

CASE REPORT

RADIOTHERAPY FOR PIGMENTED VILLONODULAR SYNOVITIS: A CASE REPORT

Navin Nayan¹, Vikas K Jagtap²

Author's Affiliations: ¹MD trainee; ²Assistant Professor, Department of Radiation Oncology, Dr. B. Borooah Cancer Institute, Guwahati, Guwahati

Correspondence: Dr Navin Nayan Email: doctornnayan@gmail.com

ABSTRACT

This is a case report of a 13 year old boy who has been operated for Pigmented Villonodular Synovitis (PVNS) and treated with adjuvant post-operative radiotherapy for incomplete surgical resection. This report mainly highlights the role of radiotherapy in managing this rare benign condition with emphasis on improving local control rates with functional joint preservation and avoiding repeated surgeries.

Keywords: Radiotherapy, PVNS (Pigmented Villonodular Synovitis), Surgery

INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a rare proliferative disorder of the synovium affecting any joint but frequently found in the knee. PVNS is an extremely rare condition affecting only 1.8 people per million.¹ Therefore, treatment data are relatively sparse, especially concerning the use of radiotherapy or other modalities (RT).

CASE REPORT

A 13 year old boy with no significant past medical history presented with a history of, gradually increasing left knee joint swelling since last 7 year. It was also associated with on & off pain with limitation of left knee joint movements. He denied any history of major trauma. He was evaluated for the same by orthopedic surgeon and diagnosed with diffuse PVNS after MRI of left knee joint. He underwent arthroscopic synovial biopsy plus total synovectomy of left knee in June 2015. The histopathology was reported as Pigmented Villonodular Synovitis (PVNS). The postoperative MRI of left knee showed minimal residual disease in the peri-articular region, posteriorly. He was referred to Oncology Department for post-operative radiotherapy. He was treated with External Beam Radiotherapy (EBRT) with cobalt machine (⁶⁰Co- γ rays) with antero-posterior opposed fields to a total dose of 36 Gy in 18 fractions. He tolerated the treatment well without any side effects during the course of radiotherapy. The patient also attended school on daily basis during the EBRT schedule. He completed treatment on 5th October, 2015. The patient was on regular follow up since then at our institute. As per the case record the follow up MRI was

done on 16th April 2016 which showed very good regression of lesion compared to previous MRI dated Jun 2015. However, mild synovial effusion and irregularity of Hoffa's fat pad is seen in the scan. Clinical examination revealed mild restriction in flexion movement. Clinically there is no significant joint pain or swelling with almost normal joint activity. He is advised to attend clinic regularly for follow up and necessary evaluation.

DISCUSSION

In 1941, Jaffe et al defined pigmented villonodular synovitis (PVNS), bursitis, and tenosynovitis as yellow-brown villous or nodular, tumor like lesions containing closely-compacted polyhedral cells, multinuclear giant cells, lipoid cells and hemosiderin-containing cells. It is known to be a benign proliferative disorder of the synovium. Lesions can be classified into localized and diffuse forms. The localized form is often effectively treated with local excision. In contrast, diffuse PVNS is typified by widespread involvement of the synovium with villous and coarse nodular outgrowths.

Diffuse PVNS is usually treated with total or near total synovectomy and have a much higher rate of recurrence, often requiring multiple surgeries and additional therapy. The goals of surgery for patients with diffuse intra-articular PVNS are to restore function of the joint and to prevent the destruction of articular cartilage by completely resecting the diseased synovium. Although, the treatment of choice is surgical excision, complete resection may be difficult depending upon the area to be resected (partial versus complete synovectomy) and the joint in-

involved. Local control rate for surgery alone varies from 44-92%.² Postoperative radiation can improve the local control rate (more than 90%). Radiation therapy may be used as the primary treatment modality for diffuse intra-articular PVNS, but it is considered to be better option for postoperative cases with total or incomplete resection. Radiation can be administered either by external beam radiotherapy (EBRT) or through intra-articular injection of radioactive isotopes also known as radiosynoviothe-sis.^{3,4} Intra-articular injection of a radioactive isotope uses several β -emitting isotopes which are colloid molecules bound to a bulky molecule to minimize resorption and leakage. But due to the cost of drug, availability and need for invasive procedures to inject the drug into the joint space, EBRT has been used widely as an easier and cheaper approach.

