PVNS talus in a patient treated for chondral lesion in ipsilateral calcaneum: A case report and review of literature

Apurv Gabrani¹, Hitesh Dawar¹, Deepak Raina¹, Surbhit Rastogi¹

Abstract

Introduction: PVNS is a locally aggressive synovial proliferative disorder of unknown etiology and has been described in the foot and ankle in previous literature. A case of PVNS in the talus has been described in a patient treated for ipsilateral calcaneal chondral lesion. Case Report: A 56 year old male presented with pain in his left ankle of 4 months duration. On investigation, he was found to have a well defined lytic lesion in the left calcaneum on x-ray. MRI showed a hyper intense lesion on T2WI. A needle biopsy revealed chondrogenic tumor which was managed by extended curettage. At 12 months follow up, patient presented with recent onset pain over the anterior aspect of left ankle which showed hypo density over the supero-anterior aspect of the talus and MRI showed ill defined hypo intense lesion on T2WI and hyper intense lesion on T1WI. The lesion increased in size on repeat MRI 6 weeks later. He was managed with synovectomy and debridement with core needle biopsy of talus. Histopathological examination revealed features consistent with PVNS. Patient remains asymptomatic at 1 year follow up after surgery. Conclusion: A double primary lesion although rare, does exist and any recurrence should be viewed at with equal degree of suspicion as the primary lesion. Keywords: Pigmented villonodular synovitis (PVNS), Talus, Calcaneum, Double Primary lesion, Chondral lesion.

Introduction

Pigmented villonodular synovitis (PVNS) is a locally aggressive synovial proliferative disorder of unknown etiology affecting the lining of joints, tendon sheaths and bursae [1]. The common sites affected are the knee joint, flexor tendon sheaths of hand and hip joints, followed by ankle and shoulder joints [2]. PVNS has been described in the mid-foot and hind-foot in recent literature as a primary lesion [3]. PVNS of the talus in a middle aged man post treatment of a chondral lesion of calcaneum is an unheard of scenario. The case report of such a double primary lesion in the ipsilateral foot is a rarest of rare cases and the same is presented which to our knowledge is the first of its kind.

Case report

A 56-year-old male patient initially presented 2 years back with pain in his left ankle for 4 months which he described as a deep seated pain that was insidious in onset, progressive, aggravated by walking and partly relieved at rest. On examination he had tenderness over the medial aspect of the heel. X-rays revealed a well defined lytic lesion in the calcaneum with presence of calcifications within. MRI revealed a lesion hyper dense on T2 with surrounding edema. A needle biopsy was done which revealed chondrogenic tumor. He was definitively managed by extended curettage through medial approach (Figure 1). After an initial period of protected weight bearing, he resumed all activities at 3 months with no symptoms. Patient presented about 12 months after the surgery with recent onset pain localized to anterior aspect of the ankle joint. On examination, the ankle dorsiflexion was limited terminally with pain. X-ray revealed hypo intensity over the supero-anterior aspect of talus (Figure 2), MRI showed ill defined hyper intense lesion in talus on T1 weighted images and a hypo intense lesion T2 weighted images, with residual deficit of surgical curettage in the calcaneum (Figure 3). A needle biopsy was done but revealed non-specific inflammatory tissue. Patient was started on anti-inflammatory treatment and was followed up after 6 weeks. MRI now showed the talar lesion increasing in size and involving the soft tissues on the dorsal aspect (Figure 4). Patient was counseled and an open biopsy was scheduled, during which synovectomy with debridement with core biopsy of talus was done. Histopathological examination of the soft tissue sample revealed non-specific inflammatory tissue, but the bone core histopathological report suggested features consistent with Pigmented Villonodular Synovitis (PVNS). The wound healing was uneventful. Patient was put...
on protected weight bearing and a prolonged course of anti-inflammatory (Indomethacin) for 6 weeks, following which he was allowed to resume his activity of daily living. The pain got relieved after the surgery. Patient reports no symptoms and continues doing all activities at 1 year follow up, with no radiological sign of any recurrence.

Discussion
PVNS or pigmented villonodular synovitis was first described in 1941 by Jaffe [4]. The disease spectrum starts from localized forms of giant cell tumor of tendon sheath, and includes up to the more diffuse form known as PVNS [5]. Typically the patient presents with pain and limitation of movement in a single joint [6], most commonly the knee [1]. In the Scottish bone tumor registry, cases of PVNS have been described in multiple locations in the foot and ankle i.e. 2 in foot phalanges, 3 in Tarsometatarsal region and 9 in the hind foot [7]. The referred case being a double primary lesion in the bones of ipsilateral foot is thus the rarest of rare presentations.
mineralization and a relatively normal joint. MRI in synovial sarcoma shows high-intensity signal [7]. On immunohistochemical analysis, synovial sarcoma shows CD34 positive but negative for S-100 whereas PVNS shows positive stain for CD3 and CD20 (localized) and CD57 (diffuse) [16]. However, calcifications within the soft tissue mass, extraarticular spread, and diffuse bone invasion may indicate malignancy [17].

Appropriate evaluation includes X-ray of the involved area which shows a well defined lesion with sclerotic margins or maybe even non specific. X-rays may also show areas of calcification which shifts the diagnosis more in favor of synovial osteochondromatosis. In early cases, there may be well defined erosions due to pressure phenomenon that are typically non-marginal [14, 15]. In late stages, severe and concentric joint space loss may be visible due to cartilage destruction [6]. CT scan is a good imaging modality for image guidance of diagnostic core needle biopsy [7]. Ultrasound is an easily available investigation to temporally compare the size of lesion on follow up visits [18]. MRI scan is the investigation of choice which shows the exact location and extent of the lesion that helps in planning the surgical approach.

The appearance on MRI depends upon the relative proportion of lipid, hemosiderin, fibrous stroma, pannus, fluid and cellular elements. PVNS lesions usually show contrast enhancement on use of gadolinium that is seen well in gradient echo sequences. Typically, low signal areas represent hemosiderin, lipids and inflammatory fibrosis and high signal areas signify the presence of either lipid laden macrophages or hemorrhage [19]. The definitive diagnosis is histological which typically shows fibrous stroma, hemosiderin deposits, histiocytic infiltrates, foam cells, multinucleated giant cells and yellow lipid within macrophages [20, 21].

Early diagnosis and aggressive surgical resection has been recommended as the treatment of choice to decrease the risk of recurrence, be it the diffuse or the localized form. The above is valid for both localized and diffuse lesions [22, 6]. In our case, aggressive synovectomy was carried out followed by use of anti-inflammatory medication (Indomethacin) for 6 weeks which yielded good results for the patient. The role of intralesional radiation therapy is controversial [23]. The use of pre-operative intralesional radiotherapy as a means to decrease the volume of disease has been described but resulted in post-operative joint stiffness and also has the risk of secondary cancer [20]. Case ending up in amputation has been described in the past due to overzealous use of radiation therapy for treatment of diffuse PVNS.

Conclusions

The described patient returned to full activity after recovering from the second surgery and reports no problems at 1 year follow up. The imaging is not suggestive of any recurrence and patient has been advised further regular follow up for surveillance. Such an incidence of double primary tumor in bones of ipsilateral foot is an unheard of scenario, and has never been previously reported to our knowledge.

References

23. Segler C.P. Irradiation as an adjunctive treatment of diffuse PVNS.
pigmented villonodular synovitis of the foot and ankle prior to tumor
24. Byers P.D., Cotton R.E., Deacon O.W., Lowy M., Newnau P.H.,
Sisons H.A., Thomson A.D.: The diagnosis and treatment of pigmented

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