KJP The Korean Journal of Pain
| Case Report |
Korean J Pain 2013 July; Vol. 26, No. 3: 295-298
pISSN 2005-9159 eISSN 2093-0569
http://dx.doi.org/10.3344/kjp.2013.26.3.295

Concomitant Glomus Tumor with CRPS in the Hand

[1]Department of Anesthesiology and Pain Medicine,
[2]Anesthesia and Pain Research Institute, Yonsei University Health System, Seoul, Korea

Hyeong Jun Jeong, MD[1,2], Chan Mi Kim, MD[1], Duck Mi Yoon, MD[1,2], and Kyung Bong Yoon, MD[1,2]

Glomus tumors are benign tumors that account for 1% to 5% of all soft tissue tumors of the hand and are characterized by a triad of sensitivity to cold, localized tenderness and severe paroxysmal pain. Paroxysmal pain is a symptom common not only in glomus tumors but also in CRPS, and the hand is one of the commonly affected sites in patients with both glomus tumors and CRPS. Therefore, it is not easy to clinically diagnose glomus tumors superimposed on already affected region of CRPS patients. We report a case of glomus tumor concomitantly originating with CRPS at the hand. (Korean J Pain 2013; 26: 295-298)

Key Words:
complex regional pain syndrome, glomus tumor.

A glomus tumor is a rare form of a soft tissue tumor which occurs on the glomus body, and normally develops on the hands causing severe pain. The disease is characterized by excruciating pain, localized tenderness and cold sensitivity around the tumor and is usually a solitary lesion. It is a benign tumor that mainly occurs underneath the finger and toe nails. On the other hand, complex regional pain syndrome (CRPS) is a rare disease associated with pains in the arms and legs. The authors report a case of a glomus tumor developed on the lesion affected by CRPS, which led to a difficult diagnosis.

CASE REPORT

A 41-year-old male patient received conservative treatment on his left wrist and hand for 3 months due to pain developed by a motor vehicular accident 6 months prior to his visit. Since he did not see any improvement from the conservative treatment, he had a wrist arthroscopy performed at a private clinic. The pain persisted even after surgery, so alternative wrist arthroscopy was performed at the orthopedics department of Severance Hospital. Afterwards, he was referred to a pain clinic with a possible diagnosis of CRPS.

At the time of admission to the hospital, the patient had spontaneous pain in his left hand, wrist and forearm with a numerical rating scale (NRS) of 9 and also paroxysmal pain, intermittent heat sensation, sweating and shiny skin with a numerical rating scale of 10. From the clinical assessment, the skin color change (redness) on the left hand, edema, allodynia, hyperalgesia, and hyperhidrosis were observed. Additionally, motion was limited in the left

Received December 21, 2012. Revised January 21, 2013. Accepted January 23, 2013.
Correspondence to: Kyung Bong Yoon, MD
Department of Anesthesiology and Pain Medicine, Anesthesia and Pain Research Institute, Yonsei University Health System, 50 Yonsei-ro, Seodaemun-gu, Seoul 120-752, Korea
Tel: +82-2-2228-2416, Fax: +82-2-2228-7897, E-mail: kbyoon@yuhs.ac

wrist and there was muscle atrophy in the left hand and forearm. There was no particularly abnormal finding from the blood test and digital infrared thermal imaging. However, the dynamic blood flow images and blood pool images in the 3-phase bone scan found an increase in radio-uptake in the left upper extremity. There was an increase in bone uptake in the left wrist and hand joint on the delayed phase images (Fig. 1).

