SHORT COMMUNICATION

Nagoya J. Med. Sci. **70**. 127 ~ 133, 2008

INTRAOSSEOUS GLOMUS TUMOR OF THE ULNA: A CASE REPORT WITH RADIOGRAPHIC FINDINGS AND A REVIEW OF THE LITERATURE

HIROSHI URAKAWA[1], HIROATSU NAKASHIMA[2], YOSHIHISA YAMADA[1], MIKITO TSUSHIMA[1], TAKEHIRO OHTA[1] and TOMOKO NISHIO[3]

[1] *Department of Orthopedic Surgery, Nagoya Memorial Hospital*
[2] *Department of Orthopedic Surgery, Aichi Cancer Center Aichi Hospital*
[3] *Department of Pathology, Nagoya Memorial Hospital*

ABSTRACT

Intraosseous glomus tumors of bone are extremely rare. We report a case of an intraosseous glomus tumor of the ulna. The patient was a 25-year-old woman who had a three-month history of a palpable mass in her right forearm with spontaneous pain. Radiographs showed cortical hypertrophy and a shell-like bone formation surrounding the small osteolytic lesion within the cortex of the ulna diaphysis. The differential diagnosis included benign bone-forming tumors, such as osteoid osteoma. The patient was treated with an *en-bloc* resection and filling with beta-TCP. Up to one year after the operation there has been no evidence of recurrence.

Key Words: Intraosseous glomus tumor, Ulna, Radiograph

INTRODUCTION

Glomus tumors are rare, with an estimated incidence of 1.6% among the 500 consecutive soft tissue tumors reported by the Mayo Clinic.[1)] Glomus tumors are benign lesions derived from neuromyoarterial glomus cells. They were originally considered a form of angiosarcoma until Masson published his classic paper on the subject in 1924.[2)]

The glomus body is involved in thermal regulation, and is located in the stratum reticularis of the dermis. It is most commonly found in the subungual region, the lateral areas of the digits, and the palm. The most common sites of glomus tumors reflect the normal glomus distribution throughout the body. The vast majority of glomus tumors occur in the distal extremities, particularly the subungual region and the forearm.

Osseous abnormalities have been reported in soft tissue glomus tumors, but only 20 cases of intraosseous glomus tumor have so far been reported.[3-20)] Most of them involve the distal phalanges of the fingers or thumb, and only two cases have been previously reported in a long bone. Here, we report a rare case of a primary intraosseous glomus tumor of the ulna, and discuss our radiographic findings. We obtained informed consent from the patient to publish this case report.

Corresponding author: Hiroshi Urakawa, M.D.
Department of Orthopedic Surgery, Nagoya Memorial Hospital,
4-305 Hirabari, Tenpaku-ku, Nagoya 468-8520, Japan
Phone: +81-52-804-1111, Fax: +81-52-806-6257, E-mail: urakawa777@hotmail.com

CASE REPORT

The patient was a 25-year-old woman who visited an orthopedic clinic with the chief complaint of a solid mass in her right proximal forearm that she noticed one month before. There was no previous known history of injury or infection at that location. A bone tumor of the ulna of the right forearm was demonstrated with X-rays and Magnetic Resonance Imaging (MRI). However, the woman did not visit the clinic after the MRI examination, preferring to monitor the course herself. Having experienced occasional right elbow pain, she visited the clinic 3 months after her first examination. She was then referred to our hospital.

On physical examination, there was a solid mass with a point of mild tenderness on the medial side of the proximal forearm. There was no sign of infection or pigmentation, nor were there any abnormal laboratory findings.

Roentgenographic examination of the right elbow revealed an osteolytic area in the proximal ulna. The lesion measured 1.0 cm in length. Its location was within the thickened medial cortex. There was a shell-like bone formation outside of the osteolytic area (Fig. 1). CT of the right elbow revealed a low-density area of approximately 1 cm within the thickened cortex of the proximal ulna, and there were light spotted calcifications in the low-density area; that area continued to the vascular groove (Fig. 2). MRI showed the lesion to be homogeneous, with a slightly high signal intensity on T1-weighted imaging, and a high signal intensity on $T2^{*}$-weighted images. The lesion did not continue to the vascular groove on MRI, and no tumor was seen outside the bone (Fig. 3).

