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Glomus tumor of the thigh: confluent with the periosteum of the femur

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Abstract True glomus tumor is rare. In the majority of cases it involves the hand, preferring the fingertips or nail beds. We report a patient with glomus tumor of the mid-thigh who presented with severe localized pain and limp. The imaging features are discussed and the English literature reviewed.

Keywords Thigh · Femur · Glomus tumor · MRI

Introduction

Glomus tumor is an uncommon benign lesion originating in the subcutaneous neuromyoarterial plexus. It typically presents as a painful subungual nodule predominantly in the upper extremity. In two comprehensive studies over

100 cases of glomus tumors have been reported. Twenty of these occurred in the superficial tissue of the knee and leg [1, 2]. We report a rare case where the tumor occurred at the mid-thigh.

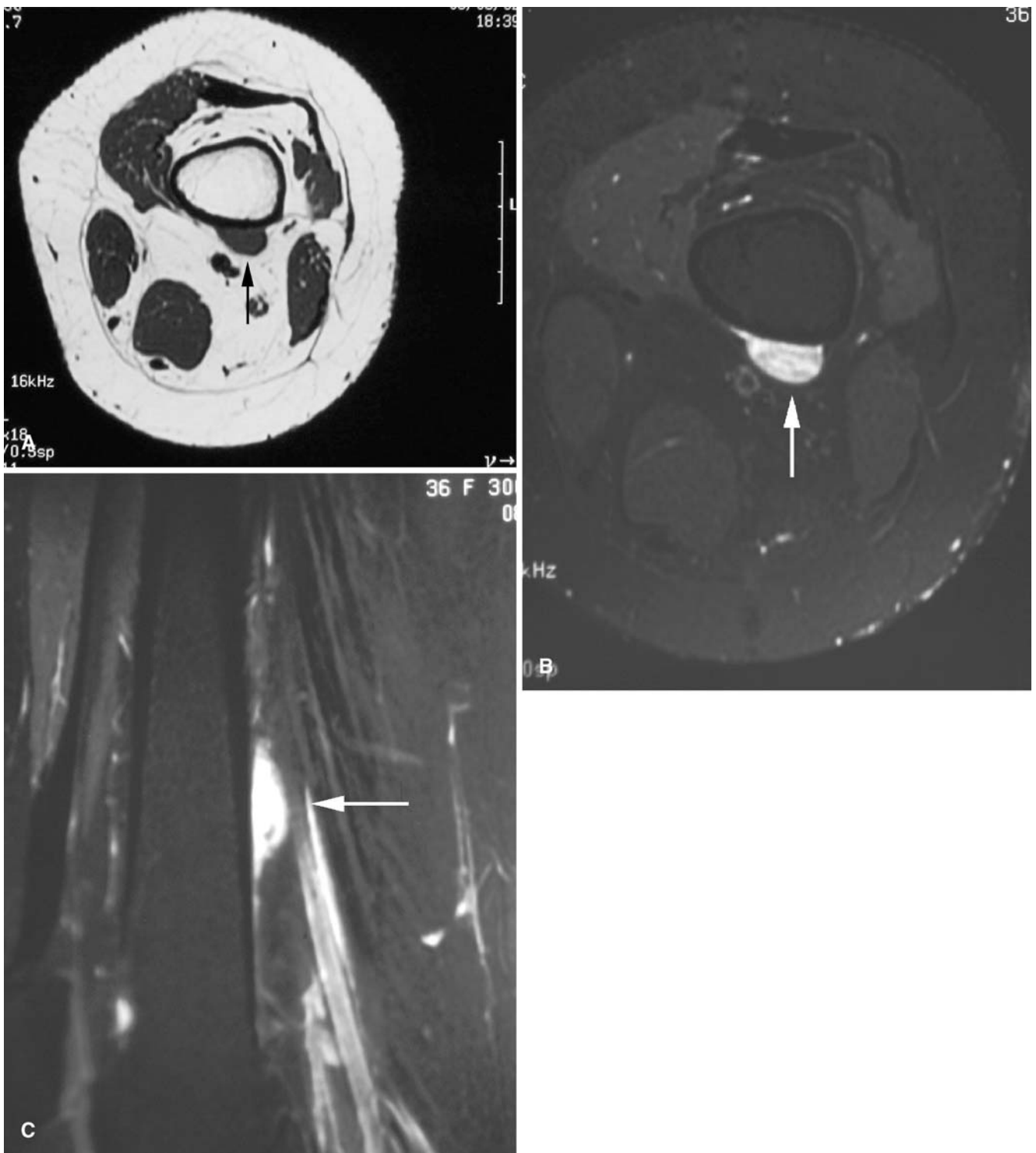


Fig. 1 **A** Axial T1-weighted image (TR/TE 750/14) of the left mid-thigh revealed a low signal intensity (SI) mass confluent with the posterior cortex of the femur (*arrow*). **B** On a STIR sequence

