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Case Report

Brace yourself: an unusual case of knee pain, an extradigital glomangioma of the knee

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ABSTRACT

The differential diagnosis for knee pain is extensive. Glomus tumors comprise approximately 1.6% of soft-tissue tumors in the extremities. Classic subungual tumors occur more frequently in women, whereas ectopic locations are more common in men. Unusual locations include the stomach; lungs; trachea; bones; intestines; fallopian tubes; and intraneural, neuromal, and intravenous locations. We present the case of a 50-year-old man with a 12-year history of enlarging right knee mass found to be a glomangioma. This case report discusses the incidence, presentation, imaging characteristics, histology, and management of glomus tumors of the knee.

Case summary

We present a 50-year-old man with a past medical history of type-II diabetes and thromboembolic disease complaining of a 12-year history of a gradually enlarging mass located in the superficial soft tissues of the right knee. The mass was described as moderately tender to friction, pruritic, and sensitive to cold exposure. The patient presented to the emergency department at our institution for persistent bleeding after an attempted incision and drainage of a knee "cyst" by his primary care provider. The physical examination revealed a 4.1 × 4.0 cm freely movable, rounded mass within the subcutaneous tissues of the right knee, immediately proximal and anterior to the patella.

Imaging findings

After the bleeding was controlled, the division of Musculoskeletal Radiology was consulted to perform an ultrasound of the lesion and assess for biopsy. The ultrasound revealed a solid mass that exhibited marked tortuous vascular flow as

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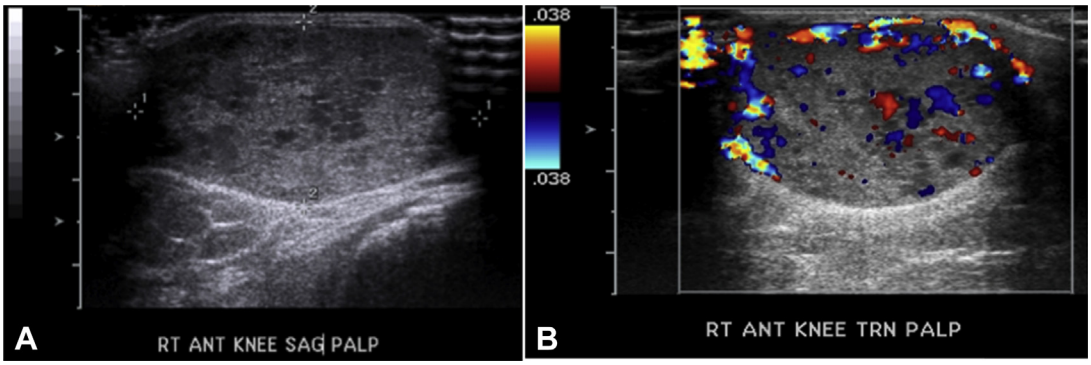

Fig. 1 – Ultrasound examination of the right knee. (A) There is a heterogeneous, hypoechoic, solid, and well-circumscribed mass corresponding to the patient's palpable complaint measuring 2.3 × 2.7 × 1.4 cm. (B) Color Doppler shows extensive vascular flow within the lesion.

evidenced on color Doppler interrogation (Fig. 1). Because of the ultrasound findings and the history of uncontrolled bleeding after a recent attempted incision and drainage, biopsy of the mass was postponed until a magnetic resonance imaging (MRI) with contrast could be obtained. Meanwhile, Interventional Radiology was consulted for the possibility of prebiopsy embolization.

The MRI revealed a mass measuring 2.6 × 5.0 × 3.0 cm (AP × CC × Trans) immediately deep to the skin surface and anterior to the proximal pole of the patella. The examination was negative for bone erosion or involvement of the quadriceps tendon insertion. A single septation was evident along the distal margin of the structure. The mass exhibited mildly heterogeneous T1 signal that was isointense to muscle. The T2 signal characteristics were also slightly heterogeneous but markedly bright. Contrast enhancement was intense and homogeneous (Fig. 2). At least 2 significant feeding vessels were associated with the mass.

