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PIGMENTED VILLONODULAR SYNOVITIS OF THE MIDFOOT

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A 60-year-old man presented with a longstanding slowly growing swelling at the dorsal aspect of the left midfoot. The lesion was imaged with CT, ultrasound, and MR. On CT bony erosions were evident. On MR the lesions appeared bifocal and one component was hypointense on T2 weighted images suggesting hemosiderin deposits. The other component was hyperintense on T2 which is more unusual for PVNS. Imaging findings, however, suggested PVNS which was pathologically confirmed. A unique finding in this case is the late age of presentation of the disorder. Also the bifocal nature of the lesion is relatively uncommon.

Key-words: pigmented villonodular synovitis, cuneonavicular joint, ankle and foot.

PVNS of the joints is not an uncommon disorder. It typically occurs in younger patients. Imaging findings usually allow a correct diagnosis. Bony erosions may be present later on in the disease, and on MR low signal intensity on T2 weighted images may suggest the diagnosis. In this case report the disorder occurred in an older patient which is unusual. Also the lesion was bifocal in nature which is not common. In addition one component of the lesion showed high signal on T2 which could be a misleading feature.

Case report

A 60-year-old male presents with a large painless swelling at the level of the dorsal aspect of the left midfoot which gradually increased in volume over the last few years. Recent complaints of increasing pain, localized tenderness and limitation of motion are evident. Further medical history was unremarkable.

CT clearly shows sharply delineated osseous destructions at the level of navicular, cuboid and cuneiform bones (Fig. 1). Ultrasound examination demonstrates prominent hypervascularization of the lesions. MR examination is performed and demonstrates the presence of a superficial mass at the dorsal aspect of the articulation, between the navicular and the first and second cuneiform bones and a second component located at the plantar aspect of the cuneonavicular joints. Superficial and deep components of the mass both demonstrate prominent enhancement of the entire lesion after contrast administration on T1 (Fig. 2). Furthermore the lesion appears isointense to muscle on proton density (PD) and hypointense on T2-weighted spin echo (SE) and gradient echo (GE) images indicating the presence of hemosiderin deposits. The deep component is rather hyperintense on T2 GE however, most probably due to inflammatory reaction (Fig. 3). There are no signs of involvement of the extensor tendons and tendon sheaths. Overall, the bony destruction and signal patterns orient the diagnosis to PVNS rather than giant cell tumor of the tendon sheath.

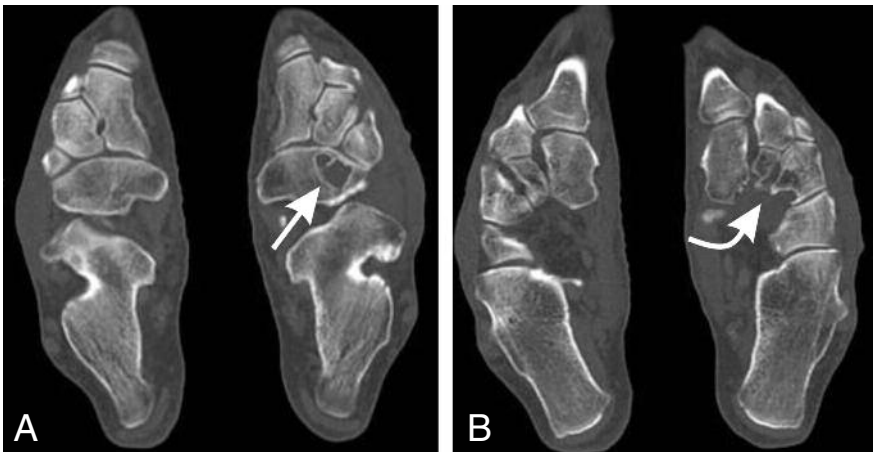

Fig. 1. — Axial plain CT images of the left foot show extensive bony erosions at the level of the navicular (A, straight arrow), cuboid, and cuneiform bones (B, curved arrow), but no lesions of the cuneonavicular articulation.

Surgery is performed and macroscopic observations confirms the characteristics of the lesion as being compatible with PVNS. The surgeon manages to resect the dorsal and plantar part of the lesion.

Microscopically the specimen is made up of a collagen tissue matrix with the presence of abundant cuboid and fusiform cells of fibrohistiocytic nature, leucocytes and macrophages loaded with hemosiderin pigment, histiocytes charged with fat, multinucleate giant cells, as well as synovial lining cells (Fig. 4).

Discussion

PVNS clinically manifests itself as pain and joint swelling in young adults. Joint effusion is observed in the knee in some cases but is uncommon in other joints. Joint aspiration typically demonstrates a xanthochromic joint effusion (1).

