from 14 radiotherapy departments for PVNS in 41 patients. The radiation dose ranged from 30 to 50 Gy. Local control was achieved in 95% of patients, and most patients (82.9%) had either no or slight functional impairment.

De Carvalho et al. [17] studied patients diagnosed with DPVNS treated with surgery followed by adjuvant radiotherapy. Recurrence was observed in 1 patient (12.5%) at 8.6 years follow-up after external beam radiotherapy in 8 patients. Patient received 2000cGy dose over 10 fractions of radiotherapy.

Blanco et al [18] described a series of 22 patients with knee DPVNS, recurrence was observed in 3 patients (13.6%) after arthroscopic synovectomy combined with postoperative external beam radiotherapy to a total dose of 2600 cGy.

Berger et al [19] added external beam radiotherapy in the range of 30 to 50 Gy after open synovectomy in 7 patients. They reported no recurrences, 1 patient complains about joint stiffness.

Nassar et al. [20] did not report recurrence in 12 patients received postoperative radiotherapy with an average follow-up of 27 months.

Chen et al. [21] reported 5 recurrences among 19 patients after 98 months of follow up. Griffin et al. [14] determined only 2 recurrences out of a series of 49 patients.

The DEGRO guidelines [22] for the radiotherapy of non-malignant disorders recommend a total dose range of 30-36 Gy with CT based treatment planning assisted by MRI for covering the entire synovial space.

4. Conclusion

Diffuse type tenosynovial giant cell tumor (D-TGCT) is a locally aggressive benign proliferation that necessitates surgery followed by postoperative radiotherapy in certain conditions to avoid the high rate of local recurrence.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study for case reviewing and publication.

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