CHAPTER **52**

PIGMENTED VILLONODULAR SYNOVITIS: Review of the Literature And Four Cases

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Introduction

Pigmented villonodular synovitis (PVS), first described by Jaffe et al[1] in 1941, is a benign proliferation of synovium of uncertain etiology. Speculations have been made that suggest PVS may be due to neoplastic process, inflammatory process, repeated trauma, or metabolic disorder. Three forms of PVS have been described depending on synovial involvement. These include nodular or localized PVS, diffuse PVS, and Giant Cell Tumor of the Tendon Sheath (GCTTS). Although different macroscopically these lesions appear to have the same histogenesis. PVS usually presents in the third to fifth decade of life and shows no predilection towards either sex. Patients generally present with monoarticular swelling out of proportion to discomfort and limitation of motion with no history of trauma to the area. Imaging studies including Roentgenography, CT and particularly MRI may play an important role in the diagnosis and treatment of PVS. Recognition of the clinical and radiographic pathology of PVS can lead to earlier biopsy and possibly less infiltration of the lesion.[2] Because pigmented villonodular synovitis can cause bone and joint destruction and has a tendency to recur, treatment consists of complete excision of the lesion and possible adjunctive radiation therapy.

Terminology

Chassaignac was the first to report a case of fibrous xanthoma of synovium.[3] Prior to 1941, pigmented villonodular synovitis had been described in the literature by various other names. These included benign or malignant polymorphocellular tumor of the synovial membrane, benign fibrous histiocytoma, chronic hemorrhagic villous synovitis, fibro-hemangioma, fibro-hemosideric sarcoma, giant-cell fibro-hemangioma, giant-cell tumor of the tendon sheath, myeloplaxoma, sarcoma fusigigantiocellulare, synovial xanthoma, synovial fibroendothelioma, synovial endothelioma, and xanthomatous giant-cell tumor.[4,5] In their 1941 paper, Jaffe et al[1] proposed the terms pigmented villonodular synovitis, tenosynovitis and bursitis, depending on the manifestation of the lesion. Granowitz et al[6] further subclassified the lesions based on clinical presentation into the more common diffuse form and the nodular, or localized form.

Etiology

The true etiology of pigmented villonodular synovitis is unknown. Speculation has centered around a few theories, including neoplastic or inflammatory processes, metabolic disorder, and chronic trauma. Recurrence rates, high mitotic activity, and a distinct difference between lesional and healthy tissue suggest a neoplastic process.[7] Ray et al[8] have reported cytogenetic clonality indicating origination from a common stem cell in a case of PVS. This hypothesis has been given further substantiation from reports of metastatic disease originating from articular PVS.[9-12] PVS has been shown to have increased uptake of Technitium-99 without gallium-67 citrate uptake which also lends evidence to a hyperplastic or neoplastic origin.[13] However, long-term survival rates and clinical course of disease are not representative of a recurrent neoplasm. Inflammation, as discussed by Jaffe et al,[1] continues to be a prevailing theory. Histologic

evidence has been reported that supports this theory.[14] Galloway et al[15] believed the lesions to be a result of abnormal lipid metabolism. Hirohata et al[16] advanced this idea, implicating the foam cell as the primary pathologic cell responsible for an abnormal cholesterol and phospholipid synthesis. Several studies have been carried out to examine trauma as the etiologic agent.[4] Myers et al[17] uncovered a somewhat common history of repeated trauma in patients who have PVS. Although a history of trauma has been reported in a minority of cases, trauma remains a possible etiology for PVS.[18,19] Interestingly, Wendt et al[20] have suggested that there may be a genetic contribution to the development of this lesion in their report on a case of PVS in two siblings and in multiple generations.

Clinical presentation

PVS occurs most commonly in the knee, followed by the hip, ankle, carpal and tarsal bones, shoulder and the elbow. PVS has been reported in patients ranging from 11 to 68 years old. PVS occurs in the third to fifth decades of life, with approximately equal distribution among males and females. The mean age at presentation is in the fourth decade.[21] The majority of patients usually present with symptoms including marked localized monoarticular swelling out of proportion to a mild or dull pain. A palpable mass can sometimes be felt. Multiple joint involvement has been reported.[5,22-24] The time of symptom onset to correct diagnosis of PVS of the knee, wrist and ankle is approximately 10 months.[25]

Laboratory studies including hemoglobulins, CBCs, sedimentation rates, urinalysis, Gram's stain and serum cholesterol are almost always normal.[19,25]

Clinical features

On gross examination of localized PVS, the synovium may appear from orange-yellow to reddish-brown. The lesion is typically lobulated. The villi are often long and appear as tangled, matted masses, demonstrative of a ìstraggly-beardî appearance. Diffuse PVS may present individual or clustered nodules along the synovium with prominent pigmentation ranging from yellow to red-brown.[4] The pigmented tissue may extend into sub-synovial structures and the joint capsule.[18] The synovial membrane may be thickened and rubbery.[1] Joint aspiration may yield a translucent straw-yellow to bloody red or brown fluid. Bone involvement has been reported as a single or confluent cluster of cyst-like areas bordered by a thin margin of sclerosis, mimicking a primary bone neoplasm.[26,27]

Classic microscopic description of the lesion includes subsynovial proliferation of large round, polyhedral or spindle shaped cells.[4] This microscopic appearance along with immunophenotype studies present evidence that PVS is of synovial cell origin.[28] The spindle shaped cells tend to appear in areas of marked fibrogenesis.[29] Some combination of histiocytes, fibroblasts and hemosiderin-laden macrophages and foam cells also has commonly been described.[6] A distinct feature of PVS is the intracellular and extra-cellular accumulation of hemosiderin and consequent diffuse pigmentation of the lesion site.

