



An Unusual Cause of Knee Pain: Periosteal Glomus Tumor of the Distal Femur

무릎 통증의 드문 원인: 원위 대퇴골 골막주위 사구 종양

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Glomus tumors are rare neoplasms that characteristically occur in subungual regions, but may also be found in other regions of the body. The clinical diagnosis of this tumor may be difficult if the tumor is located in an extradigital site. Most extradigital glomus tumors form in superficial locations. Herein, we present the case of a 34-year-old woman who experienced chronic knee pain with pinpoint tenderness resulting from a deep-seated periosteal glomus tumor of the distal femur. Extradigital glomus tumors should be considered in the differential diagnosis when characteristic clinical features and imaging findings indicative of glomus tumors are present, even if the tumor is located within deep tissues.

Index terms

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INTRODUCTION

Glomus tumors are perivascular neoplasms, derived from modified smooth muscle cells of a neuromyoarterial glomus—commonly termed a glomus body—with the function of temperature regulation through arteriovenous shunting of blood (1). Glomus tumors are rare and have been reported in approximately 1.6% of patients with primary soft tissue tumors of upper and lower extremities (2). The lesion characteristically occurs in a digital subungual location, presenting with well-localized pain that is exacerbated by temperature changes. Although tumors are most often located on the digits, up to 45% of glomus tumors are extradigital, with common locations being the hands, arms, legs, and trunk (3, 4). Glomus tumors may also develop in sites where normal glomus bodies may be

sparse or even absent, such as the gastrointestinal tract, liver, lung, and trachea, as well as intraosseous and intraneural locations (3). In the musculoskeletal system, extradigital glomus tumors have been described around the shoulder, elbow, wrist, hip, knee, and ankle (5). There have been several reported cases of extradigital glomus tumors in the thigh. In the majority of these cases, the tumors were located superficially, such as the subcutaneous and intramuscular tissues (6). To our knowledge, there have been only three reported cases of periosteal glomus tumors adjacent to the femoral cortex (7-9). We report a rare case of a histologically proven glomus tumor found in the periosteal location of the distal femur.

CASE REPORT

A 34-year-old woman presented with a 5-year history of chronic pain in her left thigh. The patient had no history of trauma, other medical, or surgical history. On physical examination, she reported pinpoint tenderness on palpation of the anteromedial distal thigh. The range of motion of the knee was normal. Radiographs did not demonstrate any abnormality. Magnetic resonance imaging (MRI) of the left thigh revealed a 10- × 8-mm, well-circumscribed mass at the anteromedial aspect of the femoral cortex that appeared hyperintense on T2-weighted images and isointense relative to muscle on T1-weighted images (Fig. 1A, B). The mass was strongly and homogeneously enhanced after contrast agent administration (Fig. 1C). The adjacent femoral cortex was normal. Ultrasonography (US) was performed for preoperative skin marking, this procedure showed that the mass was solid and hypoechoic, and there was no obvious Doppler signal (Fig. 1D). The differential diagnosis included vascular lesions such as hemangioma.

Surgery was performed to remove the lesion at the anteromedial aspect of the femoral cortex and a palpable mass, approximately 1 cm in size, was excised. Histologic examination revealed solid sheets of tumor cells that were interrupted by vessels of variable size. The round nuclei were centrally placed

