

Pigmented villonodular synovitis, radiotherapy after synvectomy

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ABSTRACT

Introduction: Pigmented villonodular synovitis (PVS) is rare, benign and proliferative disorder of synovium which consists of villous and/ or nodular formations in joints. Diagnosis is determined by clinical features, aspiration of the joint, radiographic features and magnetic resonance imaging (MRI). The definitive treatment is the complete resection of affected tissue by open or arthroscopic synovectomy. Radiotherapy is recommended as an additive treatment in the control of recurrent or refractory disease.

Case Presentation: we report a case of young woman with biopsy proven of PVS who after total synvectomy was referred for adjuvant radiotherapy. she received 36 Gy in 18 fractions and 12 month after treatment she has no sign of recurrence.

Discussion: PVS is a rare benign disorder of synovium which is managed with surgical resection. Radiotherapy can be used as an additional treatment to increase local control.

Keywords: *Pigmented villonodular synovitis; radiotherapy, synvectomy*

Abbreviations: *RT=radiotherapy; PVS =pigmented villonodular synovitis;
MRI=magnetic resonance imaging*

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Case presentation

In September 2011, a 28 years old woman was referred to radiation oncology department of Cancer institute. Her chief complaint of right knee swelling has been started a year ago. She noted pain on movement and slight limitation of motion. She had no history of prior repeated trauma to the involved joint.

MRI evaluation demonstrated a lesion with hemosiderin in the right knee which suspected to be pigmented villonodular synovitis, as conservative therapy and analgesics have made no improvement. In July 2011, synovial biopsy was performed.

Biopsy specimen consisted of creamy yellow rubbery tissue fragments with size of 2.5cm×3.5cm. Histological evaluation showed closely packed medium-sized polyhedral cells with a variable admixture of giant cells. Papillary projections containing foamy cells and hemosiderin-laden macrophages were reported and diagnosis was pigmented villonodular synovitis. Patient underwent total arthroscopic synovectomy. She was referred to our center for additive therapy after 3 weeks.

She reported no history of heavy exercise. She had no prior rheumatologic disease or similar pain in other joints. She denied history of specific disease in the family. On the observation, the appearance of the joint was normal without color changing. Arthroscopy related scar was seen on the knee. The manual examination of the knee revealed a full range of motion although she used a cane because of postoperative pain. All the motion examinations were normal in the affected knee.

After explaining to the patient about the rarity of disease and lack of standard treatment, radiotherapy option was suggested. Through patient agreement two opposed anterior – posterior field was placed with cobalt unit (*Image 1*). Treatment continued to the total dose of 3600 cGy in 18 fractions in 25 days 5 times a week.

At the end of treatment patient had no specific complaint. There was not any skin discoloration or decreased knee movement. We followed the patient every 3 month for 12 months. On the last follow up there was not any side effect or limitation in range of knee motion and also no abnormal finding on MRI.



Image 1

Discussion

Pigmented villonodular synovitis (PVS) is rare, benign, locally invasive and proliferative disorder of synovium which consists of villous and or nodular formations in joints, tendon sheaths, and bursa.^{1,2}

It has been divided into two subtypes: localized (nodular) and diffuse. The nodular variants usually involve fingers but also occur in the larger joints, especially in the knee. The diffuse form is typically a monoarticular disease and the knee is the most affecting joint in this type. It has chronic course and disease symptoms progress slowly. The most clinical presentation is painful joint swelling, however on occasion it is painless. The appearance of aspirated joint fluid is usually xanthochromic and occasionally bloody. There is not any specific changes in the synovial fluid analysis and routine peripheral blood tests.^{2,3}

Diagnosis is determined by clinical features, aspiration of the joint, radiographic features and magnetic resonance imaging (MRI) which have characteristic findings for PVNS. T2 imaging demonstrates a heterogeneous signal intensity which consists of high signal intensity with linear patterns of low signal intensity. T1 weighted

images show the same lesion with low to medium signal intensity (*Image 2, 3*). In the radiography, cystic erosion is a characteristic feature without any calcification, sclerosis or bone demineralization on both sides of joint. There is no change in the joint space.^{4,5}

Histological samples show synovial proliferation with several villi and folds which sometimes fuse and form nodular lesions. Also proliferation of mononuclear cells, infiltration of lipid-laden histiocytes and multinucleated giant cells in a fibrous stroma within the synovium and deposition of hemosiderin have been reported in the biopsy specimens.⁴

The definitive treatment is the complete resection of affected tissue by open or arthroscopic synovectomy. Diseased synovium remains in the limited surgery and increases the rate of recurrence especially in diffuse form. RT is the choice option for adjuvant therapy where achievement of complete excision is difficult. Radiotherapy is recommended as an additive treatment in the control of recurrent or refractory disease.⁵

