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Case report

Glomangiomatosis of the lower leg☆

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ABSTRACT

This paper demonstrates a case of multiple glomangiomas, or glomangiomatosis, including clinical presentation, imaging appearances, and subsequent management. Differentiating features from typical glomus tumors are described. To the best of our knowledge, this is the first reported case of a glomangioma involving the distal tibiofibular syndesmosis.

Introduction

This case illustrates a rare and clinically significant pathology in a patient presenting with atraumatic unilateral ankle pain and swelling. Glomangiomas are a rare form of atypical glomus tumor, within the pericytic (perivascular) tumor subtype in the World Health Organization (WHO) classification system for soft tissue tumors [1].

Case report

A 65-year-old female presented with a 2-year history of progressive atraumatic unilateral ankle pain and swelling. Clinical examination revealed a palpable mass at the anterolateral lower leg. MRI of the ankle demonstrated multiple extramuscular soft tissue masses within the lower leg, the largest mass measured 4.5cm and involved the distal tibiofibular syndesmosis (Figs. 1A and B, Fig. 2A). Smaller masses were seen at the level of the distal tibial metaphysis and ankle joint, respectively (Figs. 2B-C). The masses were encapsulated and internally heterogeneous. They were predominantly T1 iso-to-hyperintense, SPAIR hyperintense and demonstrated heterogeneous enhancement. There was trace perilesional edema. Ultrasound-guided percutaneous biopsy of the largest mass was performed. Histological section demonstrated numerous thin-walled blood vessels invested with abundant bland epithelioid cells with distinct cell membranes (Fig. 3A). There was no cytological atypia or necrosis, and mitoses were rare. Immunohistochemical staining demonstrated strong Actin M

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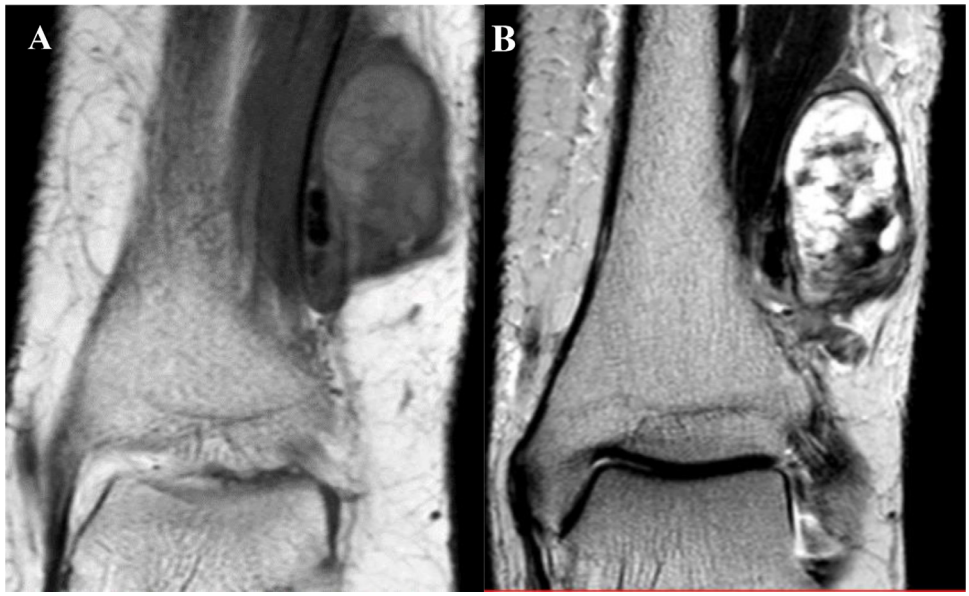

Fig. 1 – MRI of the ankle, coronal images at the distal tibial meta-diaphysis. (A) T1-W image at the demonstrates an intermuscular mass abutting and displacing the muscles of the anterior compartment. The mass is encapsulated, heterogeneous and predominantly T1 iso-hyperintense. (B) T2-W image demonstrates an intermuscular mass with an incomplete T2 hypointense capsule. The mass is heterogeneous and predominantly T2 hyperintense.

and Actin S positivity in the epithelioid cells (Fig. 3B). The vascular component was strongly CD31 and CD34 positive. S100 staining was negative. The proportion of vascular structures invested by glomus cells was supportive of a diagnosis of glomangioma, in contrast to a glomus tumor where the vascular component would be less prominent.