The differential diagnosis at that time included intraosseous hemangioma, osteoid osteoma, osteoblastoma, osteofibrous dysplasia, adamantinoma, periosteal chondroma, and abscess, all of which were suggested by the presence of spotted calcifications and the fact that no tumor was observed outside the bone.

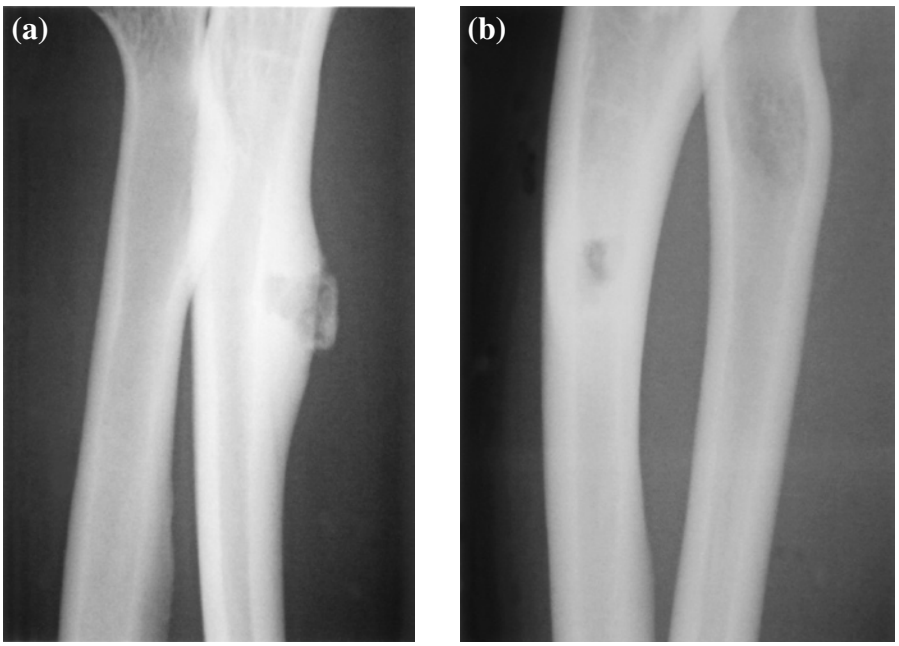

Fig. 1 Anteroposterior (a) and lateral (b) radiographs of the right elbow showed an osteolytic area in the proximal ulna, and there was shell-like bone formation outside the osteolytic area.