In a literature review postop 7 case of diffuse pigmented villonodular synovitis who underwent RT was reported from university of Tübingen in Germany. All patients had undergone radical open or arthroscopic synovectomy. RT was performed with a mean dose of 40 Gy in 20 fractions. At the assessment; they found no evidence of recurrence or persistent disease in any patient. All patients were asymptomatic except one who had persistent restriction of joint movement. This study reflected the efficacy and safety of postoperative RT for diffuse pigmented villonodular synovitis.⁷

Melissa Horoschak et al have reported a series of seventeen patients with 18 irradiated sites of localized or diffuse PVNS in the Department of Radiation Oncology at Stanford University between 1993 and 2007. They applied surgical excision or synovectomy before radiotherapy in 16 of the 18 sites. Two of the 18 sites (both primary presentations) had no cytoreductive surgery prior to radiotherapy, which showed growth after radiotherapy. They concluded that postoperative external beam radiation is effective in preventing disease recurrence when is offered following maximal cytoreduction.⁸

Molly Schnirring-Judge and Bonnie Lin reported a 36-year-old, African American female with confirmed PVNS in the ankle joint. The patient underwent synovec-

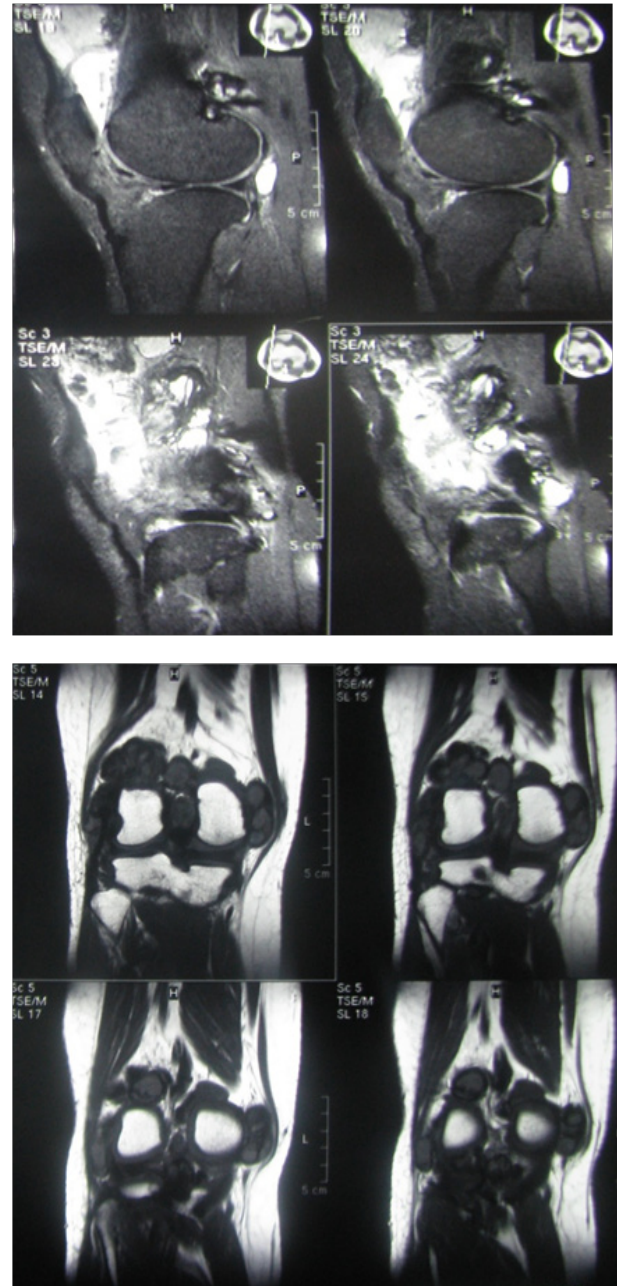


Image 2 and 3: MRI images of PVS sagittal and axial and coronal view

tomy with arthroplasty and was then treated with radiation. No evidence of recurrence was found in the 8 years period of postoperative follow up.⁹

M. Lee et al reported on their experience with seven patients with histologically confirmed diffuse pigmented villonodular synovitis in a prospective study at the Royal National Orthopedic Hospital, Stanmore. The patients

were treated with open synovectomy and adjuvant radiotherapy with a total dose of 35 Gy in 20 fractions. All cases had incomplete resection margins. With an average follow up time of 24 months, all patients, except one, had improvement in their signs and symptoms although all patient remained free of recurrence. On the basis of this research, combined approach was postulated to decrease the risk of recurrence without functional impairment.⁶

Despite lack of knowledge about PVS and rarity of the cases, most studies confirmed a good role for radiation with or without any surgical manipulation. Our case similarly gave good responses with radiation as an adjuvant therapy. With 12 months follow up, the patient was recurrence free with no radiation side effect. Of course case series and longer follow up should be designed to consider the efficacy of radiotherapy in this benign disease.

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