Patient's symptoms and signs were matched to CRPS diagnostic criteria (Budapest diagnostic criteria) [1], and treatment was initiated for CRPS. After stellate ganglion block (SGB) with 10 ml of 1% lidocaine, the pain was reduced temporarily from a NRS of 9 to 5 and after intravenous regional sympathetic block (IVRSB) with 10 µg of alprostadil and 30 ml of 0.2% lidocaine, the NRS was reduced from 9 to 7 for a couple of days. On top of the SGB and IVRSB, 75 mg of pregabalin and 1 combination tablet of tramadol/acetaminophen (ultracet®) was administered twice a day. After treatment for 3 months, the spontaneous pain in the left forearm and wrist was reduced from a NRS of 9 to a NRS of 5, but there was no improvement in the paroxysmal pain with a NRS of 10 in the little finger region. Severe tenderness of the left 5th fingernail was observed on the physical examination, and a positive result was reported for a cold sensitivity test. Magnetic resonance imaging (MRI) was conducted with the possibility of a glomus tumor and a 3 mm sized enhanced ovoid shaped mass was observed at the radial side of the subungual region in his left 5th finger (Fig. 2).

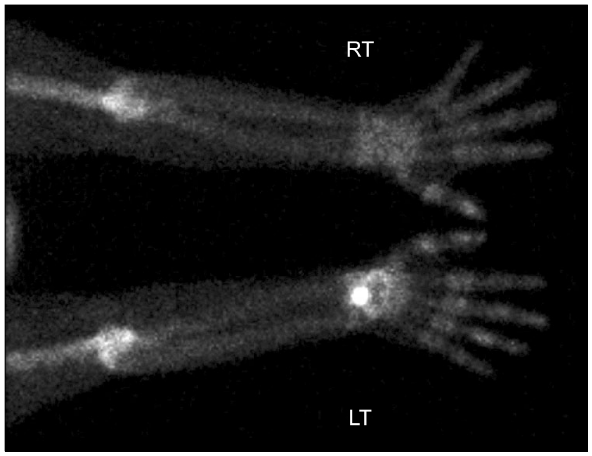

Fig. 1. The delayed bone image of 3-phase bone scan showing increased bone uptake in the left wrist and hand joints.

Surgical excision was carried out and glomus tumor was confirmed through the pathology result. Twenty µg/ml of fentanyl was administered intravenously at a rate of 2 ml/hour for 2 days after the surgery, while Ultracet® and immediate release oxycodone were administered until patient discharge after the surgery. In addition, daily SGB was conducted on the day before the surgery until the discharge date. The paroxysmal excruciating pain with a NRS of 10 in the left 5th finger disappeared after surgery. However, throbbing pain with a NRS of 5 on the left 4th and 5th finger continued even after the surgery and CRPS symptoms such as the left wrist, forearm, elbow, shoulder pain (NRS of 5-6) and skin color change are still present.

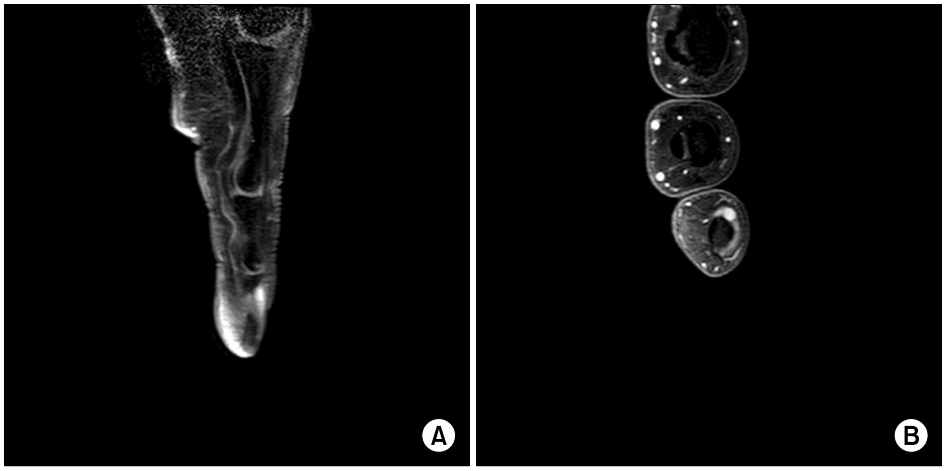

Fig. 2. Contrast enhanced T1 weighted images with fat suppression show 3 mm sized enhanced ovoid shaped mass at the radial side subungual region of the left 5th finger. (A) Saggital image, (B) Axial image