After consulting with Interventional Radiology, it was agreed that the feeding vessels could be embolized before biopsy and total surgical excision. During the angiogram, a microcatheter was used to selectively catheterize the superior lateral branch of the superior geniculate artery. An early contrast blush with venous drainage was identified (Fig. 3). Subsequently, this feeding branch was embolized using 0.2 × 2.0 cm Hilal embolization microcoils (Cook Medical, Bloomington, IN). A similar procedure was used to embolize feeding branches of the middle lateral geniculate artery. A small tortuous feeding branch was also identified arising from the popliteal artery; however, this branch was deemed too small to access.

Post-embolization angiography and ultrasound examination revealed significantly decreased vascular flow within the mass. Subsequently, an ultrasound-guided, 18-gauge core biopsy was performed without excessive bleeding. The results of the biopsy revealed the diagnosis of a glomangioma.

Discussion

The differential diagnosis for knee pain is extensive. Glomus tumors are rare and comprise only 1.6% of soft-tissue tumors in the extremities [1–3]. Generally seen in the hand, it is uncommon to see these lesions in the knee, and misdiagnosis leads to long delays in definitive treatment. The average duration of symptoms is reported to be between 7 and 11 years with 2.5 medical consultations before diagnosis is made [2,4]. The delay can be even longer for atypical locations. Classic subungual tumors occur more frequently in women, whereas ectopic locations are more common in men [1,3]. Unusual locations, where glomus bodies do not exist physiologically, include the stomach; lungs; trachea; bones; intestines; fallopian tubes; and intraneural, neuromal, and intravenous locations [2,5].

The normal glomus body is an arteriovenous anastomosis, which allows it to function as a receptor to control blood pressure and cutaneous temperature regulation by modifying peripheral blood flow [2,5,6]. Glomus cells have similar properties to smooth muscle cells, which allows for their contractile ability. Rohrich et al. suggested that changes in temperature could lead to contraction of myofilaments in the glomus cells, resulting in an increase in intracapsular pressure that could be transmitted by the unmyelinated nerve fibers, leading to the most common symptom of glomus tumors, pain [7].

Glomus-type tumors are benign neoplasms, which develop from the normal neuromyoarterial glomus body [2]. Glomus bodies are located within the reticular layer of the dermis throughout the body. In an article by Deok-Woo Lee et al., 152 cases of glomus tumors were reviewed, yielding 73.4% digital tumors and 27.6% extradigital tumors [3]. Schiefer et al. found strikingly different results. In their review of glomus tumors, of the 221 patients reviewed over a 20-year period, they found 61% of all glomus tumors to be extradigital, suggesting that an extradigital location may be more common than was initially thought [2]. Heys et al. also found similar results in their 1992 study, showing 67% of glomus tumors to be extradigital [8].

Glomus tumors of the knee have been reported in variable locations, including subcutaneously, subsynovially, within the patellar ligament, within the fat pad, laterally at the fibular head, and in the popliteal region [1]. Our glomus tumor was found within the subcutaneous tissues and was associated with pain, pinpoint tenderness, and hypersensitivity to cold exposure, all features associated with subungual glomus

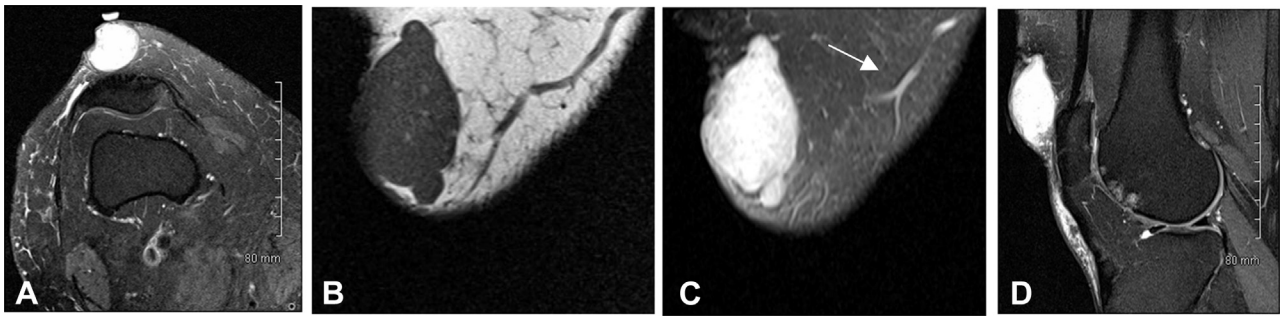

Fig. 2 – Magnetic resonance imaging (MRI) of the right knee with and without contrast. (A and D) Axial and sagittal shows a hyperintense mass anterior and superior to the patella. (B) Coronal T1 noncontrasted imaging shows a proton density fat saturation (PDFS) hypointense solid mass. (C) Coronal T1FS + C demonstrates homogenous contrast enhancement of the lesion with large feeding vessel (white arrow).

