

Case Report

Glomus Tumor in the Femoral Nerve

Dong Sun Park, M.D., Woo Jin Choe, M.D., Young Il Chun, M.D., Chang-Taek Moon, M.D.

Department of Neurosurgery, Konkuk University Medical Center, Seoul, Korea

The glomus tumor of the peripheral nerve is one of the mesenchymal tumors originating in the epineurium, and is extremely rare. A 56-year-old man presented complaining of lancinating pain on the left thigh, which was provoked by pressure or exercise. Subsequent image study revealed a mass in the femoral nerve. Total surgical excision with the aid of intraoperative ultrasonography was performed and the pain was successfully controlled. The authors report an unusual case of a patient diagnosed with glomus tumor in peripheral nerve, with a review of the clinical features, imaging, and pathological findings.

Key Words : Glomus tumor · Peripheral nerve · Femoral nerve.

INTRODUCTION

A glomus body, or glomus apparatus, is a neuromyoarterial structure in the dermis layer of the skin, involved in body temperature regulation. It consists of an arterio-venous shunt surrounded by a capsule of connective tissue. In response to sympathetic stimuli, glomus body induces vasoconstriction in the shunt arterioles, controlling blood flow and skin temperature^{1,3,10,12}.

Glomus tumor or glomangioma is a kind of perivascular tumor and a rare benign neoplasm arising from the glomus body. The vast majority of the tumors are found under the nail, on the fingertip or in the foot^{3,8,10,11}. Rarely, they may present in other body areas such as tympanic membrane, gastrointestinal organ, glans penis, or trachea^{4,8}. Since normal human nervous system does not contain glomus bodies, glomus tumors affecting peripheral nerves are extremely rare. The glomus tumor in the peripheral nerve is known as one of the mesenchymal tumors originating in the epineurium^{9,10,13}. They are often associated with constant or episodic shooting pain that is generally unresponsive to conventional therapies. The authors report a case of glomus tumor in the femoral nerve branch with a review of literature.

CASE REPORT

A 56-year-old man presented with severe lancinating pain at left anterior thigh for nine years, one year after accidentally

bumping his left thigh to a tree. The pain worsened over time and he eventually limped for five years. He initially visited the other medical center where he was diagnosed with a mass lesion in the left thigh. Surgical exploration was performed, which was unsuccessful, and the pain persisted. The effect of direct lidocaine injection was temporary and unsatisfactory. He was referred to the neurosurgery department for evaluation and surgical management.

On physical examination, severe direct tenderness in the left anterior thigh was observed. His left thigh was rather atrophic compared to the right side. The range of motion in the extremities was full, but pain was provoked by physical irritation or by knee extension. The pain graded by the Visual Analog Scale was 10. Motor weakness or radiating pain in the leg was not distinct. Electromyography (EMG) and nerve conduction velocity were consistent with left femoral neuropathy of moderate degree and chronic partial axonotmesis state. The defect was most severe at the branch leading to the vastus lateralis.

The magnetic resonance images showed a well-defined, enhancing mass of 2.5 cm in diameter at left mid-thigh level, in between the rectus femoris, vastus lateralis, and vastus intermedius muscles (Fig. 1). The mass was formed along a muscular branch of the femoral nerve, and thus resembled a neurogenic tumor of femoral nerve branch, such as schwannoma. The initial differential diagnoses before operation were schwannoma or post-traumatic painful neuroma.

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• Address for reprints : Woo Jin Choe, M.D.

Department of Neurosurgery, Konkuk University Medical Center, 120-1 Neungdong-ro, Gwangjin-gu, Seoul 143-729, Korea
Tel : +82-2-2030-2357, Fax : +82-2-2030-7359, E-mail : s1nsman@chol.com

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The patient underwent surgical exploration and gross total resection was done under spinal anesthesia. Intraoperative ultrasonography was used to localize the mass (Fig. 2). The tan, pinkish white tumor was oval in shape with moderate vascularity, and well demarcated with the nerve tissue (Fig. 3).