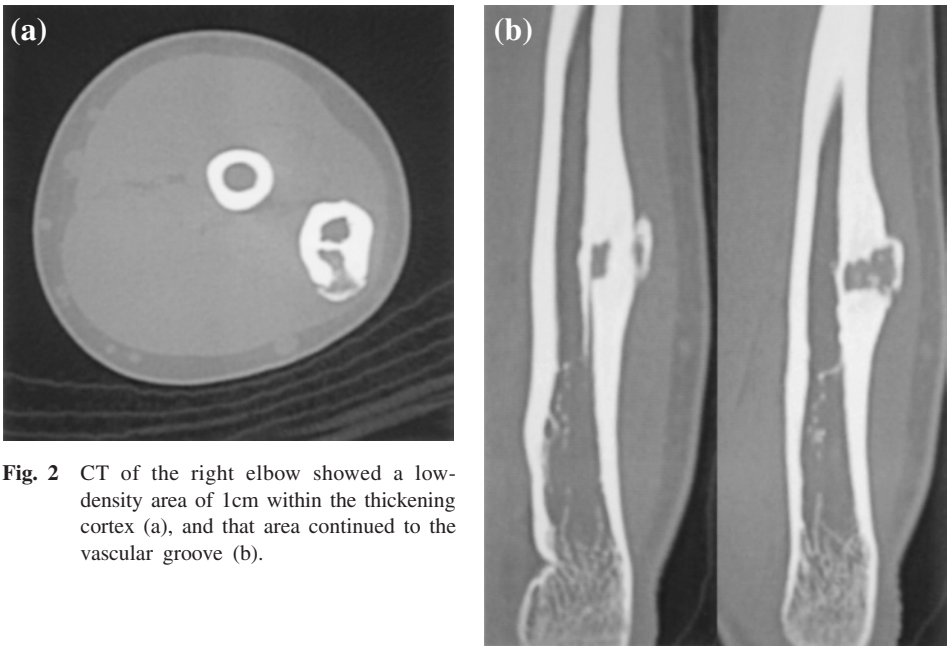

Fig. 2 CT of the right elbow showed a low-density area of 1cm within the thickening cortex (a), and that area continued to the vascular groove (b).

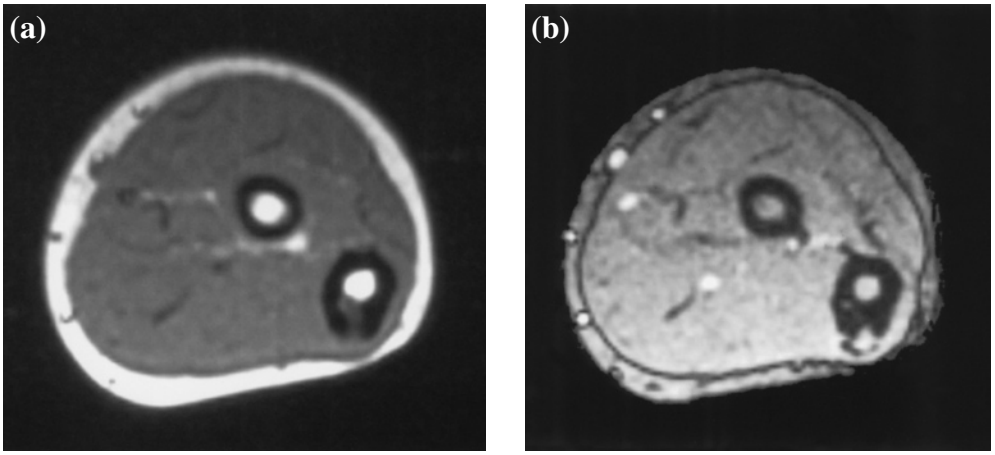

Fig. 3 MRI showed the lesion to be homogeneous with a slightly high signal intensity on T1-weighted imaging (a), and a clearly high signal intensity on T2*-weighted imaging (b).

The lesion was completely resected *en bloc* under an axillary block two months after her first visit. The operative approach between the flexor and extensor exposed the swollen bone. The resected area was reached via a bone marrow cavity using surgairtome, and was filled with beta-tricalcium phosphate (β-TCP). The tumor was located within the cortical bone, and was orange in color. No extraosseous tumor was visible to the naked eye. On pathological examination, most of the focus increased as the bone was replaced and the original trabecula was destroyed. The tumor partially destroyed cortical bone, and extended outside the periosteum. Irregular calcium