Imaging plays an important role in the diagnosis, treatment, and

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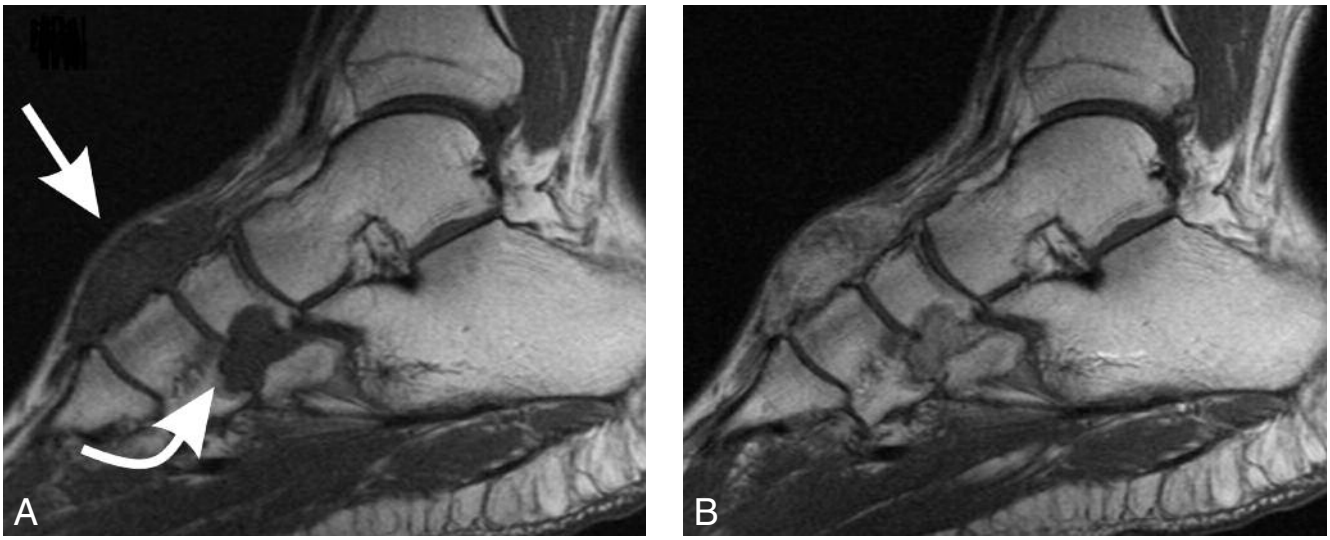

Fig. 2. — A. Sagittal T1-weighted images show a soft tissue mass at the superficial aspect of the navicular and cuneiform bones (straight arrow) with cuboidonavicular transarticular extension with bone erosions (curved arrow) and prominent heterogeneous enhancement after contrast administration (B).

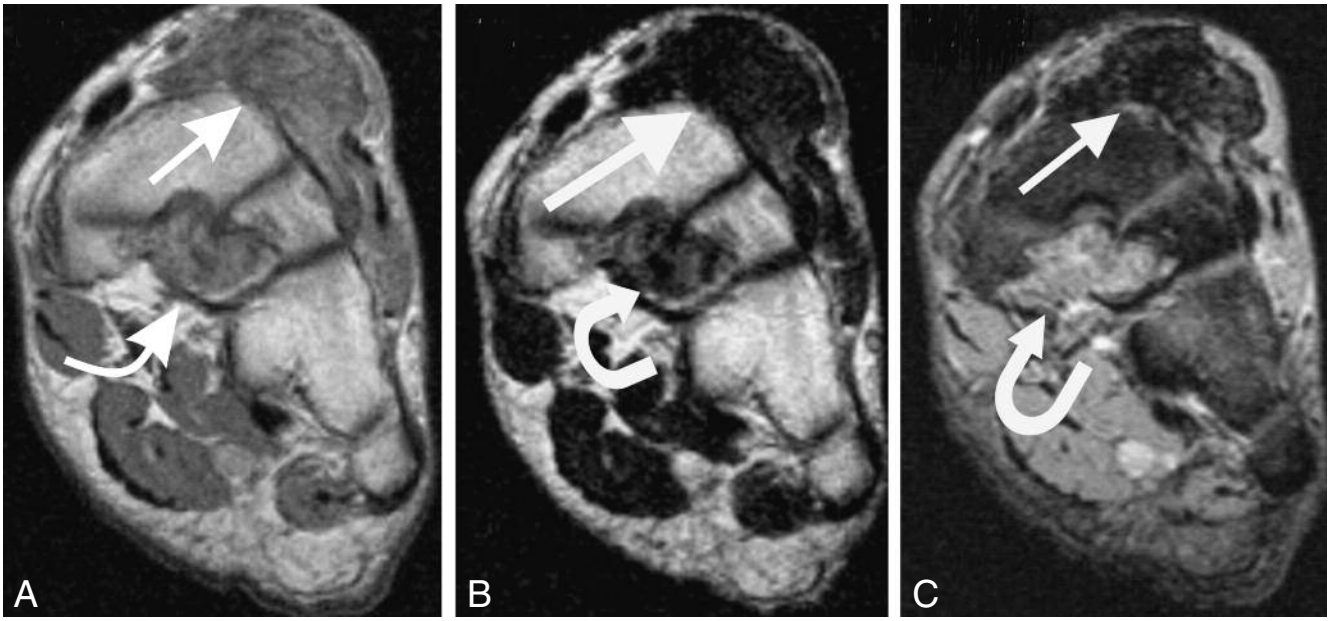

Fig. 3. — A. PD and (B) T2 SE as well as (C) T2 GE coronal images demonstrate intermediate and hypointense signal intensity respectively for proton density and T2-weighted images, whereas T2 GE sequence shows hypointense signal intensity for the superficial aspect of the lesion (straight arrow) with small areas of very low signal intensity indicating the presence of hemosiderin. A heterogeneously increased signal intensity at the deep aspect of the soft tissue mass could suggest the presence of synovitis (curved arrow).