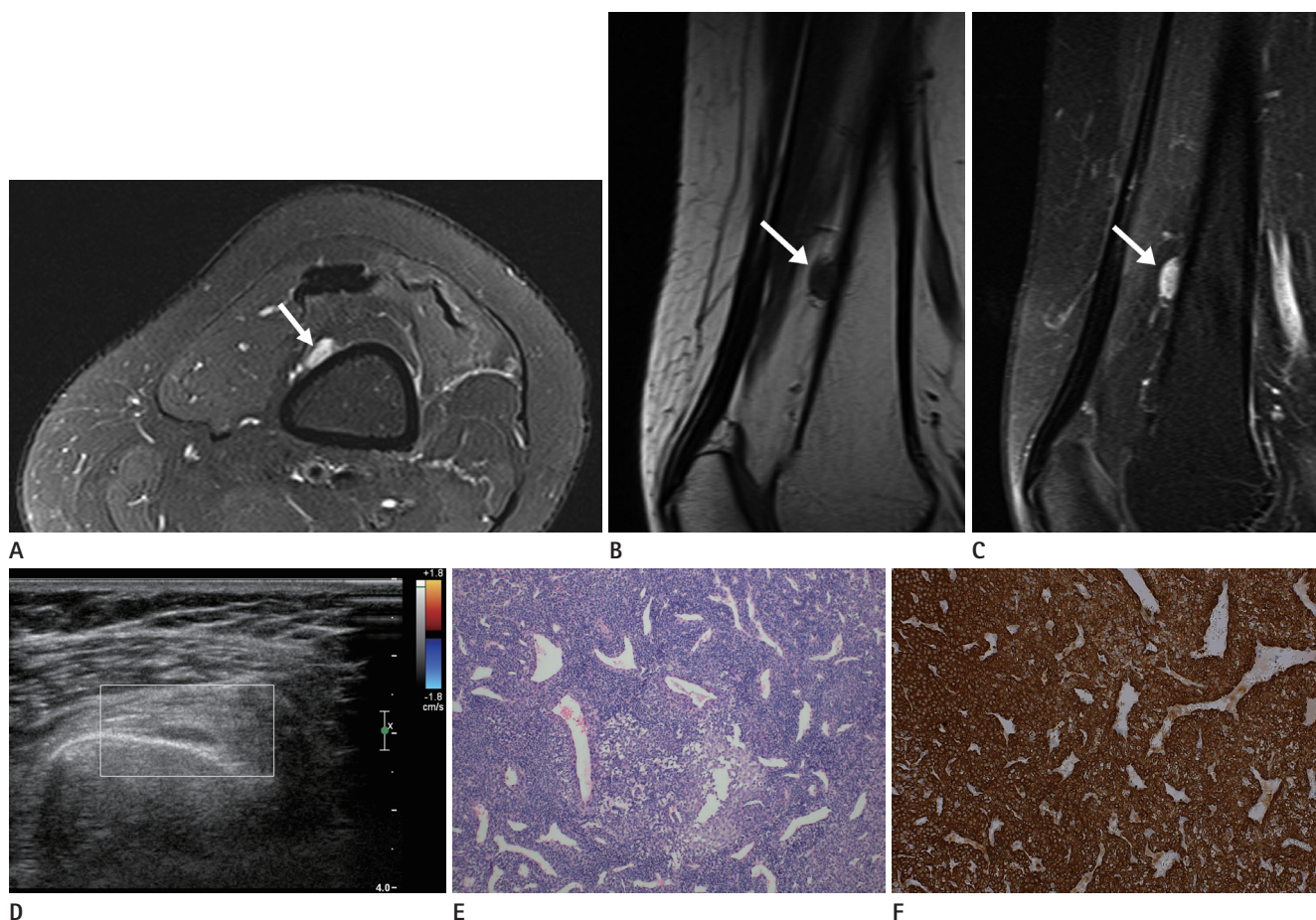


Fig. 1. Periosteal glomus tumor of the distal femur in a 34-year-old woman, complaining chronic knee pain with pinpoint tenderness.

A–C. Axial T2-weighted fat-saturated (**A**), sagittal T1-weighted (**B**), and contrast agent-enhanced T1-weighted fat-saturated (**C**) magnetic resonance images of the left thigh show a 10-mm, well-circumscribed mass at the anteromedial aspect of the femoral cortex. The mass appeared hyperintense on T2-weighted images and isointense on T1-weighted images with homogeneous and vivid enhancement after contrast enhancement (arrows).

D. Ultrasonographic image demonstrated a hypoechoic mass adjacent to the left femoral cortex. On color Doppler study, Doppler signal was not obvious.

E. Pathological examination with hematoxylin and eosin staining (× 100) revealed that the tumor comprised solid sheets of glomus cells with multiple variable-sized vessels.

F. Tumor cells show strong immunoreactivity for smooth muscle actin immunostaining (× 200).

with lightly eosinophilic cytoplasm, and no atypical mitotic figures were observed (Fig. 1E). Immunohistochemistry revealed that the tumor cells were strongly positive for smooth muscle actin (Fig. 1F). The histology was characteristic of a solid-type glomus tumor. At the 4-month follow-up, the patient reported relief from pain, and there was no evidence of residual tumor on the follow-up US.