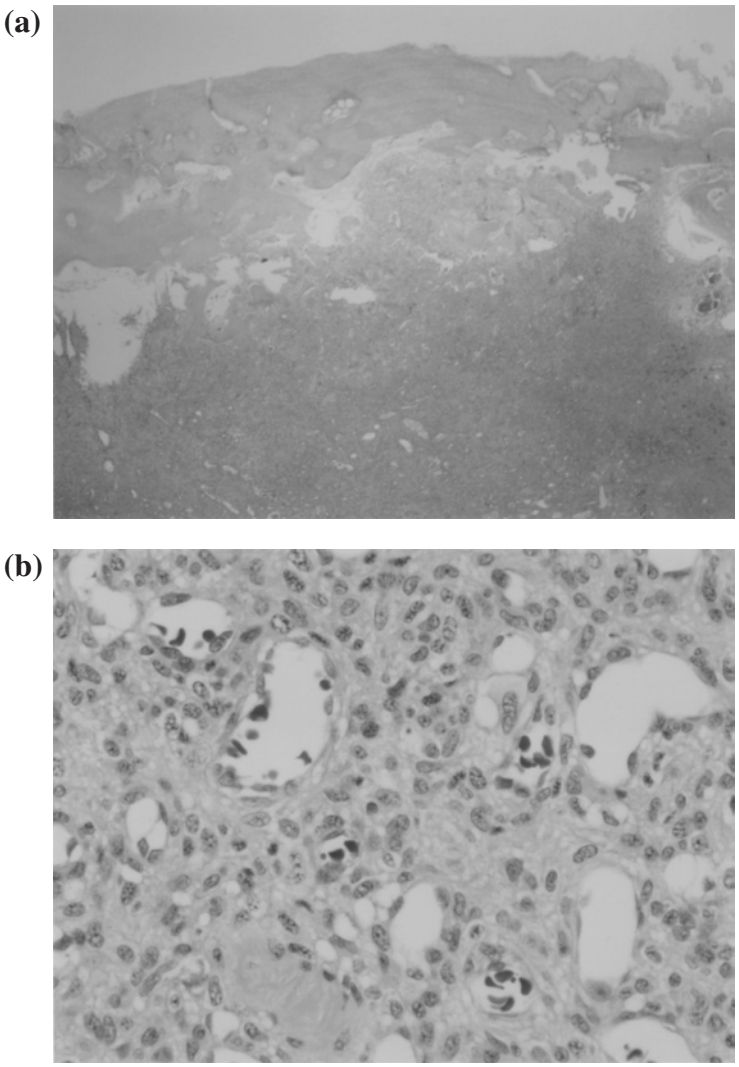

Fig. 4 On pathological examination, the tumor partially destroyed cortical bone (a). Under high-power microscopic examination, the cells appeared round to ovoid in shape and grew as surrounding small vessels (b).

deposits were found in the lesion of the destroyed bone. Under high-power microscopic examination, the cells were appeared round to ovoid in shape, and they grew as surrounding small vessels (Fig. 4). The cytoplasm was eosinophilic, and the cell boundaries were not very clear under hematoxylin-eosin staining. There was no prominent nuclear atypia, and mitotic activity was low. The immunohistochemical stains were positive for vimentin, smooth muscle actin, type 4 collagen, and partly-positive for desmin. The tumor was negative for Factor VIII, CD34, and S-100. MIB-1 positivity was low. These histological findings led to a pathological diagnosis of glomangioma, which is a variant of glomus tumors.

Simple roentgenography shows that the patient currently has no symptoms and has been completely disease-free without evidence of local recurrence for 1 year since resection of the tumor.

DISCUSSION

Glomus tumors are, with the rare exception of glomangiosarcomas, benign neoplasms derived from the neuromyoarterial plexus. Clinically, this tumor is seen as a small, red-blue, superficial nodule with symptoms of paroxysms of radiation pain due to changes in temperature or pressure. The classic clinical triad of pain, point of tenderness, and cold sensitivity is present in approximately 30% of patients.[21] A striking female predisposition (3:1) is observed in subungual lesions, although overall there is no gender-based predisposition. Adults between 20 and 40 years of age are usually affected.[22]

Glomus tumors develop in sites where the normal glomus body may exist. Such tumors are usually located around the terminal phalanx of the hand, although other common sites include the wrist, forearm, and foot. Unusual sites affected include the chest wall, stomach, colon, nerve, face, trachea, and possibly the mediastinum.[23] Extrinsic erosion of bone, often with a sclerotic margin, can be seen in 22% to 60% of nail bed cases.[24] However, an intraosseous glomus tumor of bone is extremely rare, and, to our knowledge, only 20 such cases have been reported in the English literature.[3-20] There are 12 cases of phalanges, 5 of spine and sacrum, one of ankle, and only 2 of long bone, one ulna and one fibula (Table). There is a female predisposition toward having the tumors (5 males and 13 females), and the average age is 39.4 years (22–68 years).

