

AN ANTERIOR FEMORAL NERVE GLOMUS TUMOR CAUSING TWENTY YEARS OF NEUROPATHIC PAIN: A CASE REPORT

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Background: Glomus tumors are a rare, yet painful, neoplasm commonly occurring in the subungual area of the digits. While uncommon, a glomus tumor can present in extradigital locations, leading to delays in diagnosis.

Case Report: We present a case of a 53-year-old man with a 20-year history of right thigh pain. Previous diagnostic workup and treatment with spinal cord stimulation and peripheral nerve stimulation did not yield resolution. Exam with ultrasound revealed a hypoechoic lesion. Upon targeted surgical exploration, a mass near the right anterior femoral cutaneous nerve was identified and resected. Pathology was consistent with a glomus tumor. Since the operation, the patient has experienced complete relief after a 1.2-year follow-up.

Conclusion: Glomus tumors are treated surgically. Ultrasound may be useful in well-localized, neuropathic pain of the extremity, highlighting the importance of clinicians becoming familiar with employing this modality. While rare, a glomus tumor should be included in the differential diagnosis.

Key words: Glomus tumor, chronic neuropathic pain, case report

BACKGROUND

A glomus tumor is a rare, painful neoplasm of the neuromyoarterial glomus body, often located in highly vascular regions of the body, such as the subungual area of digits (1). Glomus tumors comprise 1% to 2% of soft tissue tumors, making the condition exceedingly rare (2). The tumors are often solitary with a purplish and firm appearance with patients presenting sensitivity to palpation, cold hypersensitivity, and pain (3). While glomus tumors have been well-recognized as inciting pain in the digits, extradigital locations have been rarely reported on, which may lead to delays in diagnosis (4). Furthermore, extradigital glomus tumors may have a different presentation of symptoms, such as a lower

level of cold intolerance (5). The differential diagnosis of a painful tumor includes neuroma, angioliipoma, neurolemmoma, and dermatofibroma.

Surgical resection commonly provides immediate pain relief without recurrence in the case of a solitary tumor. Once the diagnosis of a painful mass is made, surgical intervention is the first-line treatment. In some instances, the pain associated with a glomus tumor can mimic neuropathic pain, leading to unsuccessful attempts at clinical management (6). Spinal cord stimulation (SCS) and peripheral nerve stimulation are used in the treatment of chronic neuropathic pain refractory to other treatment modalities (7,8). While these implanted devices can attenuate pain, they are also associated

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with adverse effects, such as additional pain at the generator implant site and, in the case of SCS, spinal cord damage. Additionally, the underlying pathology is not addressed. A glomus tumor with a pattern of pain mimicking a nerve distribution can be misdiagnosed and undertreated with such devices.

In this case, we present a 53-year-old man with a 20-year history of well-localized pain in the right anterior thigh. He underwent SCS placement and peripheral nerve stimulator placement before being diagnosed with an extradigital glomus tumor by the senior author. We then discuss the etiology of glomus tumors, difficulty in the differential diagnosis, and clinical management. The patient gave informed consent.

CASE PRESENTATION

The patient is a 53-year-old man with a 20-year history of pain in the right thigh. This hypersensitivity began a year after a noneventful anterior cruciate ligament repair, which localized to a region the size 3-4 cm in diameter. The pain was localized 10 cm from his groin crease, some distance from the incision site. Further knee revisions did not alleviate pain. Assuming the pain was of neuropathic origin, he was offered SCS, but this did not achieve any relief. However, pain levels were reduced when a peripheral nerve stimulator was placed in the femoral nerve. Despite the peripheral nerve stimulator, the patient continued to report constant pain. When the patient presented to the senior author's clinic, based on the patient's long history of well-localized pain that was tender to local pressure, an ultrasonography exam was performed by the senior author at time of consultation clinic targeting the location of the patient's pain on the anterior right thigh. Ultrasonography results revealed a 0.5 cm hypoechoic lesion coursing along the branches of the anterior femoral cutaneous nerve (Fig. 1). Nerve blocks targeting the anterior femoral cutaneous nerve yielded no relief of pain upon palpation of the mass, although numbness extended down the cutaneous area of the nerve distribution. Ultrasonography-guided injection with anesthetic around the actual lesion produced 100% relief. As it was unclear whether the mass was a neuroma, nerve sheath tumor, or angioliipoma, surgical exploration was recommended.