Discussion

Glomangiomas are benign glomus tumors with features of diffuse angiomatosis and no tendency towards malignant change [2]. They are a subtype of atypical glomus tumor as it falls outside the normal clinical and/or histologic spectrum of typical glomus tumors. It has an estimated incidence if 2 in 1,000,000 and accounts for 2%-3% of glomus tumors in adults [3–5]. Both typical glomus tumors and glomangiomas are painful and typically occur in the extremities [5,6]. Typical glomus tumors are most commonly subungual and single. Contrastingly, glomangiomas are deep, often extensive, and may be multiple. In comparison to typical glomus tumors [7], glomangiomas have a high risk of recurrence at 60%-90% [8,9]. The majority of glomangiomas occur in the lower (36.4%) and upper (27.3%) limbs. A significant proportion (36.4%) occur in unusual locations including the head and neck, chest wall, paravertebral regions [8]. To the best of our knowledge, this is the first reported case of a glomangioma involving the distal tibiofibular syndesmosis. Glomangiomas are typically seen as soft tissue masses that are homogenously T1 hypo-to–isointense, heterogeneously T2 hyperintense and demonstrate post-contrast enhancement [3,4,8]. This case demonstrates non-fat suppressing T1 hyperintensity (Fig. 1A), most likely reflecting internal haemorrhage.

Suspected cases of glomangioma should be managed in a tertiary tumor referral center [7]. MRI is advantageous in evaluating lesion extent, location, adjacent neurovascular structures and the detection of non-palpable lesions. Surgical resection is the treatment of choice and post-resection surveillance is required due to risk of recurrence. In cases of recurrence, further surgical intervention, including amputation, may be required and stereotactic body radiation therapy has been described [5]. Management is often conservative, balancing the need for complete surgical excision with associated surgical morbidity [10]. Following multi-disciplinary-team and patient discussion, conservative management with clinical and radiological follow-up was favored in this case.

Glomangioma should be considered when deep soft tissue masses are seen in an extremity. Differential diagnosis for soft tissue masses is broad [11]. Imaging appearances are often indeterminate, and biopsy is frequently required. As a general guide, benign neoplasms are small, superficial, and homogenous while malignant neoplasms are more often large, deep, heterogeneous and demonstrate osseous or neurovascular invasion. Vascular malformations would demonstrate serpentine morphology, phleboliths and can cross compartments.

In summary, this case describes the clinical presentation, imaging appearances, and subsequent management of multiple glomangiomas. Differentiating features from typical glomus tumors and broad differential diagnoses are also described.

