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Case Report

Embolization in the Treatment of an Intraosseous Glomus Tumor in the Upper Thoracic Spine Complicating Compression Myelopathy: A Case Report and A Literature Review

Kompresyon Miyelopatisi ile Komplike Olan Üst Torasik Omurga İntraosseöz Glomus Tümörü Tedavisinde Embolizasyon: Bir Olgu Sunumu ve Literatür Derlemesi

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ABSTRACT

Glomus tumors are very infrequent in the spine. The lesions can grow intraosseously along the entire spinal axis. A single female presenting with back pain from the upper thoracic spine is reported on. Removal of this lesion may require reconstruction of the anterior column with posterior fixation resulting in significant blood loss. The current report describes an embolization procedure prior to removal in order to reduce the significant blood loss that occurs with removal of this lesion, and summarizes the clinical and pathological characteristics of this rare tumor. A single, recent case and removal of an intraosseous tumor arising from the upper thoracic vertebra of T2-T4 is described. A 45-year-old female presenting with symptoms secondary to a glomus tumor of the upper thoracic vertebra of T2-T4 underwent resection of the lesion followed by reconstruction of the anterior column following preoperative emobolization. She had neurological symptoms for 3 years, and an irregular crescent-shaped lesion was seen going through the foramen at T3 to the chest cavity in the MRI scans. The operation was performed with a posterior approach in a single stage. The use of preoperative embolization of the T2-T4 segmental arteries resulted in significantly less blood loss as compared to without an embolization procedure. It was confirmed by histopathological examination that the glomus tumor rose from the smooth muscle cells in the right paravertebral muscles of T2. The glomus tumor has not recurred in the MRI during the five-year follow-up. Intraosseous glomus tumors are rare lesions that may extend into the epidural space and through the neural foramina and chest compartments resulting in neurological compromise. Over time, they may grow very large. Radiotherapy can be useful for eradication of this rare lesion. However, it can reoccur requiring extensive surgery resulting in significant blood loss. Preoperative embolization results in a reduction of blood loss and can be a very useful technique when performing the resection of large lesions suspected to be glomus tumors.

KEYWORDS: Embolization, Intraosseous glomus tumor, Upper thoracic spine

ÖZ

Glomus tümörleri omurgada çok nadirdir. Lezyonlar tüm spinal aks boyunca intraosseöz olarak büyüyebilir. Üst torasik omurgada sırt ağrısıyla gelen tek bir kadın hasta bildirilmiştir. Lezyonun çıkarılması için posterior fiksasyonla birlikte anterior kolon rekonstrüksiyonu gerekebilir ve önemli kan kaybı oluşabilir. Mevcut rapor, bu lezyonun çıkarılmasıyla oluşan önemli kan kaybını azaltmak üzere, eksizyon öncesinde embolizasyon işlemini tanımlamakta ve bu nadir tümörün klinik ve patolojik özelliklerini özetlemektedir. Üst torasik vertebralardan (T2-T4) köken alan bir intraosseöz tümörün çıkarıldığı, yakın zamanlı, tek bir olgu tanımlanmaktadır. Kırk beş yaşında bir kadın, T2-T4 üst torasik vertebrada glomus tümörüne sekonder belirtilerle gelmiş ve lezyon rezeksiyonu sonrasında preoperatif embolizasyon ve anterior kolon rekonstrüksiyonu yapılmıştır. Nörolojik belirtiler 3 yıldır devam etmiş ve manyetik rezonans görüntüleme (MRG) taramalarında T3'te foramen içinden göğüs boşluğuna giden düzensiz ve yarım ay şekilli bir lezyon görülmüştür. Ameliyat tek bir evrede posterior yaklaşımla yapılmıştır. T2-T4 segmental arterlerinin preoperatif embolizasyonu, embolizasyon işlemi yapılmayan duruma göre çok daha az kan kaybıyla sonuçlanmıştır. Histopatolojik incelemeyle glomus tümörünün, T2 düzeyinde, sağ paravertebral kasların düz kas hücrelerinden köken aldığı gösterilmiştir. Glomus tümörü beş yıllık takip boyunca MRG'de tekrarlamamıştır. İntraosseöz glomus tümörleri nadir lezyonlardır ve epidural boşluğa ve nöral foramenler ve göğüs kompartmanları içinden yayılıp nörolojik problemlere yol açabilir. Zamanla çok büyüyebilirler. Bu nadir lezyonun tedavisinde radyoterapi faydalı olabilir. Ancak lezyon tekrarlayabilir ve geniş cerrahi gerektirip sonuçta önemli kan kaybına yol açabilir. Preoperatif embolizasyon kan kaybını azaltır ve glomus tümörleri olduğundan şüphelenilen büyük lezyonların rezeksiyonu yapılırken çok faydalı bir teknik olabilir.