DISCUSSION

Glomus tumor is one of the rarest benign tumors accounting for 1–5% of soft tissue tumors prevalent on the hand [2]. This disease involves a tumor that occurs on the glomus body which is a cutaneous structure associated with regulating body temperature through arteriovenous shunting, and it can occur anywhere in the body where a glomus body exists, but it is more prevalent in the subungual region where a relatively large number of glomus bodies exist or in deep dermis such as the palms and the wrists. 75% of glomus tumors occur on the hands, and 75–90% of those exist on subungual region [3]. It normally exists as a solitary lesion, and it has been reported that multiple lesions in adult patients are rare. A subcutaneous nodule is frequently seen which presents as red, purple or blue along the depth of the lesion [4], and normally the size of a lesion is less than 5 mm [2], but a lesion bigger than 3 cm in size has been reported as well [4].

Most of all, paroxysmal pain, pin point tenderness and cold sensitivity are the characteristics of glomus tumor. It can be easily diagnosed though a few clinical tests if the lesion occurs in a typical region. The *Love's pin test* is a test that can be used when the tumor is suspected at the skin or nail plate region: an excruciating pain is provoked when the region is compressed with a pinhead or paperclip. The *cold sensitivity test* is an assessment to find out whether the pain gets worse when the lesion is immersed in cold water or exposed to cold environment. The *Hildreth's test* is a modified version of the Love's pin test. It assesses whether the pain and tenderness decreases in Love's pin test after exsanguinating blood using a tourniquet [5]. It is more suggestive of the diagnosis when decreased.

Subungual glomus tumors usually present as normal findings on plain radiographs. In some cases however, soft tissue thickening or bone erosion can be found on lateral radiographs of the affected finger when compared with the normal counterpart. This can be done precisely by linearly aligning the fingers so that the affected side and the normal side oppositely faces each other. [6]. Glomus tumors show increased signal intensity on T2 weighted images during high resolution MRI, thus being useful in identifying the tumor preoperatively and in evaluating its pathological process even in extradigital glomus tumors. However, it should be kept in mind that the possibility of a false negative result may arise when the tumor size is 2–3 mm [7].

Several treatment modalities exist such as sclerotherapy, laser therapy, and surgical excision. Choice of most suitable treatment can be made according to the clinical presentation [4], but most cases are treated by surgical excision [5]. The recurrence rate is reported as 1–18% and incomplete extirpation of the tumor is to a certain extent attributed to these recurrences [5]. Nail deformity may arise as a complication after surgery depending on the location of the lesion [5]. Most glomus tumors are benign in nature, and only about 1% of them are malignant [5]. The metastasis rate is greater than 25% when the tumor size is larger than 2 cm, a lesion that is positioned deep in the tissue and meets particular histological criteria [4].

On the other hand, CRPS usually occurs after trauma, and is a disease characterized by sensory, autonomic, motor, skin and bone changes with severe pain [8]. The symptoms are often limited to the limbs, and 60% of the cases occur on the arms and 40% of the cases occur on the legs [9]. The triggering events of CRPS are reported as follows: fracture (45%), sprain (18%), elective surgery (12%), etc. [9]. The prevalence rate of CRPS is not clearly known, but some studies have reported that 5.5–26.2 patients per 100,000 people develop CRPS every year [10,11].

Paroxysmal pain, which is a characteristic symptom of glomus tumors, is also common in CRPS. In our case report, the paroxysmal pain in the left 5th finger appeared after the trauma incident. Therefore it was the temporal confounding cue that led the authors to believe this symptom as one of the many constellations of CRPS and treatment was instituted accordingly for CRPS without any improvement.

