Pigmented Villonodular Synovitis of the Ankle: Radiation Therapy as Primary Treatment to Reduce Recurrence: A Case Report

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Purpose
Pigmented villonodular synovitis (PVNS) is an uncommon proliferative disease of joints, usually affecting synovial and tendon sheath. Of these lesions, 2% occur in the foot and ankle. PVNS has a high rate of recurrence, up to 45%. Radiation has been suggested as a means to reduce recurrence. Traditional treatment includes synovectomy with arthrolysis of the affected joint. We present a case of PVNS where the patient was treated in two stages: Stage I - surgical resection of the tumor and arthrolysis of the ankle (see [Fig 1 and Stage II] - External beam radiation therapy.

Case Study
A 36 yrs African-American woman complained of an insidious onset of swelling and burning in the right ankle, progressively worsening over 1.5 yrs. She denies history of trauma. With prolonged weight bearing she feels a "burning" sensation in the ankle and often feels a "crunching" from the ankle. She has morning stiffness that is accompanied by a tightness and shooting sensation radiating into the foot. The pain occurs even when not weight bearing. She feels pressure in the ankle when she is lying in bed at night and this interferes with her ability to sleep. The intensity of her pain was graded, using the ankle pain scale, as 1/10 when not weight bearing and 7/10 when at work in the factory.

A foot and ankle specialist rendered a diagnosis of rheumatoid arthritis of the right ankle joint. Physical therapy modalities and an ankle brace were prescribed but failed to relieve her symptoms. She presented to the authors’ office for evaluation and treatment recommendations for non-surgical chronic ankle pain.

A rheumatoid panel was drawn failing to support that diagnosis. An MRI confirmed the diagnosis of pigmented villonodular synovitis (PVNS). An oncology consultation suggested and encouraged surgical excision in combination with radiation therapy to reduce the risk of recurrence of this destructive process of bone and joint.

Surgical excision of the lesion and ankle arthrolysis was followed by radiation therapy of 34 Gy in 18 doses over a three week period. Beginning 5 hrs after the initial dose involved with oral AIF. At 7 yrs follow up MRI failed to reveal evidence of recurrent disease.

Radiographic Evaluation
Plain radiographs reveal joint space narrowing in all orthogonal planes. [Fig 1.2]
Other significant findings are subtle including a positive posterior ankle sign on the lateral ankle view as well as a positive synovectomy revealing the anterior aspect of the tibiotalar joint and the division of the talus neck. [Fig 1]

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Surgical Findings
The tibia overlying the mass was perforated by villous and nodular proliferations with hemosiderin deposits characteristic of the disease. [Fig 6]

This infiltrative inflammatory process has eroded the anterior aspect of the tibiotalar joint consistent w/ radiographic findings. [Fig 7] Arrow head at the tibial plateau and arrow at the talus neck.

MRI Evaluation
T1 image: Reveals the low signal intensity of a large multilobulated mass situated in the anterior aspect of the talo-plantar joint, filling the anterior recesses of the joint. A multilobulated soft tissue mass is seen within the posterior ankle capsule. [Fig 5]

STIR images: Reveals intermediary edema throughout the talus and talar articular. The fluid within the posterior ankle space appears to communicate with the increased masses noted on plain films. [Fig 4]

Discussion
Pigmented villonodular synovitis has been described as a progressive and destructive inflammatory, inflammatory condition affecting both peripheral and large joints. The tumor grows and infiltrates the joint capsule and the periarticular structures. It is believed the condition to be rheumatic in origin while others believe this is a focal inflammatory response initiated by trauma. It is interesting to note that the majority of articles in the current literature suggest this condition affects patient’s ranging from 20 to 50 yrs of age. The authors were able to cite 47 articles which suggests that this condition affects a much wider patient population. Based upon a meta analysis of those articles in the English language 11–50 yrs of age more accurately describes the age range of reported cases in the current literature. This statistic may support the notion that the condition is more likely due to chronic inflammatory association with trauma or repetitive injury.

Although the etiology remains uncertain there is a consensus that proliferation therapy is beneficial to subject to surgical excision and debulking. We present a case using this technique in an attempt to treat this debilitating disease.

Consultations from oncology and rheumatology were insightful pre-operatively. Based upon a 7-year clinical follow up and updated MRI evaluation the authors support the use of adjunctive radiation therapy for surgical excision and debulking for PVNS. Although large group studies would be more valuable we feel this case affecting an otherwise healthy and active female may provide insight for others faced with this challenging condition.

References
Pigmented villonodular synovitis (PVNS) was first termed by Jaffe et al. in 1941 and is an uncommon proliferative disease usually affecting the synovium of the joints and tendon sheaths, resulting in effusions and bony erosion. Only 2% affect the joint.