ANAHTAR SÖZCÜKLER: Embolizasyon, İntraosseöz glomus tümörü, Üst torasik omurga

INTRODUCTION

The glomus body is involved in thermal regulation, and is located in the stratum reticularis of the dermis layer in the skin (19). Sometimes the glomus body is in the subungual region, the digits, or the palm. The glomus body is a temperature-sensitive organ of modified perivascular smooth muscle cells, which is involved in the vascular regulation of skin temperature. The role of the glomus body is to shunt blood away from the skin surface when exposed to cold temperature, thus preventing heat loss, and allowing maximum heat flow to the skin in warm weather to allow heat to dissipate. The glomus body has high sympathetic tone and potentiation leads to near complete vasoconstriction. Glomus tumors are very scarce, with an estimated incidence of 1.6% among the 500 consecutive soft tissue tumors reported by the Mayo Clinic (17). Glomus tumors are benign lesions that are derived from neuromyoarterial glomus cells. The vast majority of glomus tumors occur in the distal extremities, particularly the subungual region and the forearm (8). Intraosseous glomus tumors are extremely rare. We report a case of an intraosseous glomus tumor of the upper thoracic spine, discuss the imaging and histological findings, and the treatment.

CASE REPORT

A 45-year-old female was first hospitalized in December 2004. She had presented with progressive numbness and weakness of the lower limbs with difficulty walking for a year. Physical examination showed that the spinal process of T2-T4 was hypoesthetic with decreased sensation below the papilla plane. The muscle strength of the bilateral iliopsoas and quadriceps femoris muscles was level 2. The myodynamia with extended halluces and toes was level 3, and with extended and flexed feet was level 4. Abdominal reflexes were absent.

In December of 2004, CT imaging revealed osteolytic areas at levels T2-T4 with osteosclerotic changes. There was a mucinous mass in the T2 paravertebral space (Figure 1A). The lesion involved paravertebral soft tissue, and was shaped like a crescent. It measured 5 cm x 6 cm in size in the vertebral body. Magnetic resonance imaging (MRI) showed the lesion to be homogeneous, with slightly higher signal intensity on T1-weighted images, and equal signal intensity on T2-weighted images. The mucinous mass was seen outside the T2 paravertebral space with cord compression. The pathological findings of a CT-guided biopsy revealed this to be a glomus tumor. The patient was subsequently treated with radiotherapy. The first choice of the radiotherapy for the patient was the financial condition as the surgical treatment was more expensive than the radiotherapy in China. The radiotherapy was carried out with a single photon less than 40 Gy, 50 mm depth by a linear accelerator. Thereafter, a month later, the daily fractions of 2 Gy was additional. After three months of radiotherapy treatment, the symptoms of the patient gradually improved. All symptoms were relieved almost completely after a year, and the patient's neurological function was entirely normal. However, in November 2006, the patient complained of new onset of pain at the dorsal part of the chest again. Subsequent physical examination showed the muscle strength of the bilateral iliopsoas and quadriceps femoris muscles to be level 5. The myodynamia with extended halluces and toes was level 4, and with extended and flexed feet was level 5. Abdominal reflexes were again absent.

MRI showed a compressive fracture of the T3 vertebra, with obvious destruction and a crescent-shaped lesion involving the upper thoracic vertebra of levels T2-T4. This lesion extended through the neural foramen at T3 to the chest cavity (Figure 1A). In comparison with the CT and MRI imaging performed in Dec 2004, the mass was noted to have grown slightly.

After informed consent was obtained, the patient underwent posterior decompression, tumor resection and corpectomy.