Rozmaryn *et al.* reported an intraosseous glomus tumor of the ulna.[14] Radiographs of the right elbow showed an area of bone destruction in the anteromedial region of the proximal ulna. It was located within the medial cortex, and there was no sclerotic border. In a case reported by

Table 20 cases of intraosseous glomus tumor reported in the English literature

Authors	Date	Age (yrs)	Sex	Site	Duration of symptoms	Treatment	Recurrence
Lattes *et al.*[9]	1948	28	F	Thumb	4 years	Curettage and bone graft	(–)
Lehman *et al.*[10]	1949	48	F	5th finger	6 years	Curettage	(–)
Lehman *et al.*[10]	1949	36	F	Thumb	14 months	Excision	(–)
Mackenzie[11]	1962	30	F	5th finger	2 years	Amputation	(–)
Mackenzie[11]	1962	61	F	Thumb	6 years	Excision	(–)
Siegel[16]	1967	24	M	2nd finger	10 months	Curettage	Unknown
Sugiura[18]	1976	33	F	5th finger	2 years	Curettage	(–)
Chan[6]	1981	42	F	4th finger	20 years	Amputation	Unknown
Bjorkengren *et al.*[5]	1986	68	M	5th finger	5 years	Curettage and bone graft	(–)
Rozmaryn *et al.*[14]	1987	24	F	Ulna	2 years	Curettage	(–)
Kobayashi *et al.*[8]	1990	22	F	Sacrum	1 year	Cutettage	(–)
Bessho *et al.*[4]	1991	49	M	Spine	Unknown	Decompression and fusion	(–)
Simmons *et al.*[17]	1992	30	M	Thumb	2 years	Amputation	(–)
Johnson *et al.*[7]	1993	28	M	Thumb	1.5 years	Amputation	(–)
Robinson *et al.*[13]	1996	45	F	Spine	4 years	Curettage	(–)
Bahk *et al.*[3]	2000	34	F	Fibula	1 month	Resection	(–)
Settakorn *et al.*[15]	2001	53	F	2nd finger	3 years	Resection	(–)
Payer *et al.*[12]	2002	55	F	Spine	Unknown	Decompression and fusion	(–)
Massari *et al.*[19]	2006	38	F	Calcaneus	2 years	Resection	(–)
Bambakidis *et al.*[20]	2007	44	M	Spine	1 year	Resection/decompression/ fusion	(–)
Present case	2008	25	F	Ulna	1 month	Resection and filling with β-TCP	(–)

β-TCP: β-Tricalcium Phosphate (–): no recurrence

Bahk *et al.* involving an intraosseous glomus tumor of the fibula, radiographs of the right leg revealed an ovoid, eccentric, expansive lytic lesion of the midshaft of the fibula with an outer complete, thin shell of bone.[3)]

In our case, the radiographs of the right elbow revealed an osteolytic area in the proximal bone shaft of the ulna, with the lesion measuring 1.0 cm in length. It was located within the medial cortex thickening. There was a shell-like bone formation outside of the osteolytic area. Our case is similar to that of Rozmaryn *et al.* in that the radiolucent area existed in the cortical bone on X-ray, but there was no thickening of the cortex in their case. All 3 cases of intraosseous glomus tumor of the long bone exhibited similarities, in showing on X-ray a radiolucent area with a thin cortical shell, although there was some variation in the cortex or in the thickening or ballooning of the shaft.

In the case of Rozmaryn *et al.*, the glomus tumor originating in the soft tissue presumably eroded through the opening in the bony cortex. However, Bahk's patient showed no evidence of this phenomenon. They believed that most, if not all, reported cases of bone lesions are not primary tumors of the bone but soft tissue glomus tumors that erode into the adjacent bone, followed presumably by a glomus tumor arising within the bone, either from normal glomera that occasionally exist in the terminal phalanges, or from pericytes in the blood vessel walls.

There was no evidence of an opening of the cortex during our operation nor any microscopic evidence of continuity between the tumor and the vessel. However, CT did demonstrate the continuity of the tumor lesion to the vascular groove. We considered that this intraosseous glomus tumor might have arisen from pericytes in the blood vessel walls, given the CT findings.