DISCUSSION

Glomus bodies are located in the stratum reticularis of the dermis and are particularly numerous in the hands and feet. They are composed of an afferent arteriole, an arteriovenous complex with a neurovascular reticulum, and efferent venules. Glomus tumors usually occur in areas rich in glomus bodies, such as the subungual regions of digits or the deep dermis of the palm, wrist, and forearm (1). However, these tumors have also been observed in extra-cutaneous locations not known to contain glomus bodies, where they presumably arise from perivascular smooth muscle cells that differentiate into glomoid cells. Recently reported review articles suggest that extradigital tumor distribution may be more frequent than has been generally assumed (3, 4). When glomus tumors occur outside the fingers, they are found in a different demographic group, with a higher male to female ratio of 4:1 as compared to 1:2 in the fingers (4). The most common symptoms are pain and localized tenderness. Glomus tumors in extradigital locations contain nerve bundles that correlate with the presenting symptoms of pain. However, the classic clinical features of well-localized pain and cold hypersensitivity may not be present in extradigital sites, and preoperative diagnosis can be difficult. Moreover, the unusual locations of these tumors make application of diagnostic tests cumbersome. The average duration from symptom onset to diagnosis is 7 years (4).

On radiographs, erosive changes may be present in a minority of subungual glomus tumors. However, radiography is less useful in diagnosing extradigital lesions. On US, glomus tumors present as circumscribed, solid hypoechoic masses with small cystic-appearing spaces. Marked blood flow within the masses and adjacent feeding vessels can be observed on Doppler examinations (10). However, similar to our case, a case with juxtacortical glomus tumor of the distal femur was identified on US with

no Doppler signal (8). MRI has proved to be the most sensitive modality for diagnosis, especially for smaller tumors. Most often, a characteristic, well-circumscribed, T2 high and T1 intermediate signal intensity lesion is found with homogeneous enhancement (10). Although MRI and US were not shown to provide a specific diagnosis of the extradigital glomus tumor, they did show its precise location and size. The imaging findings in our case are consistent with the findings in previous studies of extradigital glomus tumor.

On histopathological analysis, glomus tumors are typically composed of three components: vascular, smooth muscle, and neural components (3). The classic histological features of the glomus tumor include angiocentric uniform sheets of cells with oval nuclei, forming a perivascular “collar” around vessels. The histologic features differentiate the three tumor variants. The solid form has poor vasculature and scant smooth muscle components, while glomangiomas have a prominent vascular component, and glomangiomyomas are composed of prominent vascular and smooth muscle components (1). Solid glomus tumors, the subtype observed in our case, are the most common (about 75%), followed by glomangiomas and glomangiomyomas. Although the exact etiopathogenesis and cellular origins of this tumor are poorly understood, several lines of evidence suggest a modified pericytic/modified smooth muscle phenotype for glomus tumor.

In conclusion, this case details the clinical presentation of exaggerated pinpoint tenderness on palpation and a well-enhanced periosteal mass at the distal femur on MRI. Extradigital glomus tumors should be considered in the differential diagnosis when characteristic clinical features and compatible imaging findings are present, even if the tumor is found within deep tissues.

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무릎 통증의 드문 원인: 원위 대퇴골 골막주위 사구 종양

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사구 종양은 주로 손발톱 밑에서 특징적으로 발생하는 드문 종양이며, 이 외에도 신체의 다른 어느 곳에서도 발생할 수 있다. 종양이 손가락이나 발가락 이외의 위치에서 발생하는 경우 임상적 진단이 어려울 수 있으며, 대부분이 얇은 위치에서 발생하는 것으로 보고되었다. 본 저자들은 원위 대퇴골의 골막에 인접하여 발생한 사구 종양으로 인한 촉진 시 무릎 통증을 경험한 34세 여자 환자의 증례를 보고하고자 한다. 자기공명영상 및 도플러 초음파 검사의 소견과 임상 증상을 함께 제시하는 바이다.

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