Imaging studies

On plain film radiographs, two out of three cases show some sort of soft tissue density.[18] Approximately 50 percent show some osseous changes.[30,31] Excluding the knee greater than 80 percent of cases have demonstrated bony involvement[30] Classic findings of PVS include monoarticular involvement, preservation of cartilage, synovial swelling, absence of calcification within the swelling, and normal bony mineralization of the affected joint.[32] Joint space narrowing, just as polyarticular involvement, is an uncommon finding. If there is bone involvement one or more intraosseous cysts may be apparent, each delineated by a thin rim of sclerosis and surrounding a bone of normal density.[27] These defects may be lobulated. PVS may be difficult to distinguish from tuberculosis, rheumatoid or osteoarthritis.[5] Most authors conclude that radiographs alone should not be used in confirming the diagnosis of PVS.[4,33,34]

Rosenthal et al[34] have reported increased attenuation on computed tomography (CT) scan of a PVS lesion. The increased absorption is believed to be due to the high iron content of the growth. Arthrographic study of diffuse PVS may reveal multiple irregular filling defects that project into an enlarged joint and are fixed to the joint capsule.[4,32] Johansson et al[25] report that athrograms provide little if any diagnostic aid. Arthrograms are generally not indicated for this lesion.

Angiography, although nonspecific, may be utilized for defining the extent of the lesion, due to the increased vascularity, presence of numerous arteriovenous shunts and irregular vessels commonly seen with PVS.[30,36] The presentation of fine caliber smooth walled arteries, late phase contrast pooling in dilated vascular spaces and early visualization with this study suggests synovial hemangioma rather than PVS.[37] Rosenthal et al[34] have reported a case of PVS with little vascularity. Lesions of this type are believed to be more mature, and demonstrate fibrosis as a more important tissue component.[38]

Ultrasonograms can be utilized to evaluate the anatomic dimensions and structure, density, and vascularity of PVS lesions. The use of color doppler sonography has revealed hypoechoic synovial proliferation and hypervascularity of a lesion involving the ankle joint.[39] Ultrasound may also be useful for tissue sample enhancement during joint aspiration or biopsy.[40]

The magnetic resonance (MR) appearance of PVS lesions essentially depends on the intrinsic amount of hemosiderin and lipid deposited within the growth,[41,42] and often appears as a heterogeneous mass. PVS appears dark due to low signal intensity on both T1- and T2- weighted MR images caused by the ferromagnetic property of the deposited hemosiderin.[43] Nodules with sufficient hemosiderin deposition have low signal intensity on T1- and even more signal void on T2-weighted images.[44] Although literature has claimed the signal intensity to be equivalent to or lower than muscle, this is true in only half the cases of PVS. Fat equivalent signal intensities can be seen in the mass due to lipid filled macrophages associated with PVS. There are likely to be small pockets of tissue with variable signal intensity that contributes to the heterogeneity of the mass. About half of the lesions will demonstrate a low signal capsule and septa.[45] At high field strength the random distribution of hemosiderin in the synovium alters the local magnetic sensitivity resulting in shortened T2 relaxation time and loss of signal on T2-weighted images.[12,35] The diseased synovium and inflammatory process may best show up on T2-weighted images. This signal void is seen with other synovial lesions (synovial osteochondromatosis, rheumatoid arthritis, hemangioma, chronic traumatic synovitis, and fatty synovial tumor); therefore, biopsy is necessary for proper diagnosis.[32] When associated with hemorrhagic or xanthochromatic joint effusions these hypointense lesions strongly suggest PVS.[46]

Treatment

Treatment generally depends on the type of lesions present. The localized form of PVS is sufficiently treated by local excision of the tumor. Curettage of bone is performed if there is bony involvement. Recurrence rates are low, and these patients generally have a good prognosis. Borton et al[47] have successfully utilized arthroscopic synovectomy for a lesion localized in the first metatarsophalangeal joint. For arthroscopic approach to treating PVS complete arthroscopic synovectomy has resulted in substantially lower recurrence rates than partial synovectomy.[48] Diffuse and recurrent PVS require a different approach. Wide synovectomy is recommended and has had satisfactory results.[25,49] Radiation therapy and combination surgery plus radiation therapy have both been utilized. Injections of radioactive gold colloids such as Au-198 postoperatively have been employed with some success.[50] Currently intraarticular Yttrium-90 injection is an alternative to Au-198.[14,51] This therapeutic modality has been shown to be effective in cases without extra-synovial spread.[51] Franssen et al[52] demonstrated areas of persistent synovitis after repeated injections of this colloid, although their studies did show decreased numbers and prominence of villi on the diseased synovium. Moderate doses of radiation (35Gy in 15 fractions) have been shown to be effective in diffuse PVS especially after total gross removal of the lesions, with satisfactory limb function maintained.[50] Additionally, arthrodesis, arthroplasty, and radiation synovectomy (with dysprosium-165-ferric hydroxide macro-aggregate) alone or in conjunction with surgical synovectomy have also been utilized. Radiation therapy can cause joint stiffness or radiation induced sarcomas[21] and thus radiation therapy should be reserved for patients in whom surgical intervention is contraindicated.[53]

Case Report 1

A twenty-six-year-old female presented with swelling on the outside of the right foot which had been present greater than four months and getting progressively worse. The patient denied any pain with the swelling. On physical examination, the neurovascular status was intact. There was a three by two centimeter nodular mass over the cuboid area of the right foot that was elevated approximately one centimeter (Figure 1). The mass was firm to palpation. The patient was negative for pain on palpation over the entire area of the mass. The mass was negative for discoloration, and otherwise the dermatologic examination was normal. There were no other relevant findings on the orthopedic examination.

