

Diffuse-Type Giant Cell Tumour (Diffuse Pigmented Villonodular Synovitis) of the Ankle

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Abstract: Diffuse-type giant cell tumor (Dt-GCT), also known as pigmented villonodular synovitis, is a rare benign synovial tissue disease, that affects the foot and ankle joint only in a minority of cases. It may be locally aggressive and destructive and has the risk of recurrence after excision. The presentation is unspecific, so diagnosis is often delayed. Magnetic Resonance Imaging with contrast presents specific characteristics for Dt-GCT and is the best imaging modality for its diagnosis. Complete surgical resection is the preferred treatment when technically feasible. Adjuvant therapies such as radiotherapy may be considered in recurrent or selected cases. We present a case of a 37-year-old patient, that was diagnosed with Dt-GCT of the anterior ankle only six years after disease onset, though still with minor local destruction. Complete surgical excision was obtained and resulted in remittance of symptoms and an excellent functional result, without disease recurrence.

Keywords: Diffuse-type giant cell tumor; Pigmented villonodular synovitis; Foot and ankle; Surgery.

INTRODUCTION

Pigmented villonodular synovitis (PVNS) is a rare synovial tissue disease that despite being benign may be locally aggressive [1]. Its first description is attributed to Chassignac [2] in 1852, but it was Jaffe who coined the term PVNS.

Later, Granowicz and Mankin [3] classified the disease in two forms, localized and diffuse, which may represent a spectrum of disease. The localized form presents as a defined, pedunculated mass, while diffuse types correspond to infiltrative lesions of the synovial lining, which may lead to bony erosions and subchondral cysts.

The World Health Organization (WHO) classified these tumors as giant cell-rich tumors (GCT). The diffuse type is now categorized as diffuse-type giant cell tumor (Dt-GCT), which differentiates it from the localized form and those tumors arising from tendon synovium (giant cell tumors of the tendon sheath)[4]. Although the WHO terminology is the one currently in use, in surgical literature is still common practice referring to these entities as PVNS.

The estimated incidence of Dt-GCT is 1.8 new cases per million per year. The knee is by far the most commonly affected joint, followed by the hip. The ankle and foot are affected only in 2,5 to 7% of cases [5,6].

Total synovectomy is usually the procedure of choice for the treatment of symptomatic patients, but given the anatomical characteristics and multiple joints present in the foot and ankle region, it can be technically challenging in some cases.

Besides the local aggressiveness of the Dt-GCT lesion, recurrence is also a concern and is estimated to be around 12,5% when treated with surgery alone[7].

Clinical Case



Fig-1: Lateral radiograph of the right ankle, showing no significant changes.



Fig-2: Antero-posterior radiograph of both ankles, without significant bony changes.

A 37-year-old woman teacher, previously healthy, presented to our outpatient clinic with complaints of episodic but persistent dull pain in the anteromedial and proximal dorsum of her right foot, which had been present for almost six years. She described the pain as deep and aching, usually worsened by prolonged standing or walking. She denied any relevant previous trauma and did not practice sports regularly. She had a slight increase in volume in the anteromedial ankle, and a small (2-3 cm) elastic nodule

was palpable in the same region. Her primary care physician requested an ultrasound early in the course of the disease, which described the presence of a hypochoic 30mm bosselated formation deep to the flexor hallucis longus, compatible with synovial disease. She also had an x-ray, which was unremarkable (Fig. 1 and 2). It had been assumed to be a case of synovitis and treated with NSAIDs and physical therapy, but her symptoms had little improvement.



Fig-3: Sagittal T1 weighted image.



Fig-4: Sagittal T2 weighted image.



Fig-5: Sagittal T1 weighted image with gadolinium contrast enhancement. Intense contrast capitation.

After the evaluation at our outpatient clinic, a Magnetic Resonance Imaging (MRI) exam was ordered, which showed an expansive lesion with 2,8 x 2,7 x 3,1cm, with slight erosive changes in the dorsum of the talar neck and head, that was hypointense in T1 and T2 weighted images (Fig. 3 and 4), but showed intense gadolinium contrast enhancement with magnetic susceptibility artifacts (Fig. 5). These findings were very suggestive of a Dt-GCT.

Given the clinical presentation with persistent symptoms, we opted for surgical treatment through

open synovectomy. An anteromedial linear incision was performed over the palpable mass, along the tibialis anterior tendon. After subcutaneous dissection, the tibialis anterior tendon was identified and retracted laterally, and deeply to it, a firm, nodular, chocolate-colored mass was identified. Dissection was performed around the mass, and it was possible to excise it completely (Figs. 6 to 10). The erosions on the talar neck were superficial, did not involve articular surfaces and required no specific treatment.



Fig-6: Superficial dissection and surface of the mass.



Fig-7: Partial dissection and reflection of the mass.



Fig-8 Talar neck and head exposed after complete resection.

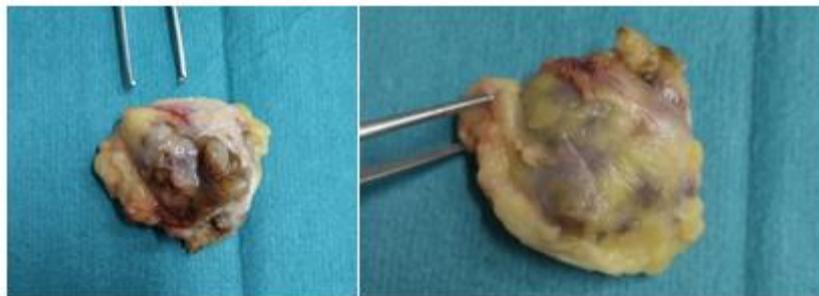


Fig-9 and 10: The excised mass, with nodular contours and a irregular brown/chocolate coloured surface.