It is quite common to encounter no treatment response in CRPS patients. However, in this case report, the lesions on the other parts such as the left forearm and wrist showed improvement after treatment with SGB and IVRSB etc., but the left 5th finger particularly did not respond to the treatment. This prompted us to seek other causative factors for the pain other than CRPS. Temporary severe excruciating pain was elicited at the left 5th finger even with a slight contact. The pin point tenderness and cold sensitivity were confirmed thereafter, and MRI was carried out to confirm a concomitant glomus tumor. It took us an overall three months of time to finally arrive at the diagnosis of CRPS and comorbid glomus tumor since the patient's first visit to the outpatient clinic.

If the region for surgery coincides with the CRPS lesion as in this case, the management of postsurgical pain can be very difficult because surgery itself may trigger exacerbation of the preexisting CRPS symptoms. In order to prevent this, the surgery should be scheduled when the CRPS symptoms are well managed, the duration of ischemia time caused by the operation should be minimized, a tourniquet should be used, and the surgery should be conducted with the least invasive method [12]. In addition, the administration of calcitonin before and after the surgery, early initiation of functional exercise after the surgery and the active management of the pain are known to be helpful in preventing exacerbation of the CRPS symptoms, and recently, there was a study reporting that the administration of vitamin C assists in the prevention of CRPS [12].

Both glomus tumors and CRPS usually involve the hand and cause severe pain and since both are rare diseases, the prevalence is low. If a glomus tumor coexists with pain due to CRPS, the diagnosis of glomus tumor can be overlooked due to the similarities of clinical manifestations with CRPS, especially the presentation of pain. The following characteristics can be useful in differential diagnosis: unlike CRPS, there is no spontaneous pain in a glomus tumor, and severe pain appears only with the presence of pressure, and the presence of pin point tenderness and cold sensitivity. The other possible differential diagnoses should always be considered when treating CRPS patients.

REFERENCES

1. Harden RN, Bruehl S, Stanton-Hicks M, Wilson PR. Proposed new diagnostic criteria for complex regional pain syndrome. Pain Med 2007; 8: 326-31.
2. McDermott EM, Weiss AP. Glomus tumors. J Hand Surg Am 2006; 31: 1397-400.
3. Carroll RE, Berman AT. Glomus tumors of the hand: review of the literature and report on twenty-eight cases. J Bone Joint Surg Am 1972; 54: 691-703.
4. Gombos Z, Zhang PJ. Glomus tumor. Arch Pathol Lab Med 2008; 132: 1448-52.
5. Nazerani S, Motamedi MH, Keramati MR. Diagnosis and management of glomus tumors of the hand. Tech Hand Up Extrem Surg 2010; 14: 8-13.
6. Vandenberghe L, De Smet L. Subungual glomus tumours: a technical tip towards diagnosis on plain radiographs. Acta Orthop Belg 2010; 76: 396-7.
7. Glazebrook KN, Laundre BJ, Schiefer TK, Inwards CY. Imaging features of glomus tumors. Skeletal Radiol 2011; 40: 855-62.
8. Goebel A. Complex regional pain syndrome in adults. Rheumatology (Oxford) 2011; 50: 1739-50.
9. de Mos M, de Bruijn AG, Huygen FJ, Dieleman JP, Stricker BH, Sturkenboom MC. The incidence of complex regional pain syndrome: a population-based study. Pain 2007; 129: 12-20.
10. Drummond PD. Sensory disturbances in complex regional pain syndrome: clinical observations, autonomic interactions, and possible mechanisms. Pain Med 2010; 11: 1257-66.
11. Sethna NF, Meier PM, Zurakowski D, Berde CB. Cutaneous sensory abnormalities in children and adolescents with complex regional pain syndromes. Pain 2007; 131: 153-61.
12. Asaad B, Glass P. Perioperative management for patients with complex regional pain syndrome. Pain Manag 2012; 2: 561-7.