The pathological findings were compatible with benign glomus tumor. The tumor cells were basophilic, round and fairly uniform. They had rich cytoplasm and centrally placed nuclei. The cellular borders were sharply defined and angulated, and dilated vessels were surrounded by the tumor cells. The immunohistochemical staining showed positive for smooth muscle antigen (SMA) and negative for S-100 protein, and the Ki-67 proliferative index was less than 1.0% (Fig. 4).

Postoperatively, the patient's symptom improved dramatically. The sharp pain was completely resolved and new neurological deficit was not observed. No evidence of recurrence or pain was noted at 24 months after resection.

DISCUSSION

A glomus tumors, a benign neoplasm arising from the glomus body, usually occurs in the subungual region, on the fingertip or in the foot³. The tumor was first described by Hoyer in 1877, while the first complete clinical description was given by Barre and Masson in 1924^{5,9}. Glomus tumors are made up of afferent arterioles, anastomotic vessel, and collecting venules accompanying modified smooth-muscle cells. Anastomoses of the afferent arterioles with collecting veins are called Sucquet-Hoyer canals and the entire complex, with abundant unmyelinated nerve fibers, is surrounded by a collagenous capsule^{5,9,12}. Hyperplasia or hamartomatous proliferation of this structures is the pathologic feature of the tumor.

Glomus tumors account for nearly 2% of the soft-tissue tumors¹². They show preference for young adults and females. Episodic pain, abrupt and lancinating in nature, is the most common symptom at presentation. This pain can be provoked by pressure or cold and often unresponsive to conventional therapies. Interruption of afferent blood flow may be effective, which is the basis of the tourniquet test^{3,5}. Multiple variant is not common, and approximately 10% of these tumors occur in multiples^{4,6,9}. An association with neurofibromatosis Type 1 has also been rarely mentioned in multiple variant glomus tumors¹².

Glomus tumor associated with apparent peripheral nerve is extremely exceptional, despite normal glomus bodies have a micro-anatomical connection with small, unmyelinated nerve fibers¹⁴. So far, only nine cases including the present case have been documented to our knowledge^{1,2,7,9,13-16}. Their clinical features are summarized in Table 1. The tumors have affected various sites such as the common peroneal nerve¹⁶, the radial nerve¹³, the sciatic nerve⁹, the tibial nerve¹⁵, a digital branch of the ulnar nerve⁷, a dermal nerve in the shoulder¹, the median nerve⁹, and the sural nerve². The present case is the first report of a glomus tumor associated with the femoral nerve branch. All le-

sions occurred in adults : six in male patients and three in female patients. The nine tumors reported ranged from 1 mm to 7 cm in size and had benign clinical and pathological nature. All the patients clinically improved with surgical excision and no evidence of recurrence was reported.

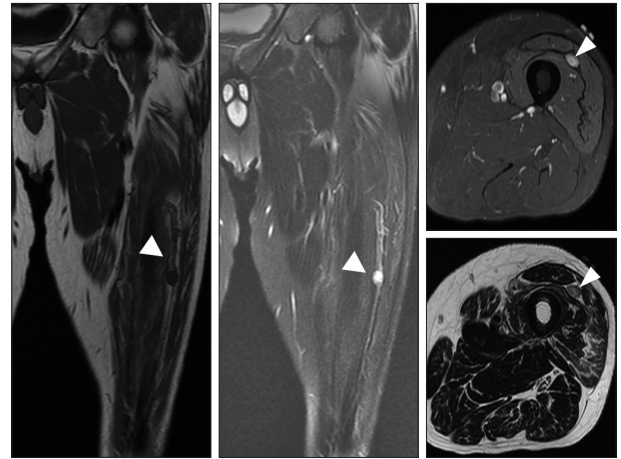


Fig. 1. MRI shows a 2.5 cm sized well-defined enhancing mass (arrow-head) in the femoral nerve among rectus femoris, vastus lateralis and vastus intermedius at mid-thigh level.