Radiographic findings included a semi-circular erosion of the plantar aspect of the calcaneal cuboid joint. Mild sclerosis of the bone was noted along the erosion into the cuboid. MRI images including T1, T2, inversion recovery, and post gadolinium T1 weighted with fat saturation were taken in combinations involving all three planes (Figures 2 and 3). The images clearly demonstrated a soft tissue mass approximately 3 cm x 3.5 cm x 4cm in the frontal, sagittal and transverse planes respectively. The mass was located along the lateral border of the foot infiltrating the cuboid and to a lesser degree the calcaneus along the plantar aspect of the calcaneal cuboid joint. It completely surrounded the peroneal tendons. The signal intensity was slightly decreased on T1 weighted images and markedly decreased on T2 weighted sequences. Post gadolinium studies enhanced the images somewhat. The impression from the MRI studies was pigmented villonodular synovitis as the leading diagnosis followed by tophaceous gout and

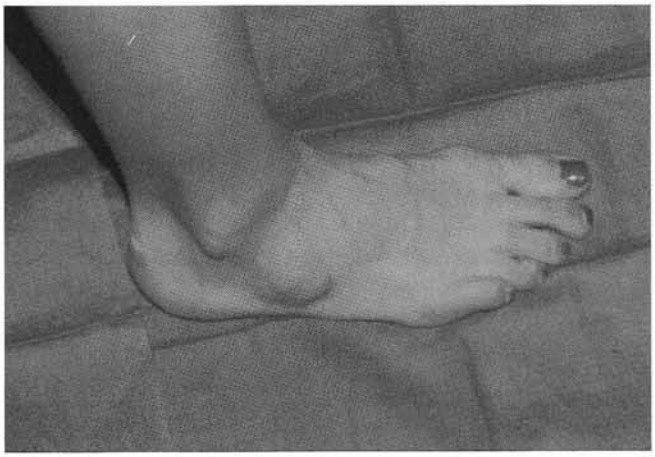
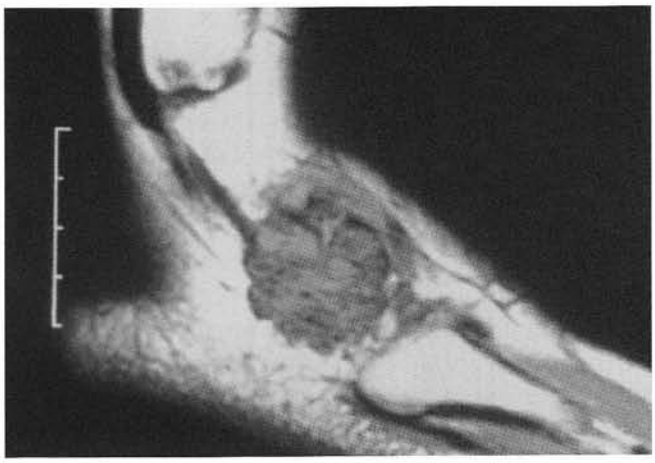


Figure 1. Case 1. Clinical photograph demonstrating a nodular mass over the lateral aspect of the foot.



Figures 2 . Case 1. T1 weighted MR images defining the anatomic boundaries of the mass.

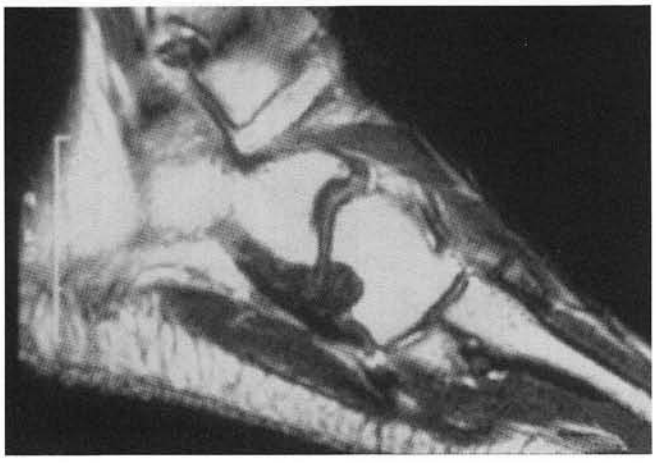


Figure 3. Case 1. The mass can be seen extending into the calcaneocuboid joint. The patient had no pain with this mass.

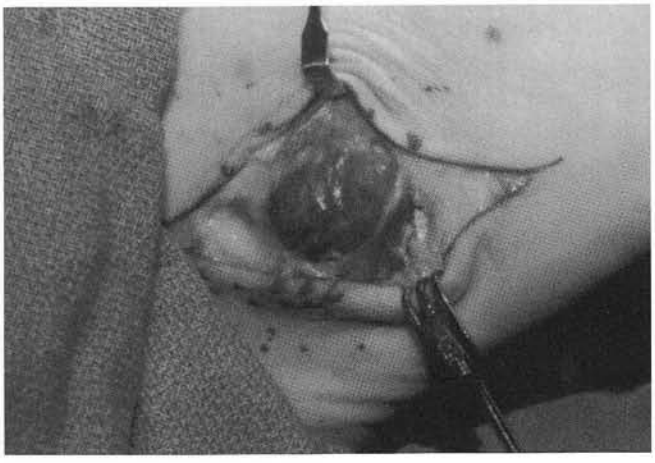


Figure 4. Case 1. Intraoperative view of the mass once the deep fascia was incised.

synovial cell sarcoma. The decreased signal intensity on the T2 images rendered sarcoma as unlikely.