The patient was marked and prepared for surgery (Fig. 2A). Upon surgical exploration, a purple-colored mass adjacent to the anterior femoral cutaneous nerve was noted (Fig. 2B). The 1 x 1 cm mass was external to the epineurial layer and was easily separated from the

nerve. Neurolysis of 2 branches of the anterior femoral cutaneous nerve was performed. The mass was excised (Fig. 2C), and the tissue was sent to pathology. Immunohistochemistry was positive for muscle-specific actin and caldesmon and negative for S100, AE1/AE3, chromogranin, synaptophysin, and melan-A (Fig. 3A-3C). The well-circumscribed tumor was analyzed for morphology, as well which found variable-sized nests of small, epithelioid cells surrounded by a fibrous capsule (Fig. 3D). These findings were consistent with a glomus tumor with no evidence of malignancy.

The patient was seen one week postoperatively via telemedicine and reported 95% pain relief. There has been no recurrence of symptoms at the 1.25-year follow-up. He has had his SCS battery removed and reports relief of pain in the original pattern since the operation. The patient is able to partake in strenuous activities he enjoys pain free.

DISCUSSION

We demonstrate a case of a 20-year history of neuropathic pain caused by a mass adjacent to the anterior femoral cutaneous nerve. After surgical excision, pathology was consistent with a glomus tumor, and since excision, the patient reported relief from all pain at the 1.25-year follow-up.

The glomus body consists of an arteriovenous anastomosis that acts as a thermoregulator through the regulation of blood flow (9). This neuromyoarterial receptor is present throughout the body in the stratum reticularis of the dermis. However, glomus bodies are concentrated in subungual regions and the classical presentation of a glomus tumor is a purplish mass under the nail bed (10) (Fig. 2B). When a neoplasm of glomus cells occurs in an unusual location, such as the upper thigh, diagnosis may be missed and management of the patient is suboptimal (11). In this case, a glomus tumor in proximity to the anterior femoral cutaneous nerve was discovered. As the patient elicited dysesthesias in the distribution of this nerve, his chronic pain was suspected to be of neuropathic origin.

Neuropathic pain is managed with a broad spectrum of pharmacological and nonpharmacological interventions. Two options in the latter category are SCS and peripheral nerve stimulation. The electrical lead of a SCS is placed in the epidural space, directly above the stimulated fibers. Through electrical stimulation, SCS modulates the activation of dorsal column axons and higher-order organization of nociceptive input (12).

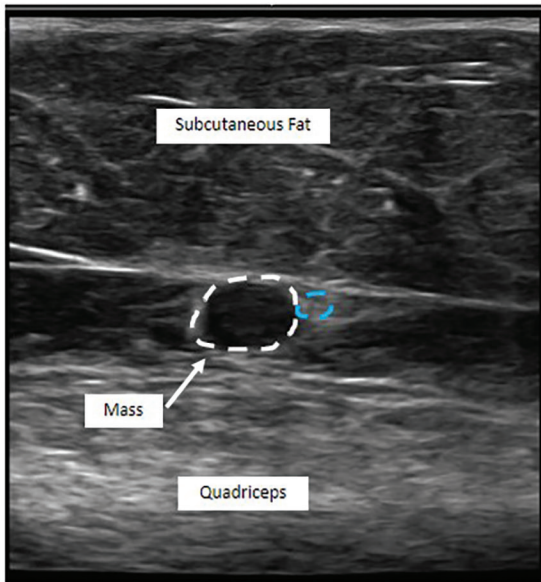


Fig. 1. Ultrasonography of the 0.5 cm noncompressible, painful hypoechoic mass located near the anterior femoral cutaneous nerve. Dotted white circle demonstrates the mass. Dotted blue circle demonstrates the anterior femoral cutaneous nerve.

Peripheral nerve stimulation has a similar mechanism of action but avoids the risk of spinal cord injury. Electrical stimulation is applied to a targeted nerve with implanted electrodes, disrupting nociceptive afferents (13,14). Our patient underwent implantation of both types of devices as a result of his refractory pain. However, neither form of stimulation resolved his symptoms and he went on to experience over 20 years of daily pain. With the etiology of his pain revealed to be a glomus tumor, it is clear that SCS and peripheral nerve stimulation were not correct management in retrospect. Only surgical excision can provide a complete cure, with low rates of recurrence being reported (15). This highlights the key principle of identifying the root cause of a patient’s pain instead of relying on nerve stimulation to ameliorate the symptoms.