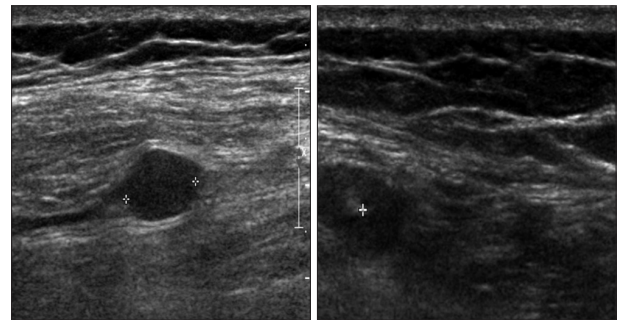


Fig. 2. Intraoperative ultrasonography (USG) showing an oval hypoechoic mass. With the aid of intraoperative USG, the deep seated tumor could be located exactly.

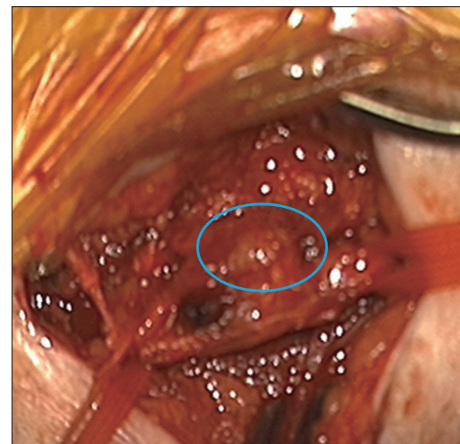


Fig. 3. Operative view of the glomus tumor (inside the circle) : oval in shape with moderate vascularity, and well demarcated with the nerve. The tumor was removed without difficulty using microscopic technique. The tumor was connected to a vessel.

The rare occurrence of glomus tumors in nerves where normal glomus bodies have not been found can be approached by a number of theories⁷⁾. First, the tumor might infiltrate from an extraneural tissue such as accompanying vessels by direct extension. Second hypothesis is that tumors are developed from a possible normal intraneural glomus body or ectopic glomus cells, but such histological findings have never been described in nerves so far. The third possibility is that the tumor developed through differentiation from unspecialized perivascular wall, or pericytes in

particular. The cells of glomus tumors are similar to pericytes in morphological features. Pericytes are also modified smooth-muscle cells, widely distributed around small-sized vessels like vasa nervorum. Also, smooth muscle cell and glomus tumor cell show a resemblance in immunophenotype study (SMA+, collagen IV+, S100-, and epithelial membrane antigen; EMA-). The differentiation of pericytes into tumor cells can be a potential explanation for the origin of glomus tumors where glomus body do not exist in general^{3,5-8,10,12)}. In this case, we are in favor of first and

the third hypotheses, because the tumor was obviously connected to a small vessel and the tumor was discrete to the associated neural tissue. These theories need further investigation and clarification.

When a patient complains severe lancinating pain aggravated by exercise or physical irritation, and especially has history of trauma as is this case, surgeons easily consider the diagnosis of painful neuroma. However, neoplastic conditions can show similar manifestations. When presenting at an uncommon site such as a nerve, other entities including a schwannoma, a meningioma, an epithelioid leiomyosarcoma, a metastatic melanoma, a carcinoma or a glomus tumor should be considered as differential diagnosis¹⁰⁾. Mere medication and loose observation may be catastrophic, and image study and surgical confirmation must be considered at an