The space occupying size of the lesion and the patient's concern of malignancy lead to surgical intervention. The lesion was sent for frozen section to verify the diagnosis of PVS at the time of surgery. The procedure was performed under an ankle tourniquet. The lesion was accessed through a straight-line incision over the mass from the tip of the fibular malleolus over the calcaneo-cuboid joint onto the base of the fifth metatarsal. After dissection through the subcutaneous tissues an incision was made through the deep fascia just plantar to the extensor digitorum muscle belly. A brownish red mass with hemosiderin deposits consistent with PVS was visualized at this level (Figure 4). The communicating branch from the lateral dorsal cutaneous to the intermediate dorsal cutaneous nerve was located over the mass and retracted throughout the procedure. The extensor muscle belly was dissected and mobilized dorsally. The mass was noted to completely circumscribe the peroneus brevis tendon from 1.5 cm distal to the fibular malleolus to 1 cm proximal to its insertion into the base of the fifth metatarsal as well as the peroneus longus proximally.

A major portion of the mass was excised and sent to pathology. Upon excision of the mass a brownish fluid exuded from the wound with the viscosity and color of blood tinged synovial fluid. A portion of the mass was sent for frozen section to pathology and the fluid was cultured and sent to microbiology for culture and sensitivity.

Attention was then directed to the calcaneo-cuboid joint. Dissection was carried out through the capsular and periosteal tissue at this level. Upon reflection of the periosteum two channels were noted with lobules of the mass infiltrating the calcaneus distally and the cuboid proximally at the calcaneo-cuboid joint. The entire plantar aspect of this joint had been eroded. Curettage was used to remove the lobules and send a portion of the calcaneus and cuboid to pathology for identification. What appeared to be the stalk of the mass was then followed plantar to the calcaneo-cuboid joint into the plantar aspect of the foot and removed. Synovectomy of the calcaneo-cuboid joint was performed. The lesion surrounding the peroneus brevis and a portion of the longus was excised with the tendon sheath. Prior to completion of the procedure the frozen section from pathology was evaluated as chronic synovitis consistent with PVS. The wide excision and vascularity of the tissues resulted in a moderate amount of bleeding. The tourniquet was deflated and all bleeders were tied or coagulated. Absorbable suture was then used to close the wound in layers over a penrose drain. Owens silk and a dry sterile dressing were then applied.

The largest mass that was sent for pathology was 28mm x 20mm x 12mm in size with papillary and shaggy fragments weighing a total of 9 grams. The surface was brown in color. Upon sectioning the mass was solid with color variation from brown through tan with areas from yellow to grayish white. The microscopic description of the tissue was that of a nodular mass of synovial tissue that was intensely inflamed. The predominant cells were macrophages with some multinucleated giant cells. The macrophages contained an abundant brown granular pigment of hemosiderin (Figure 5). There was abundant vascular tissue and moderate lymphoid infiltrate. There was no atypia or malignancy seen. The bone from the cuboid and calcaneus showed erosion with invasion of macrophages, multinucleated giant cells, fibroblasts and a few lymphocytes. There was a moderate degree of vascular proliferation. The appearance was similar to pannus. The bone from the cuboid was significantly more infiltrated than the calcaneus. All cultures were negative

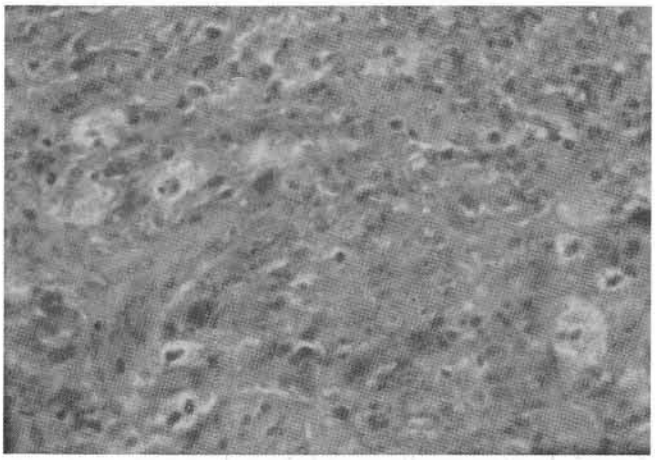


Figure 5. Case 1. A micrograph of the mass. The mass contained predominantly macrophages with brown pigment consistent with hemosiderin. Few multinucleated cells are also present.

Case Report 2

A twenty-eight-year-old female initially presented to the emergency room in March of 1999 with a chief concern of episodic ankle pain. She had been having these symptoms since October of 1998. She related periods of acute ankle swelling. Pertinent history included prior excision of lipoma along the posterior lateral of the same leg at the level of the ankle. On presentation she had fullness along the posterior lateral aspect of the leg anterior to the Achilles tendon just above the superior surface of the calcaneus. There was no significant edema or joint effusion appreciated on examination. Palpation revealed mild to moderate tenderness over the area of a suspected mass. Range of motion of both the ankle and subtalar joints was within normal limits, without crepitation and pain free at the time of the exam. X-rays taken during her emergency room visit did not reveal osseous or joint pathology. An MRI was taken to verify the presence and determine the nature and size of the soft tissue mass. Medical history was unremarkable with the exception of the prior excision of the lipoma. The patient's only medication was birth control and occasional ibuprofen for the ankle pain and swelling.

The MRI was completed in May of 1999, but the patient was not able to follow up until July of the same year. During that time the pain had subsided. She denied any swelling of the area during this period. The MRI revealed a soft tissue mass 1.5 x 1.2 x 2.5 cm mass along the superior margin of calcaneus abutting the subtalar joint (Figures 6 and 7). The mass exhibited a fairly heterogeneous appearance with an intermediate intensity on the protein density images and low signal intensity on the T2 weighted images. The primary diagnosis based on the MRI and clinical presentation was pigmented villonodular synovitis. The patient was advised regarding the nature of the presumptive diagnosis and the patient elected to have excisional biopsy of the mass.

