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

Molly Judge, DPM, FACFAS and Bonnie Lin, DPM

St. Vincent Charity Hospital, Section of Podiatry, Division Orthopedics, Cleveland, Ohio

Purpose

Pigmented villonodular synovitis (PVNS) is an uncommon proliferative disease usually affecting synovial joints and or tendon sheaths. 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 arthroplasty 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 arthroplasty of the ankle joint and **Stage II: E**xternal beam radiation therapy.

Case Study

A 36 y/o 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 "feverish" sensation in the ankle and often can feel 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 analog 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 reduce her symptoms. She presented to the author's office for evaluation and treatment recommendations for recalcitrant chronic ankle pain.

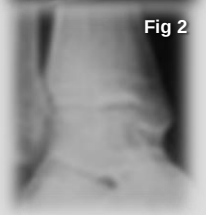
A rheumatoid panel was drawn failing to support that diagnosis. An MRI confirmed the diagnosis of pigmented villonodular synovitis (PVNS). An oncology consultation suggested and encouraged surgical excision in combination with radiation therapy to reduce the risk of recurrence of this destructive process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

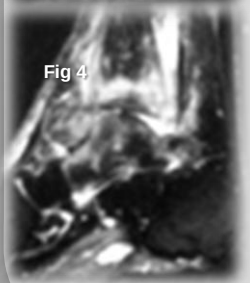
Surgical excision of the lesion and ankle arthroplasty was followed radiation therapy of 34Gy in 15 doses over a three week period. Radiation burn of the lateral ankle skin resolved with local care. At 7- years follow up and MRI failed to reveal evidence of recurrent disease.

Radiographic Evaluation

Plain radiographs reveal **joint space narrowing** in all orthogonal planes. **Fig 1,2**

Other significant findings are subtle including a **positive posterior hiatus sign** on the lateral ankle view as well as a **deep concavity** involving the **anterior aspect of the tibio talar joint and the dorsum of the talar neck** **Fig 1**





MRI Evaluation

T-1 image: Reveals the low signal intensity of a **large multi-nodular synovial-based mass** situated in the anterior aspect of the tibio-talar joint, filling the anterior recess of the joint. A **multi lobulated synovial proliferation** is seen within the posterior ankle capsule **Fig 3**

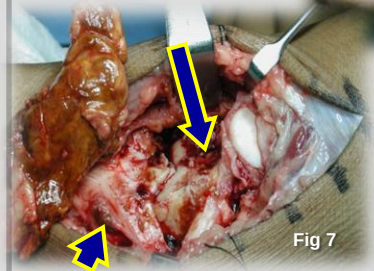
STIR image: Reveals **intramedullary edema** throughout the **talus and tibial plafond**. The **fluid within the posterior ankle joint** is commensurate with the increased recess noted on plain films.

Fig 4

Surgical Findings

The fascia overlying the mass was perforated by villous and nodular proliferations with hemosiderin deposits characteristic of the disease **(Fig 5)**

Nodular proliferation is composed of mono & multinuclear cells, Hemosiderin & lipid deposits **(Fig 6)**



This infiltrative inflammatory process has eroded the antero aspect of the tibio talar joint consistent w/ radiographic Findings **(Fig 7)** Arrow head at the tibial plafond and arrow at the talar neck

This infiltrative inflammatory process has been stalled at the antero lateral aspect of the tibio talar joint consistent with radiographic findings. There is no recurrence of nodular proliferation and residual structural deformity appears stable despite non compliance with the use of AFO devices (**Fig 8) When comparing this image with Fig 3 it is apparent that there is no evidence of recurrence of the space occupying and that the inflammatory process has been Stalled if not arrested by the combination of surgical and radiation therapy.**

Discussion

Pigmented villonodular synovitis has been described as a progressive and destructive infiltrative, inflammatory condition affecting both periarticular and intra articular structures. Some believe the condition to be rheumatic in origin while others believe this is simply a florid inflammatory response incited 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 years of age. The authors were able to cite 47 articles where the condition affected infants, children and adolescents which suggests that this condition affects a much wider patient population. Based upon a meta analysis of those articles in the English language 11-mos to 50 years 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 inflammation associated with trauma or repetitive injury.

Although the etiology remains uncertain there is a consensus that radiation therapy is a beneficial adjunct to surgical excision and debridement. We present a case using this technique in an attempt to forestall if not halt 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 debridement for PVNS. Although large group studies would be more valuable we feel that this case affecting an otherwise healthy and active female may provide insight for others faced with this challenging condition.

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Pigmented villonodular synovitis (PVNS) was first termed by Jaffe et al. in 1941 is an uncommon proliferative disease usually affecting the

Figure 1