External beam irradiation is usually employed after the incomplete resection of disease. Radiation doses ranging from 20 Gy to 50 Gy are administered in 15–25 fractions, beginning about 6–8 weeks following surgery.⁴ The optimal dose and fractionation schedule of radiation for PVNS is unclear. Some have reported using doses as low as 16 – 20 Gy without recurrence, whereas others have used doses as high as 50 Gy without complications. Myers et al recommended a dose of 36 Gy with conventional fractionation.¹

Positive surgical margin, history of previous surgery and joint location are the reported factors that increase the likelihood of recurrence.^{5,7} PVNS recurs more frequently in the knee, owing to the large capacity of the joint and the difficulty in performing a true total synovectomy. The final pathway of PVNS of knee is total joint destruction leading to joint replacement. Incomplete synovectomy leads to higher recurrence rates and same has been reported in literature.^{8,9} (56% and 50%, compared with those for complete resection 7% and 20%, respectively). Addition of radiotherapy can minimize these recurrences with less need for repeat surgery. Postoperative external beam radiotherapy has surely shown to minimize recurrence rates in young healthy individuals. Blanco and colleagues reported 14% recurrence rate for patients who underwent external beam irradiation after partial synovectomy, a rate that was comparable to the recurrence rate for patients who underwent total synovectomy for diffuse intra-articular PVNS.¹⁰ In our case the patient had a diffuse type PNSV with incomplete surgical excision. So the deci-

sion was taken to treat with postoperative radiotherapy considering the young age and functional preservation of joint activity.

CONCLUSION

Pigmented villonodular synovitis is a rare benign proliferative disorder of the joints. Local recurrence rates are common after surgery especially in cases with incomplete resections. Postoperative adjuvant external radiotherapy is safe and effective adjuvant modality to improve the local control rates with reduction in repeated surgical procedure and maintaining functional joint capacity.

REFERENCES

1. Myers BW, Masi AT. Pigmented Villonodular Synovitis and Tenosynovitis: A Clinical Epidemiological Study of 166 Cases and Literature Review. *Medicine (Baltimore)* 1980; 59:223-38.
2. Mohler DG, Kessler BD. Open synovectomy with Cryosurgical Adjuvant for Treatment of Diffuse Pigmented Villonodular Synovitis of the Knee. *Bull Hosp Jt Dis* 2000; 59:99-105.
3. Wiss DA. Recurrent Villonodular Synovitis of the Knee: Successful Treatment with Yttrium-90. *Clin Orthop Relat Res* 1982; 169:139-44.
4. O'Sullivan B, Cummings B, Catton C, et al. Outcome following radiation treatment for high-risk pigmented villonodular synovitis. *Int J Radiat Oncol Biol Phys* 1995;32:777-786.
5. Flandry F, Hughston JC. Pigmented villonodular synovitis. *J Bone Joint Surg Am* 1987;69:942-949.
6. Schwartz HS, Unni KK, Pritchard DJ. Pigmented villonodular synovitis: a retrospective review of affected large joints. *Clin Orthop Relat Res* 1989; 247:243-255.
7. Bravo SM, Winalski CS, Weissman BN. Pigmented villonodular synovitis. *Radiol Clin North Am* 1996; 34:311-326.
8. Ogilvie-Harris DJ, McLean J, Zarnett ME. Pigmented villonodular synovitis of the knee: the results of total arthroscopic synovectomy, partial arthroscopic synovectomy, and arthroscopic local excision. *J Bone Joint Surg Am* 1992;74:119-123.
9. De Ponti A, Sansone V, Malcher M. Result of arthroscopic treatment of pigmented villonodular synovitis of the knee. *Arthroscopy* 2003;19:602-607.
10. Blanco CE, Leon HO, Guthrie TB. Combined partial arthroscopic synovectomy and radiation therapy for diffuse pigmented villonodular synovitis of the knee. *Arthroscopy* 2001;17:527-531.