The following month the patient underwent excision of the mass. Hemostasis was achieved with epinephrine 1;200,000 with the local infiltrate. A straight to mildly curved incision was used just anterior to the Achilles ending just above the calcaneus. The incision was carried down through the dermis down to the level of the subcutaneous tissue. Blunt and sharp dissection was carried out down through the subcutaneous tissue and deep fascia. Once the deep fascia was incised a brown to yellow mass protruded that was well encapsulated (Figure 8). Blunt dissection was carried out to isolate the mass. The mass had a stalk that originated from the subtalar joint. A hemostat was applied to the stalk prior to excision of the mass. A stick tie was performed with non-absorbable suture to tie off the stalk. The mass was sent to pathology for identification (Figure 9). The incision was closed in layers in a running fashion.

The mass was identified as PVS. The patient healed the incision uneventfully. The patient has no residual pain or swelling in the area and has returned to full activity. At 18 months follow-up the patient has no signs of recurrence.

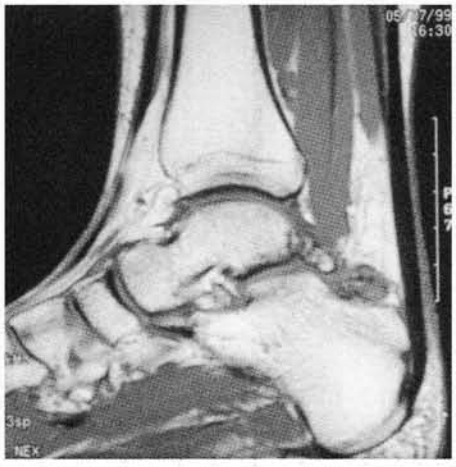


Figure 6. Case 2. MRI of the mass extending from the subtalar joint out over the superior aspect of the calcaneus on a TI- weighted image. The mass did cause significant pain and symptoms for the patient.

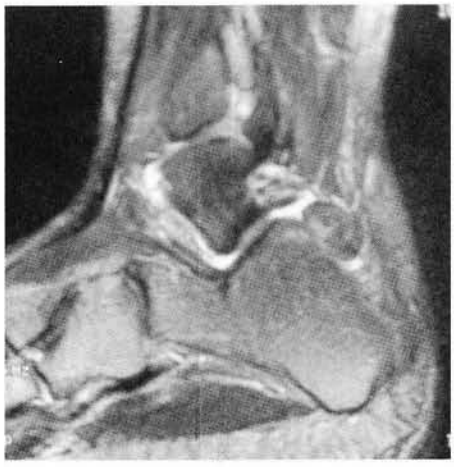


Figure 7. Case 2. A T2- weighted image of the mass illustrating a well emarginated mass with heterogenous appearance. The mass is isointense with muscle.

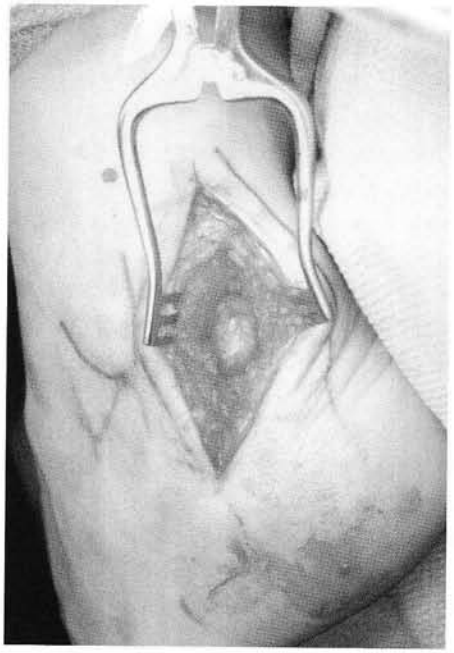


Figure 8. Case 2. An intraoperative photograph of the well encapsulated mass once the incision was carried out through the deep fascia

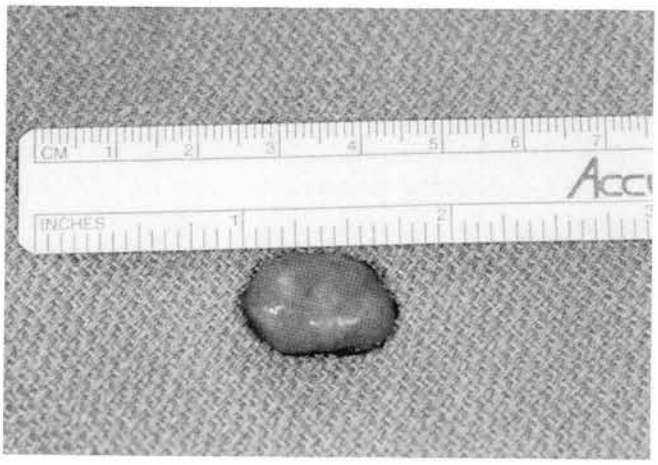


Figure 9. Case 2. The mass was removed in its entirety and measured approximately 2cm in length.

Case Report 3

An 81-year-old female originally presented in May of 1996 with a mass over the dorsum of the left foot (Figure 10). At that time she related swelling present for over one year. Pain was present while walking secondary to shoe gear pressure. Another physician had previously seen her and ordered x-rays and an MRI series. On physical exam she had a cystic mass over the dorsum of the left foot. X-rays revealed a calcified mass 1.5 cm. in diameter adjacent to the second metatarsal (Figure 11). MR imaging demonstrated a heterogeneous soft tissue mass involving the extensor hallucis longus tendon and the presence of fluid in the intermetatarsal space (Figure 12). The cystic mass was aspirated. Culture of the fluid was negative for organisms. There were PMN's, and mononuclear cells. Synovial fluid analysis revealed a bloody fluid with WBC's of 1,413, decreased viscosity, 43% neutrophils and an absence of fibrin. Crystal fluid analysis revealed cholesterol like crystals in the fluid. At that time the patient was offered an oncology consult or excisional biopsy of the mass. The patient elected to defer treatment and did not follow-up again for a year and a half.