REFERENCES

1) Shugart RR, Soule EH, Johnson EW Jr. Glomus tumor. *Surg Gynecol Obstet,* 1963; 117:334–340.
2) Masson P. Le glomus neuromyo-artériel des régions tactiles et ses tumeurs. *Lyon Chir,* 1924; 21: 257–280.
3) Bahk WJ, Mirra JM, Anders KH. Intraosseous glomus tumor of the fibula. *Skeletal Radiol,* 2000; 29: 708–712.
4) Bessho Y, Kataoka O, Sho T, Kitazawa S, Okada S. Intraosseous glomus tumor in the upper thoracic spine complicating compression myelopathy: a case report. *Spine,* 1991; 16: 988–990.
5) Bjorkengren AG, Resnick D, Haghighi P, Sartoris DJ. Intraosseous glomus tumor: report of a case and review of the literature. *AJR,* 1986; 147: 739–741.
6) Chan CW. Intraosseous glomus tumor: case report. *J Hand Surg,* 1981; 6A: 368–369.
7) Johnson DL, Kuschner SH, Lane CS. Intraosseous glomus tumor of the phalanx: a case report. *J Hand Surg,* 1993; 18A: 1026–1028.
8) Kobayashi Y, Kawaguchi T, Imoto K, Yamamoto T. Intraosseous glomus tumor in the sacrum: a case report. *Acta Pathol Jpn,* 1990; 40: 856–859.
9) Lattes R, Bull DC. A case of glomus tumor with primary involvement of bone. *Ann Surg,* 1948; 127: 187–191.
10) Lehman W, Kraissl C. Glomus tumor within bone. *Surgery,* 1949; 25: 118–121.
11) Mackenzie DH. Intraosseous glomus tumours. Report of two cases. *J Bone Joint Surg,* 1962; 44B: 648–651.
12) Payer M, Grob D, Benini A, Varga S, Hodler J, Martin JB. Intraosseous glomus tumor of the thoracic spine. Case illustration. *J Neurosurg,* 2002; 96: 137.
13) Robinson JC, Kilpatrick SE, Kelly DL Jr. Intraosseous glomus tumor of the spine: case report and review of the literature. *J Neurosurg,* 1996; 85: 344–347.
14) Rozmaryn LM, Sadler AH, Dorfman HD. Intraosseous glomus tumor in the ulna: a case report. *Clin Orthop,* 1987; 220: 126–129.
15) Settakorn J, Chalidapong P, Rangdaeng S, Arpornchayanon O, Chaiwun B. Primary intraosseous glomus tumor: a case report. *J Med Assoc Thai,* 2001; 84: 1641–1645.
16) Siegel MW. Intraosseous glomus tumor. a case report. *Am J Orthop,* 1967; 9: 68–69.

17) Simmons TJ, Bassler TJ, Schwinn CP, Forrester DM. Case report 749: primary glomus tumor of bone. *Skeletal Radiol,* 1992; 21: 407–409.
18) Sugiura I. Intra-osseous glomus tumour. *J Bone Joint Surg,* 1976; 58: 245–247.
19) Massari L, Caruso G, Chinni C, Ferron P, Gaban A, Traina F. Intraosseous glomus tumor of the ankle: a case report and review of the literature. *Foot Ankle Int*, 2006; 27:1148–1151.
20) Bambakidis NC, Gore P, Eschbacher J, Coons S, Albuquerque FC. Intraosseous spinal glomus tumors: case report. *Neurosurgery,* 2007; 60: 1152–1153.
21) Drape JL, Idy-Peretti I, Goettmann S, Wolfram-Gabel R, Dion E, Grossin M, Benacerraf R, Guerin-Surville H, Bittoun J. Subungual glomus tumors: evaluation with MR imaging. *Radiology,* 1995; 195: 507–515.
22) Weiss SW, Goldblum JR. Perivascular tumors. *In Soft tissue tumors.* 4th edition. pp.985–1035, 2001, Mosby, St Louis.
23) Kransdorf MJ, Murphey MD. Vascular and lymphatic tumors. *In Imaging of soft tissue tumors.* 2nd edition. pp. 150–188, 2006, Lippincott Williams & Wilkins, Philadelphia.
24) Mathis WH, Schulz MD. Roentgen diagnosis of glomus tumors. *Radiology,* 1948; 51: 71–76.