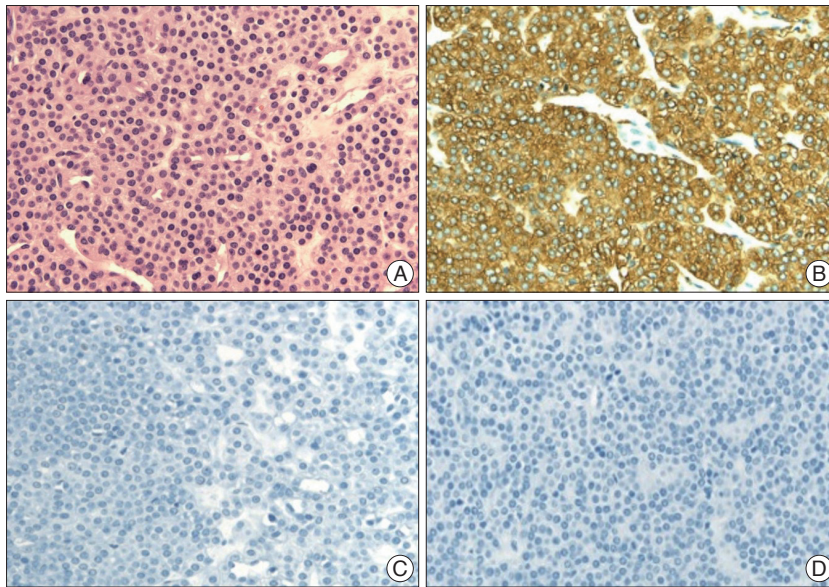


Fig. 4. A : H&E, original magnification $\times 200$. Cells are round, uniform and basophilic. Rich cytoplasm and centrally placed nucleus are observed. Dilated vessels are surrounded by tumor cells. B, C and D : Original magnification $\times 200$. Tumor cells are positive for smooth muscle antigen; SMA (B), negative for S-100 protein (C), and showing low Ki-67 proliferative index less than 1.0% (D). SMA : smooth muscle antigen.

Table 1. Summary of clinical features of glomus tumors involving peripheral nerves

Authors & year	Sex/age	Nerve involved	Symptoms	Size	Follow-up results
Wing & Levine ⁶⁾ , 1962	M/37	Common peroneal n.	Radiating pain diffuse muscle atrophy	1 cm	NA
Kline et al. ⁷⁾ , 1990	M/46	Digital branch of ulnar nerve n.	Numbness & tingling of fingers point tenderness; cold insensitivity	"Pea size"	Complete resolution of pain with residual numbness
Tropet et al. ¹⁵⁾ , 1991	F/16	Tibial n.	Posterior thigh pain & tenderness	5 cm	Partial recovery of motor function after n. graft
Smith et al. ¹³⁾ , 1992	F/62	Radial n.	Upper arm & finger pain	2.5 cm	Immediate resolution of pain t recurrent symptoms (-)
Calonje and Fletcher ¹⁾ , 1995	F/67	Dermal n. in shoulder	Shoulder pain	1 cm	No evidence of recurrence in 14 yrs
Donato et al. ²⁾ , 2006	M/56	Sural n.	Distal sensorimotor neuropathy	400 μ m	NA
Scheithauer et al. ⁹⁾ , 2008	M/32	Sciatic n.	Lower extremity pain hypesthesia & weakness	7 cm	Improved symptoms recurrence (-)
Scheithauer et al. ⁹⁾ , 2008	M/31	Median n.	Hand weakness and forearm atrophy, burning pain at thumb	6 cm	Resolution of pain in the hand and elbow
Present case	M/56	Femoral n.	Anterior thigh pain	2.5 cm	Complete resolution of pain

n. : nerve, NA : not available

early stage. Operative excision has benefits in both diagnosis and curative treatment of the tumor affecting peripheral nerves.

Although glomus tumors are closely associated with the functioning peripheral nerves, complete surgical excision can be achieved without difficulty thorough microsurgical techniques. According to the literatures along with our experience on a glomus tumor in the peripheral nerve, a nerve-sparing surgical excision is the curative treatment of choice. When a tumor is deeply seated between muscles and is without a palpable mass, it can be hard to locate the exact lesion site. At first surgery in the present case, surgical exploration had failed because the surgeon could not locate the tumor by palpation. The authors were able to remove the tumor successfully with the aid of intraoperative ultrasonography. Intraoperative EMG monitoring may also be helpful, but it was omitted in the present case because the operation was performed under spinal anesthesia. The debilitating lancinating pain was resolved and no recurrence was seen after total resection of the tumor as were the reported cases.

CONCLUSION

A glomus tumor can be encountered at an uncommon site, such as a peripheral nerve. Severe shooting pain and pressure tenderness are characteristic clinical features. A nerve-sparing surgical excision is the curative choice of the treatment.

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