tumors in the literature. Multiple studies have found that extradigital glomus tumors often present with pain but not temperature sensitivity, making these ambiguous lesions even harder to identify.

These lesions can be either solitary or multiple. The etiology of solitary glomus tumors is unknown, although the development of multiple glomus tumors is associated with an autosomal-dominant mutation in the *glomulin* gene, localized to chromosome 1p21-22, which encodes for the glomulin protein [5]. Most glomus tumors are found in the 30- to 60-year old-age group. Glomus tumors have not been observed in individuals who are aged less than 1 year, presumably because glomus bodies form after that age; and they are not seen in the elderly population because glomus bodies undergo atrophy and degeneration [5].

On histopathological analysis, glomus tumors are comprised of vascular, smooth muscle, and neural components [4]. Glomus-type tumors have been classified into 3 groups based on the predominant cellular components: glomus tumor, glomangiomas, and glomangiomyomas. Specifically, solid-type glomus tumors consist of monomorphic round cells with eosinophilic cytoplasm and central round nuclei (Fig. 4). Glomangiomas consist of cavernous vascular spaces surrounded by several layers of glomus cells. Glomangiomyomas consist of spindle-shaped smooth muscle cells blended with glomus cells near the vascular spaces. Substance P and TRPV1 are highly expressed in glomus tumor cells, allowing easy differentiation from the surrounding stroma [3].

Malignant transformation is exceedingly rare but should be considered when the tumor presents with atypical features such as size >2 cm, deep location, infiltrative growth pattern, mitotic activity, nuclear pleomorphism, and necrosis [2,4,9]. Diagnosis is based on histologic features rather than clinical findings. These tumors generally do not metastasize, however, when they do it is often fatal. Glomangiosarcomas are painful dermal lesions with a predilection for the lower extremities [6].

The preferred methods of diagnosis are magnetic resonance imaging and ultrasound. An ultrasound may be used as the initial test of choice because of its low cost, speed, and ease of convenience for the patient. On ultrasound, glomus