follow-up of the disorder. Conventional radiographs of joints affected by PVNS may appear normal or may demonstrate periarticular soft-tissue swelling. Joint spaces and bone mineralization are characteristically preserved until late in the disease. Bony erosions are common in joints with a tight capsule, such as the hip and ankle. On MR images, the mass-like proliferative synovium has a lobulated margin, and it may be extensive in diffuse PVNS or limited to a single nodule in the focal form. The lesions tend to bleed, causing hemosiderin deposition and a characteristic low signal intensity with all pulse sequences. Areas of high signal intensity on T2- weighted images may be present and are likely caused by inflamed synovium or joint effusions (2-6). This is a somewhat misleading finding that was present in our case, because more typically the signal intensity on T2 is low. PVNS should be considered in ankle masses with bony erosions and low signal intensity masses on T1 and T2 MR imaging.

PVNS is a benign proliferative disorder of the synovium that may affect the joints, bursae, or tendon sheaths. PVNS most often occurs in young to middle aged adults. A unique finding in this report was that the disorder occurred in an elderly patient. Two primary forms are described, including a diffuse form that affects the entire synovial lining

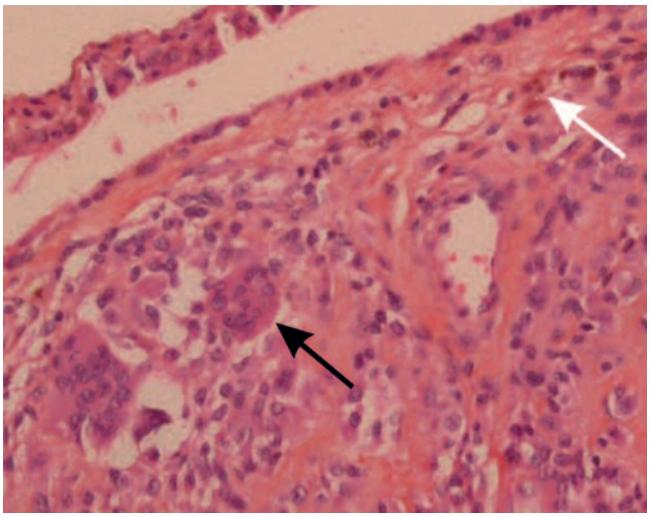

Fig. 4. — Hematoxylin-eosin staining (10 x) shows the hyperplastic synovium consisting of fibrous stroma containing foamy macrophages, lymphocytes, and plasma cells. On the left, two multinucleate giant cells are seen of which one is indicated (black arrow) and deposits of golden brown hemosiderin lie within the stroma itself as well as within the cytoplasm of synovial cell lining and macrophages (white arrow).

of a joint, bursa, or tendon sheath, and a focal or localized form also called localized nodular synovitis (7). The knee is the most frequently involved joint, followed by the hip, ankle, and shoulder. Local recurrence following surgical or arthroscopic synovectomy occurs in almost 50% of cases. Polyarticular involvement is extremely rare. Another unique finding in this case was the bifocal presentation of the lesion.

PVNS is currently classified in the group of fibrohistiocytic tumors according to the WHO classification.The differential diagnosis for intraarticular masses is limited. Anatomic knowledge helps the radiologist in localizing masses to the joint space. Familiarity with the typical imaging characteristics allows one to make confident diagnoses of many of the diseases causing intraarticular masses, the most important ones being noninfectious synovial proliferative processes such as lipoma arborescens and synovial osteochondromatosis, infectious and inflammatory diseases such as tuberculous arthritis and rheumatoid arthritis, deposition diseases such as gout and amyloid arthropathy, vascular malformations such as synovial hemangioma and arteriovenous malformations, malignancies with synovial osteochondromatosis and metastasis (8-11).

At pathologic analysis, PVNS is characterized by synovial inflammation with multinucleate giant cell proliferation, collagen, and lipid-laden macrophages. Treatment of PVNS often consists of resection of the lesion. However, a recurrence rate of 10%-20% is reported in the focal forms and of up to 50% in the diffuse forms (3, 12).

In conclusion we report a case of PVNS of the midfoot. Unique findings in our case were the older age of the patient and the bifocal presentation of the lesion. PVNS should be suspected with any periarticular or intraarticular mass with low signal intensity on all MR imaging sequences.

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