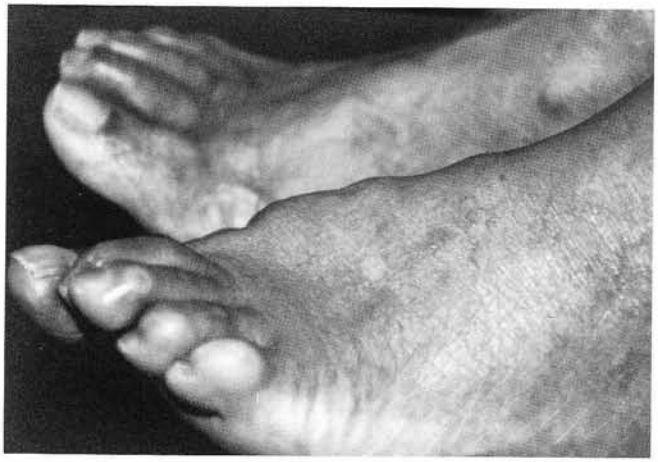


Figure 10. Case 3. Clinical photograph of the dorsal foot showing the mass along and medial to the externsor hallucis longus tendon.

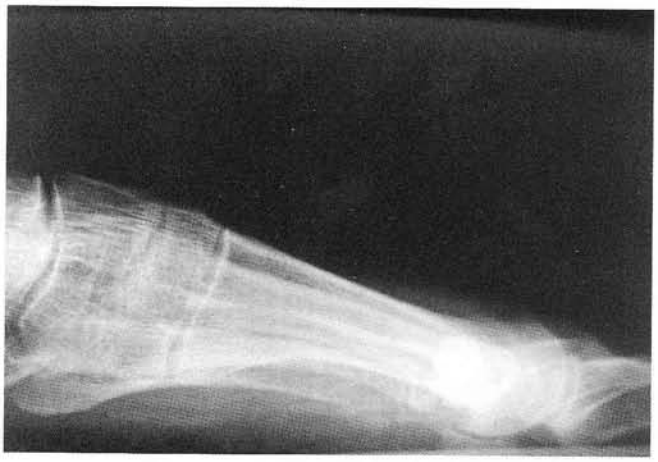


Figure 11. Case 3. Sclerotic mass is seen medial to the neck of the 2nd metatarsal.

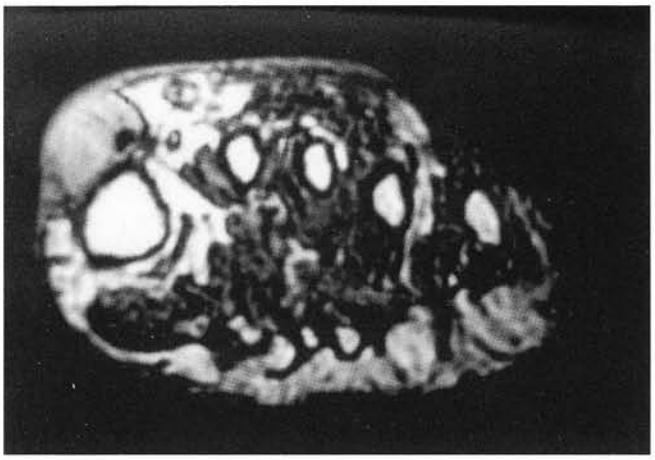


Figure 12. Case 3. MR demonstrated heterogenous appearance of the mass with involvement of the extensor hallucis longus tendon. There is also fluid in the intermetatarsal space.

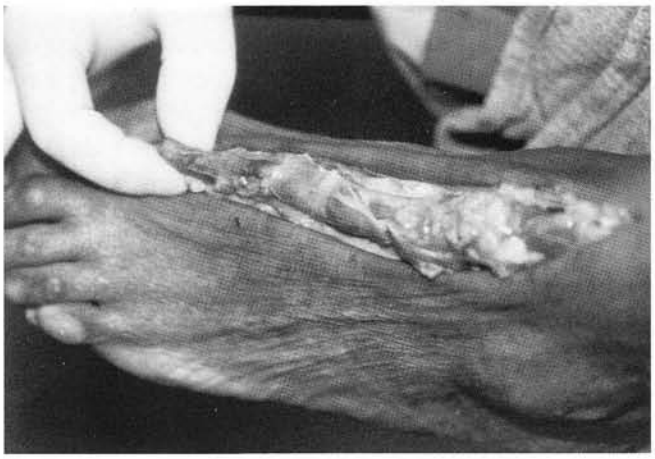


Figure 13. Case 3. Intra-operative view illustrating the mass encompassing the externsor hallucis longus proximal to the ankle.

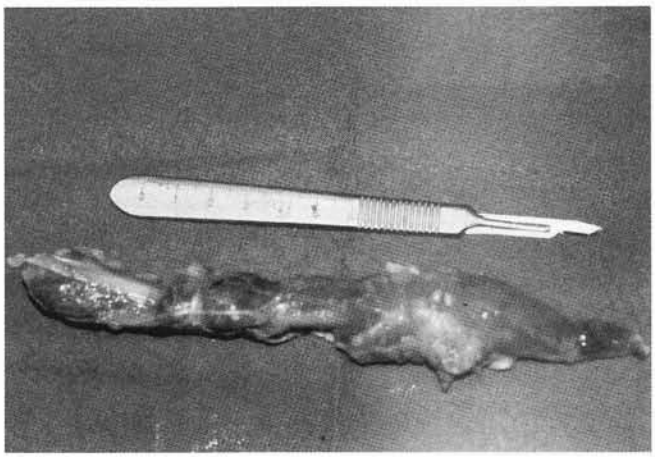


Figure 14. Case 3. The mass along with the extensor hallucis longus tendon excised with a length of about 15 centimeters.