The specimen was sent for pathology analysis. The report indicated that the sample showed villous and nodular proliferation, as well as multiple mononuclear

and multinuclear cells present and abundant hemosiderin deposition (Fig. 11, 12 and 13), which was consistent with the diagnosis of Dt-GCT.

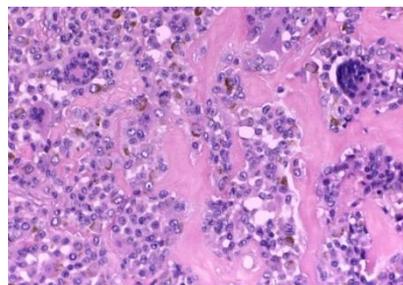


Fig-11: High powered hematoxylin-eosine stained specimen. Visible multiple mononuclear cells (some containing hemosiderin), some giant multinuclear cells, and collagen deposition.

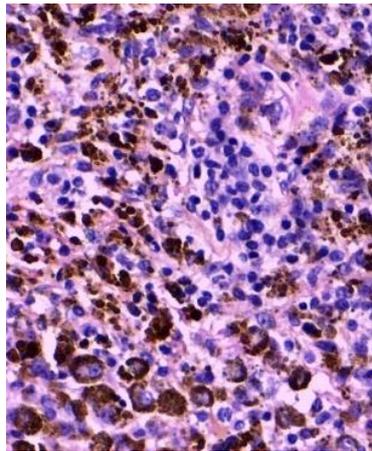


Fig-12: High powered Hematoxylin-eosine stained specimen. Segment with abundant siderosomes.

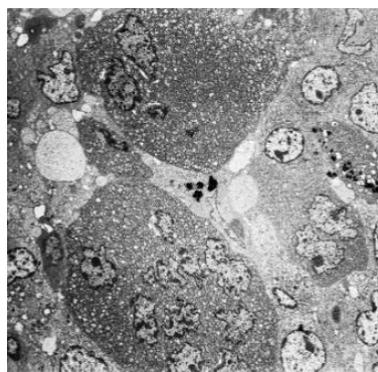


Fig-13: Electron microscopy of the specimen, detail of a multinuclear giant cell.

The patient had an uneventful recovery, had sutures removed at two weeks postoperatively, was allowed partial weight bearing with crutches for three weeks, full weight bearing onwards, and did not require physical therapy. She was symptom-free afterward and remained well and without signs of recurrence over a three-year follow-up period.

DISCUSSION

Patients with GCT of the foot and ankle may present with variable symptoms depending on the location and type of disease, pain and swelling being the most common complaints. Stiffness, locking, and catching may be present less frequently. In some cases, Dt-GCT may cause erosive and destructive joint changes, which create complaints of their own and may further confuse the diagnosis in late appearing patients.

Given the unspecific and often intermittent complaints, diagnosis is often delayed, with case series referring mean times to diagnosis from 15 to 28 months (range 3 to 70 months)[7,8].

The etiology of Dt-GCT is still under debate. The finding of hemosiderin deposition in these tissues led to proposing local hemorrhage or trauma as a possible cause[9]. Others proposed a neoplastic or inflammatory origin[6].

Regarding investigation, MRI is the method of choice. It allows to accurately assess location, the extent of the lesion, as well as possible erosions or local invasion present, which are important elements to take into account in surgical planning. Dt-GCT is typically low to intermediate signal in T1 and T2 and shows intense post contrast enhancement. The difference between the hemosiderin laden synovial tissue and adjacent structures produces a typical “blooming” picture on gradient echo sequences[10].

Total surgical excision is considered the procedure of choice if the lesion is accessible. The main concern lies with the risk of local recurrence, which was reported in older smaller case series as 0 to 40% in the cases of foot and ankle[11,12]. More recently, a study pooling the results of 49 cases treated with surgery alone indicated that the risk of recurrence was around 12 with 2%, and most of the recurrent cases corresponded to either incomplete or intralesional excision[13]. Open surgery is still commonly performed, though there have been some cases treated arthroscopically or with arthroscopy assistance with good results[7,11]. However, there are far too few cases to provide definite recommendations.

Given the concern of recurrence, some authors have used adjuvant radiotherapy as part of primary treatment[13,14]. There were no recurrences in the

group treated with this method, but there were some cases of local morbidity. Given the relatively low rate of recurrence with surgery alone, most advocate for that option for primary cases, leaving the role of radiotherapy for cases of recurrent disease or incomplete resection.

Local instillation of Yttrium-90 has been used before also with the same intent of lowering recurrence[15]. However, significant local adverse effects have led to its discontinuation for this indication[16].

More recently, the use of imatinib and other tyrosine kinase inhibitors has been under investigation as systemic therapy for this condition. The largest case series reported beneficial effects in 20 of 27 patients[17, 18] ranging from pain improvement to complete remission. It was offset by toxicity in 22% of patients and cost. Despite this, it might be an option to consider in patients with recurrent, advanced, or disease present in difficult anatomical locations.

If present, local bony or cartilage changes might require specific treatment, and it should be included in the surgical plan in advance. Simpler defects such as subchondral cysts may be curetted and filled with bone graft, while larger defects might benefit from osteochondral auto or allografts[8], and more advanced joint destruction might even need fusion or arthroplasty. Finally, the complex anatomy and multiple joints and tendons in the foot and ankle may create specific challenges depending on the location of the tumor, again stressing the paramount importance of adequate surgical planning.

CONCLUSION

Dt-GCT, formerly known as PVNS, is a rare, benign but locally aggressive disease. It presents uncommonly in the foot and ankle. Diagnosis is frequently delayed, and local destructive changes may be present. Adequate surgical resection is the treatment of choice if feasible, with adjuvant procedures in mind for specific cases.

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