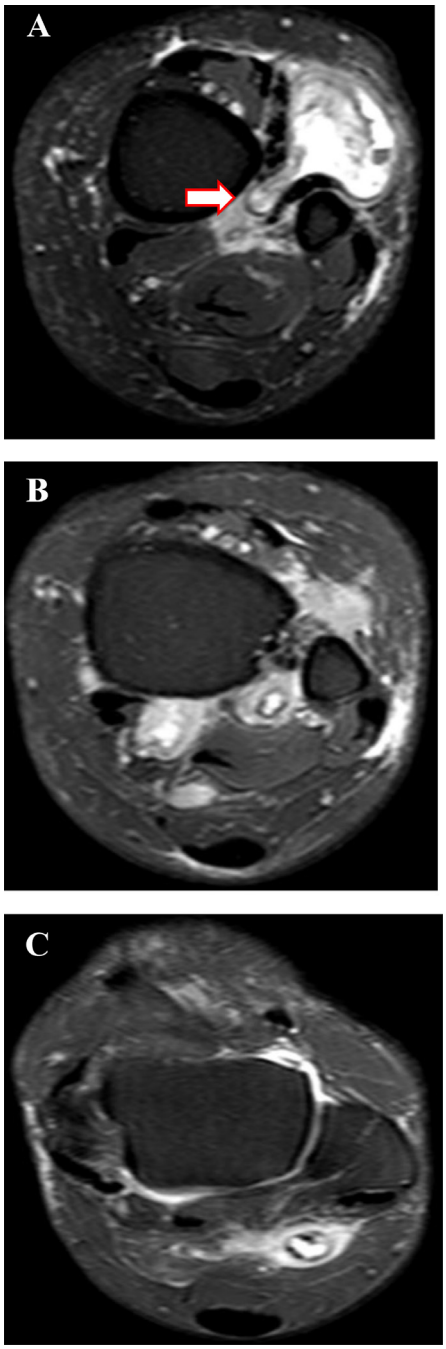

Fig. 2 – MRI of the ankle, axial SPAIR-W images. (A) Image at the distal syndesmosis demonstrates a large intermuscular mass in the anterior compartment with extension to involve the distal tibiofibular syndesmosis (arrow). The mass is encapsulated, SPAIR heterogeneously hyperintense, and there is trace perilesional edema. (B) Image at the distal tibial metaphysis demonstrates a small intermuscular mass in the posterior compartment interposed between flexor hallucis longus and tibialis posterior muscles. (C) Image at the ankle joint demonstrates a small intermuscular mass posterior to the peroneal muscles and anterior to the distal achilles tendon.

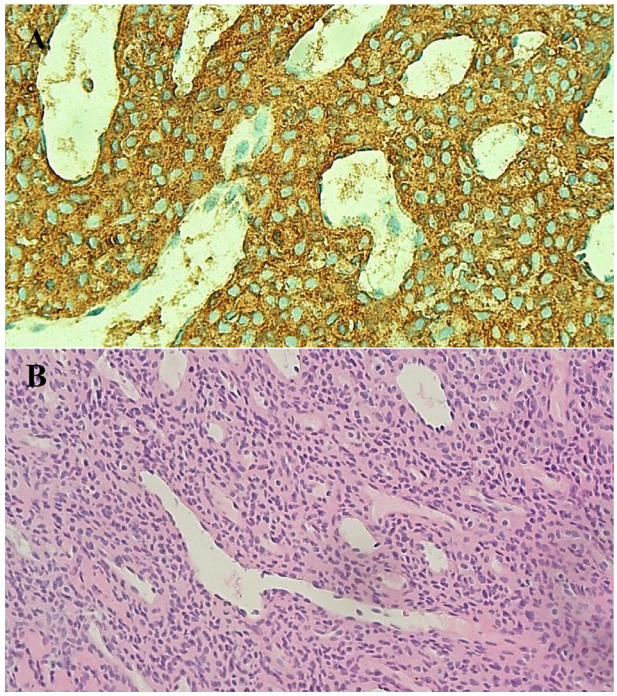

Fig. 3 – (A) Smooth muscle actin staining is diffusely positive in glomus cells (ACTS, original magnification 400x). (B) Uniform, small round blue cells with distinct cells membranes (arrow) are seen investing vessels of varying sizes, including staghorn vessels (starred) (hematoxylin and eosin, original magnification 200x).

Patient consent

Informed consent was obtained from the patient involved in this case.

Authors' contributions

DG is primary author and responsible for drafting the manuscript. JH is responsible for assisting in drafting of manuscript and manuscript review. KR is responsible for drafting pathology component, acquiring pathological images and drafting pathology component of manuscript. EK is the supervisor and responsible for idea generation. EK, GO'T and CO'K are responsible for proof reading and manuscript review.

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