When she presented over a year later she related increase in pain and size of the mass now extending onto the ankle. Her symptoms were similar to initial presentation with pain from shoe pressure. Examination at that time revealed a fluctuant mass extending from the first MTPJ to the anterior ankle. There were multiple lobulated regions that were fluctuant on palpation. She had point tenderness over the distal second metatarsal corresponding to the location of the calcified mass. There was no change in size or appearance of the calcified mass. There was no trabeculation within the mass, however there was erosion into the second metatarsal.

Although cytology did not reveal any neoplastic cells, it was explained to the patient that this could not be completely ruled out until biopsy was performed. The patient elected to undergo excisional biopsy of the mass. In November of 1997 the patient underwent excision of both masses. Hemostasis was achieved with a thigh tourniquet at 250 mm Hg. A 5 cm incision was carried out over the calcified mass extending from the first interspace proximally to the base of the first metatarsal. Sharp and blunt dissection was performed in layers providing surgical hemostasis throughout. A calcified mass was located adjacent to the second metatarsal with erosions noted into the second metatarsal. The lesion was dissected down between the interossi muscles into the plantar vault. There was no proximal or plantar extension of the mass. The mass measured 2 x 4 cm. in dimension. The mass was well encapsulated, appeared yellowish and had a rubbery consistency.

Attention was then directed to the level of the first metatarsophalangeal joint. A 15 cm incision was carried out from this level over the extensor hallucis longus tendon proximally over the anterior ankle and leg. Dissection was carried out to expose the mass located within the subcutaneous tissues completely encircling the extensor hallucis longus tendon (Figure 13). The tendon was transected distal to the extent of the tumor. Care was taken to completely dissect out the mass (Figure 14). The color of the mass was variegated with spotty areas of yellowish color and a preponderance of chocolate brown color. The mass passed over the deep peroneal nerve and anterior tibial vessels and ended at the myotendinous junction of the extensor hallucis longus. The muscle tendon complex

was distracted distally and transected 2 cm proximal to the most distal aspect of the mytotendinous junction. There was no visible tumor after excision. The wound was closed in layers with absorbable and non-absorbable sutures. A moist dressing was applied and a posterior splint was applied. The specimen was incised on the back table were several milliliters s of a low viscosity brownish-reddish colored fluid. Cultures were sent for aerobic and anaerobic culture with a gram stain. Specimens were sent for pathology with acid fast and fungal cultures prior to fixating in formalin.

The mass was negative for organisms. The patient did experience pain, swelling and decreased range of motion. The patient was treated with physical therapy, compressive dressings and ultimately compressive stockings. The pain, edema and decreased ankle range of motion resolved on a gradual basis. There were no signs of recurrence during her follow-up care and the patient was pain free and ambulating well.

Case Report 4

A 35-year-old white female presented initially with a painful mass at the medial aspect of the left ankle which had been increasing in size and discomfort over a 6 month period. She denied any history of trauma to the region. She could not walk with a normal gait due to the pain in the rearfoot. On physical exam, there was a palpable mass along the medial ankle which was extremely painful to palpation (Figure 15). There was a neuritic component to the mass with shooting pain radiating proximally into the leg upon percussion. A MRI was obtained which revealed a 1.5 x 3cm well circumscribed mass intertwined in the long flexor and posterior tibial tendons. The signal was isointense with normal muscle. The differential diagnoses considered at this point was leiomyoma, fibroma, leiomyosarcoma, and giant cell tumor of the tendon sheath (PVS).

The patient underwent excisional biopsy 3 weeks later. Under adequate intravenous sedation with local anesthesia and with a mid-calf tourniquet inflated a 7.0 centimeter incision was made directly over the lesion. The incision was carried down through the laciniate ligament, with care to identify both the posterior tibial artery and nerve to preserve and retract these vital structures. Just deep to the laciniate ligament a multinodular, loculated, encapsulated soft tissue mass was identified measuring 5 cm x 3 cm x 1.5 cm (Figure 16). The mass was yellow with admixed spots of brown. The flexor digitorum longus and brevis tendons and the posterior tibial tendon were identified. The mass was dissected free from the surrounding tissues in entirety (Figure 17). The neurovascular structures were again assessed and determined to be intact. The wound was closed in layers with absorbable suture and a dry sterile dressing was applied over a wet saline gauze.

The specimen was sent for pathologic and histologic evaluation. Microscopic analysis revealed numerous giant cells with admixed intervening fibrohistiocytic cells, some of which contained hemosiderin pigment. Foamy macrophages were also prominent, as well as some fibroconnective tissue. This analysis was consistent with and confirmed the diagnosis of pigmented villonodular synovitis.

After five months, the patient has residual neuritic-type pain over the operative site. There is no evidence of recurrence present on follow up, and the patient is propulsive with the ability to bear full weight on the heel.

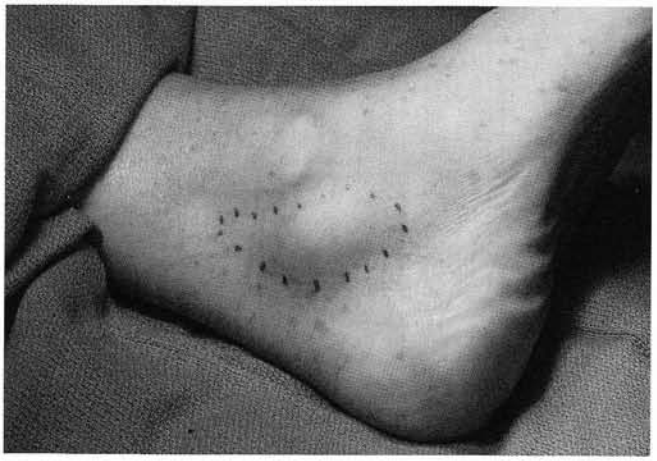


Figure 15. Case 4. Clinical photograph demonstrating a soft tissue mass at the medial aspect of the ankle.