(TR/TE 5000/90) the mass was heterogeneously bright (*arrow*). **C** Sagittal image (TR/TE 750/14 fat suppressed) following contrast administration showed patchy enhancement of the mass (*arrow*)

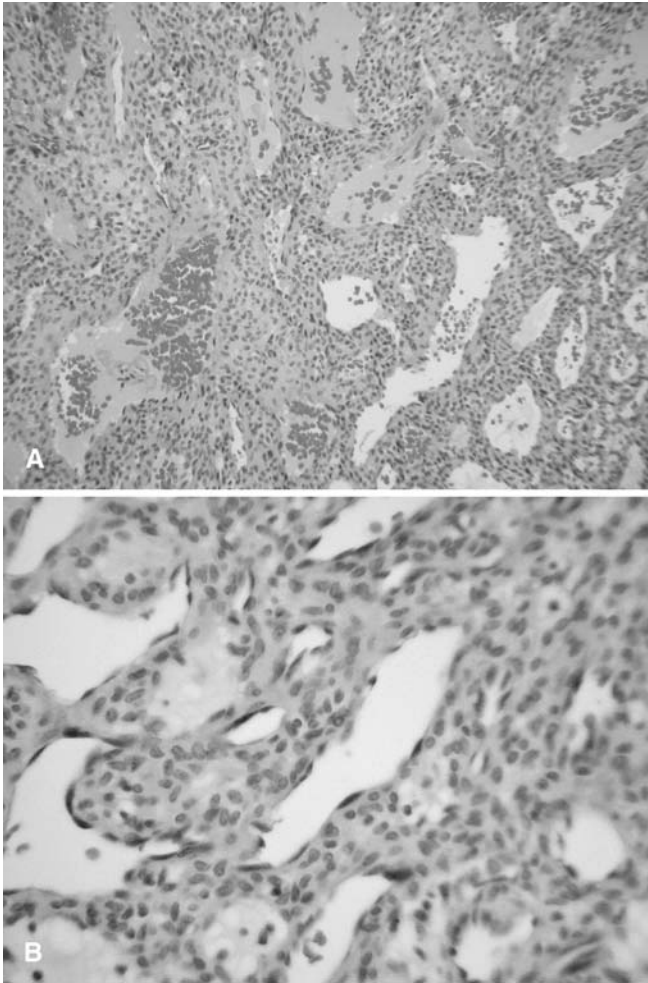


Fig. 2A, B Glomus tumor. **A** Multiple irregularly configured vascular spaces lined by bland endothelium compressed by round regular glomus cells ($\times 150$). **B** Glomus cells proliferating between vascular spaces demonstrate bland, round nuclei with finely divided chromatin, no hyperchromatism and minimal proliferative activity ($\times 350$)

In our case, based on imaging findings, an erroneous prebiopsy diagnosis of an osseous lesion was made and a soft tissue lesion was not considered.

Case Report

A 36-year-old woman presented at the orthopedic office with a complaint of discomfort in her left thigh and knee that had persisted for 6 months. She stated that 2 months previously she had been involved in an automobile accident. Since then, the pain had increased and become consistent, predominantly in the lower extremity, in the posterior part of the left thigh. Her past medical history was unremarkable.

She showed a slight limp. On physical examination, the posterior aspect of her left thigh was tender and this prevented evaluation of the area by palpation. The range of motion of the hip and knee was normal. The neurovascular examination was unremark-

able. Radiographs failed to demonstrate an abnormality. Nuclear scan ($^{99m}\text{Tc-MDP}$) revealed focal, slightly increased uptake at the distal one-third of the posterior aspect of the left femoral cortex. The remainder of the skeleton showed no abnormality.

MRI of the left thigh (Fig. 1) revealed a 0.9×2.0 cm mass at the posterior aspect of the cortex that appeared homogeneously hypointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images. Following contrast administration the mass showed heterogeneous enhancement. The differential diagnosis included infection of bone and osteoid osteoma.

At operation the mass was located at the posterior part of the femur, adjacent to the periosteum and adherent to it. It did not appear to scallop the periosteum or outer cortex. The mass was excised.

Histologic examination demonstrated a tumor composed of monotonous round to polyhedral cells arranged in small nests and intimately associated with variably sized capillary vessels. The cytoplasm was faintly staining and the nuclei appeared bland and occasionally clear. The cell proliferation associated with capillary vessels was primarily external to the vessels in distribution. There was almost no mitotic activity and no pleomorphism noted (Fig. 2).

On follow-up 12 months later, the patient was free of symptoms and resumed her normal activity.