The patient’s history and examination indicated that the mass may have been an angiolipoma, nerve sheath tumor, or neuroma of the anterior femoral cutaneous nerve. Only upon surgical exploration and resection was the lesion identified as a glomus tumor. Immunohistochemical studies are necessary to confirm diagnosis (16). As glomus tumors are rare, and are even rarer when presenting in an uncommon location, the cause of the patient’s pain was overlooked for decades.

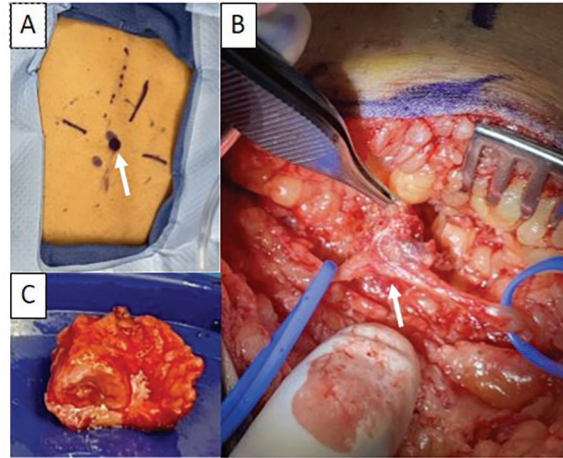


Fig. 2. Intraoperative images and photo of gross specimen. A) Preoperative markings on the anterior thigh. Ultrasound-guided assistance was used to plan the markings and the arrow designates the location of the mass. Left is proximal thigh, Right is distal thigh, Up is medial thigh, Down is lateral thigh. Picture is taken approximately upper third to mid-thigh. B) Within the subcutaneous tissue of the anterior thigh superficial to the deep fascia adjacent to the anterior femoral cutaneous nerve, was a purplish mass, designated by the arrow, continuous with the anterior femoral cutaneous nerve. C) Gross specimen of the 1 x 1 cm mass just after resection.

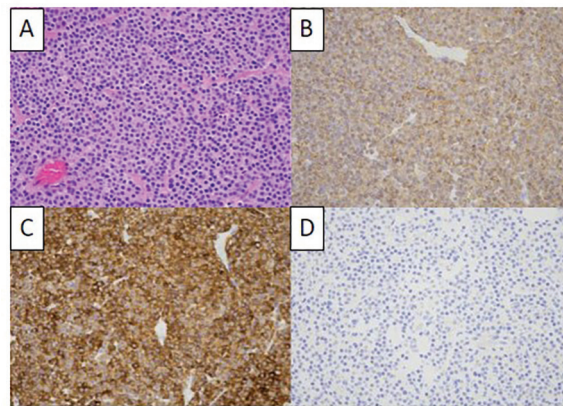


Fig. 3. Immunohistochemistry of the mass consistent with a glomus tumor. A) Hematoxylin and eosin, 20X. B) Muscle-specific actin positive, 20X. C) Caldesmon positive, 20X. D) Synaptophysin negative, 20X.

This case shows the importance of a thorough clinical workup in the case of well-localized chronic pain of unclear etiology. Targeted ultrasonography was critically important in helping make the diagnosis of this well-localized lesion. We suspected that if this mass was

identified earlier, this patient may have not undergone peripheral nerve stimulator or SCS placement though these 2 modalities are reasonable in the treatment of neuropathic pain. Our experiences suggest that ultrasonography, which is noninvasive and painless, should be taught to clinicians and made more readily accessible, especially in the specialties treating chronic pain. Overall, the pain management physician and care team should complete a meticulous evaluation for treatable causes of well-localized chronic pain.

CONCLUSIONS

A 53-year-old man with a 20-year history of persistent,

well-localized right thigh pain was seen in the clinic. Both SCS and peripheral nerve stimulation yielded no resolution of symptoms. Upon ultrasonography examination of this well-localized area of pain, a hypoechoic painful mass was identified adjacent to the anterior femoral cutaneous nerve. The mass was surgically excised, resulted in full relief of pain after 1.25 years of follow-up, and was identified as a glomus tumor. While rare, including a glomus tumor in the differential diagnosis may possibly identify similar patients promptly, such that surgical intervention can provide complete resolution of debilitating pain.

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