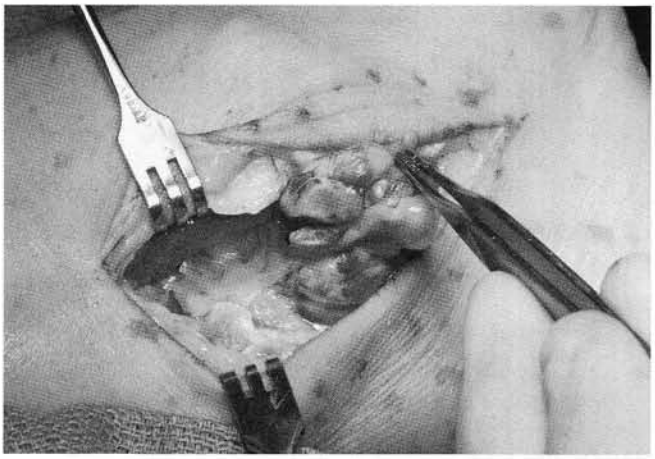


Figure 16. Case 4. The mass was noted to envelope the flexor tendons and neurovascular structures.

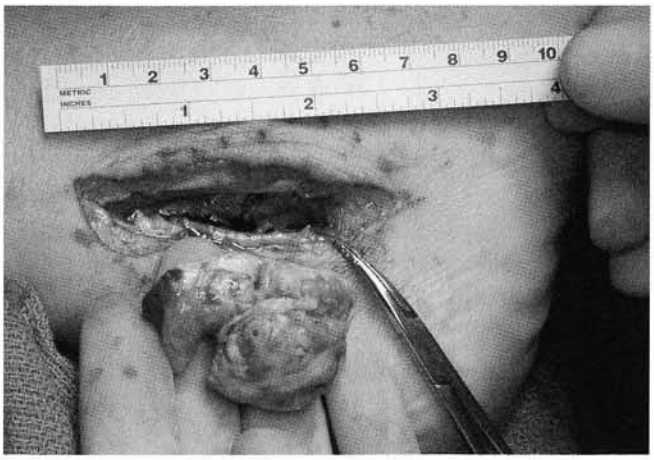


Figure 17. Case 4. The mass was excised in toto and sent for histopathologic diagnosis.

Discussion

PVS is rarely reported in the foot. It is unclear whether PVS does not occur in the foot or it goes undiagnosed. Three of the patients in this study were in the fairly typical age range associated with this condition (the third decade) and the other was much older. The presentations of PVS were varied. The first case presented with monoarticular swelling without pain. Pain played a small role in cases 2 and 3, although was the dominant symptom in the fourth case. The masses in all four had nodular components that could have been confused with a mature ganglion. The erosion of the calcaneus and cuboid on radiographs placed synovial sarcoma, and although less likely, gout further up on our differential in the first case. The heterogeneity of the signal with the low intensity on T1 and T2 images lead to a presumptive diagnosis of PVS with a possibility of synovial sarcoma in the first two cases. The size of the mass and the growth over a year in the third case was suspicious for malignancy, although cytology did not reveal any neoplastic cells. When the patients were informed of a differential diagnosis that included malignancy they all ultimately chose to undergo excisional biopsy. All four cases had either a yellow, dark chocolate or reddish brown appearance within the mass. This was consistent with PVS and we became more comfortable with this presumptive diagnosis once the mass was visualized. The frozen section was performed in the first case to confirm our diagnosis prior to discussion with the patient and her family. These four cases all had similar gross appearance with significant differences such as size, location, erosion of bone and encapsulation or diffuse nature. The first case involved the peroneal tendons, the calcaneo-cuboid joint with infiltration of both bones and plantar extension of the mass. The second case was nodular and well encapsulated and originated from the posterior subtalar joint. The third case primarily involved the extensor hallucis tendon with a secondary mass in the first interspace. The fourth case was intertwined between the flexor hallucis and digitorum longus tendons although no clear origin from these tendons could be established.

Although nodular in appearance, characterization of PVS as nodular or diffuse was difficult in the first and third cases. The second and fourth cases were clearly encapsulated and thus categorized as nodular. The first involved the peroneal tendons and the entire calcaneo-cuboid joint. The third case extended along the entire extensor hallucis longus tendon with a possible second mass ultimately diagnosed as fibrocollagenous tissue with amorphous calcification. The diffuse variety has the propensity of recurrence whereas the nodular form does not. The surgical excision of these two masses was aggressive due to the recurrence rate noted in the diffuse type of lesion. The patients had no significant complications secondary to the procedure and healed uneventfully. At follow-up the patients had not had recurrence of the lesion. The second case involved a mass that was well

encapsulated with an origin from the subtalar joint. The mass was categorized as nodular and is unlikely to involve recurrence. However, it is interesting that the patient had a history of a lesion removed in this area that she related as a lipoma in the same area years earlier. The fourth case was hallmarked by intense neuritic pain, however there was no evidence of nerve involvement intraoperatively. It was evident that the mass was compressing the nerve, however removal of the mass failed to relieve the neuritic pain.

Conclusion

This report reviews the literature on pigmented villonodular synovitis and presents four different cases. Few cases of PVS have been reported in the foot. The four cases presented varied widely in their clinical presentation. MRI proved quite helpful in these cases for diagnosis and surgical planning. Our treatment protocol was wide excision to prevent recurrence in the more diffuse cases, since they are more likely to recur. At follow-up there has not been recurrence in the cases reported. Further reports of PVS in the foot are needed to determine the likelihood of recurrence in this area.

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