Discussion

True glomus tumors arise from the neuromyoarterial glomus bodies. There are direct connections between preterminal arterioles and efferent veins. The normal glomus cells control arteriovenous shunting and serve as a regulator of body temperature. True glomus tumors should be differentiated from glomus jugulare and glomus vagale tumors that derive from chromaffin cells occurring in the head and neck. Glomus tumors are rare. Since the first description of glomus tumor as a painful subcutaneous nodule by Wood in 1812 [3], it has been observed that in the majority of cases it occurs in the area of the fingertips or nail beds [4]. More than two-thirds of the cases involve the upper extremity [5, 6]; up to 75% occur in the hand [7, 8]. The incidence of this lesion ranges between 1% and 4.5% of all tumors of the hand [9, 10, 11] but it may be found in the stomach, mediastinum, nasal fossa and synovial membrane. The tumor arises almost exclusively in the soft tissue. Secondary involvement of the neighboring bone occurs in up to 60% of reported cases. Primary intraosseous glomus tumors may occur in the phalanx metacarpal bones or long tubular bones such as the femur, ulna and fibula [12, 13]. Only a few case reports have described primary intraosseous glomus tumor of the spine [10, 14].

True glomus tumor may be observed at any age. In most instances it occurs in the fourth or fifth decades of life [4]. The usual presentation is a tiny blue tumor beneath the nail that manifests with sharp pain, exquisite point tenderness over the area and extreme sensitivity to cold. The tumor may be small and not palpable. Exposure to cold or minor trauma may cause an acute attack of pain. It is benign and complete removal is the treatment of choice. Secondary involvement of the bone by soft tissue

glomus tumor is fairly common and may occur in up to 65% of the cases [6, 14, 15, 16], particularly in the hand. In a comprehensive study Masazumi et al. [1] reviewed 63 patients with glomus tumor of the soft tissues. Forty-six involved the upper extremity, 15 occurred in the lower extremity, seven around the knee and six in the leg. The majority were found on the body surface, mostly in the corium and adjacent to subcutaneous tissue. Heys et al. [2] in 1992 reviewed their material. Only seven of 43 cases occurred in the lower extremity. The majority involved the upper extremity, neck and abdomen. More recently, Abou Jaoude et al. [17] found glomus tumor in two instances in the soft tissue of the thigh. None of them extended to the bony cortex.

Primary intraosseous glomus tumor is rare. Bahk et al. [18] recently reviewed the world literature and found 17 such cases. In the majority of cases the tumor involved the phalanges of the fingers. In one instance the tumor arose in the ulna, in another in the fibula.

Pambakian and Smith [19] have described coccygeal glomus tumor in two patients who presented with coccygodynia. Excision of the coccyx and pericoccygeal tissues, containing glomus tumor of the coccygeal body, relieved the symptoms. They concluded that glomus tumors might be the cause of symptoms of coccygodynia [19].

Radiologic Features

Primary glomus tumor of the soft tissue may extend into the adjacent bone producing a well-margined erosion. The margin may be sclerotic. Intraosseous glomus tumor appears as a well-defined osteolytic lesion confined to the bone, usually in the distal phalanx, that mimics inclusion cyst. Lytic lesions in the metacarpal should be differen-

tiated from aneurysmal bone cyst (ABC) enchondroma or giant cell tumor. Considering the slowly growing patterns, malignant lesions such as metastatic carcinomas of breast, lung or kidney are easy to eliminate. The borders are well defined. MRI shows decreased signal intensity on T1-weighted images and increased signal intensity on T2-weighted images without evidence of edema in the surrounding bone. In our case the nodule measured approximately 2×1 cm in diameter. It was located deep in the posterior aspect of the thigh. The nodule abutted the periosteum. It was not palpable; however, the severe localized pain pointed to the exact site, which was clinically suggestive of an infection. The unusual location was very atypical for a glomus tumor. MRI demonstrated decreased signal intensity on T1-weighted images and localized heterogeneous increased signal intensity on T2-weighted images.

Pathology

The pathologic appearance of the glomus tumor is a pink or purple mass that measures a few millimeters in diameter [6]. Reactive capsule may surround it [20]. Glomus tumor is typically a highly vascularized neoplasm with uniform rounded cells encircling capillary-sized vessels [21]. These cells are round to polyhedral and contain granular eosinophilic cytoplasm [8, 22]. The nuclei have finely dispersed chromatin. The stroma may show loose, hyalinized or myxoid pattern. In such cases the vascular channels and cells are separated from one another. Occasionally, the glomus cells are tightly packed with little stroma. The cells lack nuclear atypia and mitotic activity is rare. Excision of the tumor as well as of the involved part of the bone is curative.

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