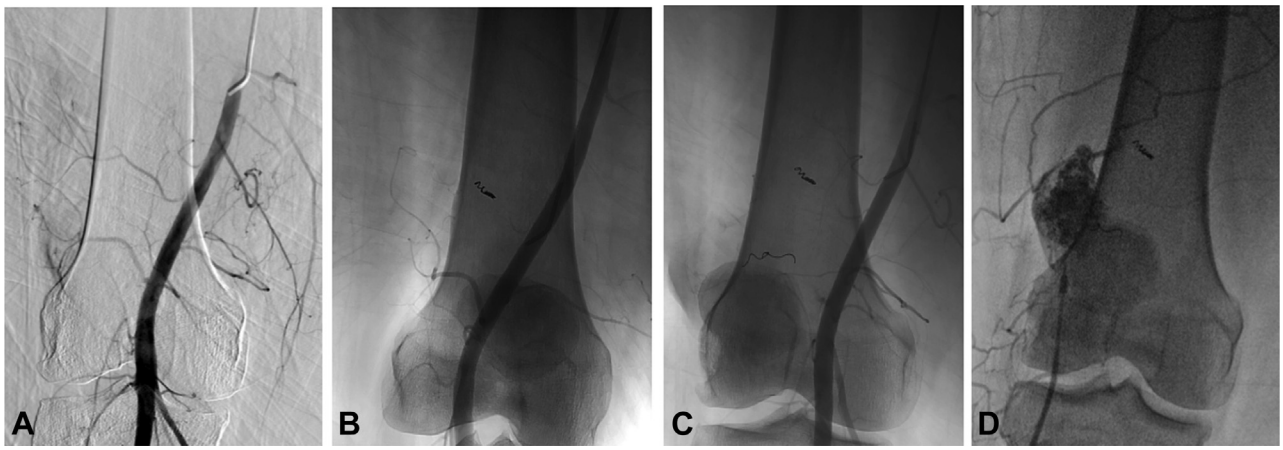

Fig. 3 – Right lower extremity angiography and intervention. (A) Pre-embolization angiogram via selective catheterization of the right superficial femoral artery (SFA) demonstrates 2 branches arising from the distal SFA/proximal popliteal artery supplying the lesion. (B and C) Successful coil embolization of the proximal (B) and distal (C) feeding vessels. (D) Direct puncture of the nidus of the glomangioma before alcohol ablation demonstrates a blush within the mass itself with multiple surrounding small serpiginous draining veins.

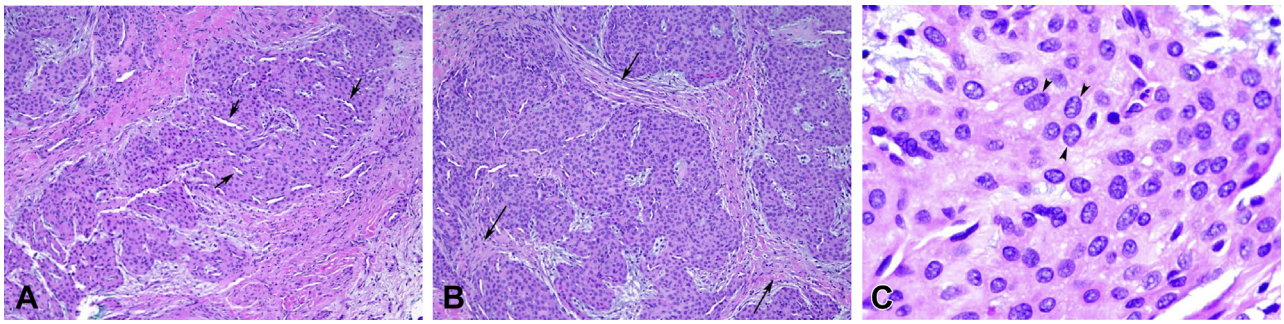

Fig. 4 – (A) Glomus tumor cells arranged around vessels, arrows pointing to vessels (H&E, 10×). (B) Glomus cells in nests and aggregates separated by stroma, arrows pointing to stroma (H&E, 10×). (C) Glomus cells have small, round, regular nuclei with indistinct nucleoli, arrows pointing to nuclei (H&E, 40×).

tumors are seen as round or ovoid hypoechoic masses. They have been reported to be hypervascular. Magnetic resonance imaging has proven to be the most sensitive imaging modality for the diagnosis of glomus tumors. The typical appearance of a glomus tumor by MRI is decreased signal intensity on T1-weighted images and increased signal intensity on T2-weighted images. When clinical diagnosis suggests glomus tumor, a negative MRI result should not impede excision because small size may be responsible for lack of detection and false-negative results [2]. The specificity of MRI has been estimated to be around 50% [2].

Treatment

The treatment of choice for an isolated glomus tumor is complete surgical excision. Small size and superficial location facilitates complete removal of these lesions. This often results in immediate resolution of symptoms, full recovery, and improved quality of life. Approximately 10%-33% of glomus tumors recur after conservative excision [2,3,7]. This is felt to represent either inadequate removal or multiple tumors. Inadequate excision may result in tumor recurrence within days to weeks, and in reoccurrence, symptoms may appear 2-3 years postoperatively [10].

Conclusion

Rapid detection and diagnosis of glomus tumors is important to avoid treatment delays, chronic pain, and disuse syndromes. Only 9%-20% of extradigital glomus tumors are correctly diagnosed by the primary physician [2,3]. The most common misdiagnoses in the remaining patients were neurogenic tumors, vascular tumors, pigmented nevi, epidermal cysts, lipomas, leiomyomas, and sarcoidosis [3]. Therefore, increasing our understanding and recognition of these benign lesions is essential in avoiding unnecessary costs, consultations, and complications. Glomus tumors should be added to the differential diagnosis for subcutaneous nodules when the classic triad of pain, cold sensitivity, and point tenderness is recognized.

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