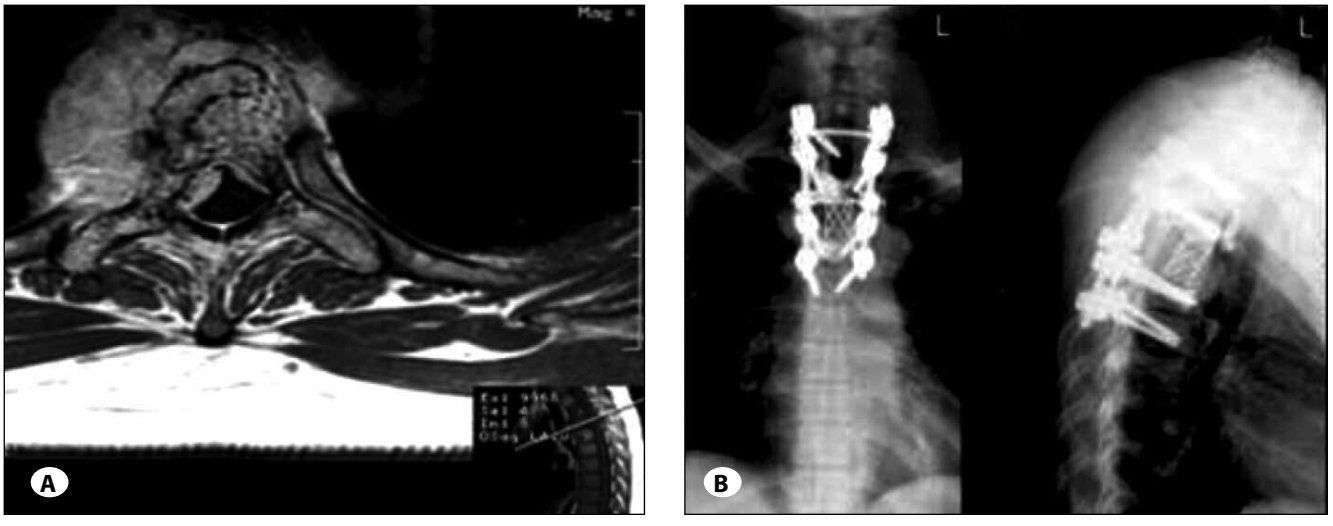

Figure 1: A) MRI showed compression fracture of vertebra of T3, and destruct obviously and an irregular ball-shaped lesion involving the upper thoracic vertebra of T2-T4, which was extended through the neural foramen at T3 to chest cavity. **B)** Postoperative x-ray showed reconstruction of the anterior column with a titanium cage, and posterior instrumentation.

This was followed by reconstruction of the anterior column with a titanium cage filled with autograft, and placement of pedicle screws from T2 to T6 by a one-stage posterior approach (Figure 1B). This procedure results in significant intraoperative blood loss. Therefore, in order to decrease blood loss during the operation, embolization of the tumor arteries was performed preoperatively. Access to the segmental arteries was performed using digital subtraction angiography (DSA)-guided arteriography under local anesthesia through the femoral artery. Superselective catheterization was performed with injection of contrast media to verify the vessels (Figure 2A). Once verified, embolization of the arteries was done using a gelatin sponge (Figure 2B).

The percutaneous embolization technique is now presented. When the tumor blood supply was derived from an artery, it was necessary to embolize by prior angiography. However, the vascular supplying segments of the spinal cord were not advocated for embolization for safety reasons. At the endpoint of embolization, an angiographic series was performed to verify reduction of tumor vessels and hemostasis. The adjacent segments of the embolization should also be checked after injection to determine if the tumor will blush or the additional collaterals have been reduced. This embolization procedure resulted in a blood loss of 650 ml during the operation, which was significantly less than the 1500-2000 ml of blood loss reported in the literature.

The glomus tumor was verified by postoperative pathology evaluation (Figure 3A). Microscopic examination showed a hypercellular neoplasm. There were round, polygonal epithelioid cells with the monotonous nuclei in the neoplasm. It was found that scattered, dilated or small-to-medium vascular channels were also in the focus. Nuclear atypia and

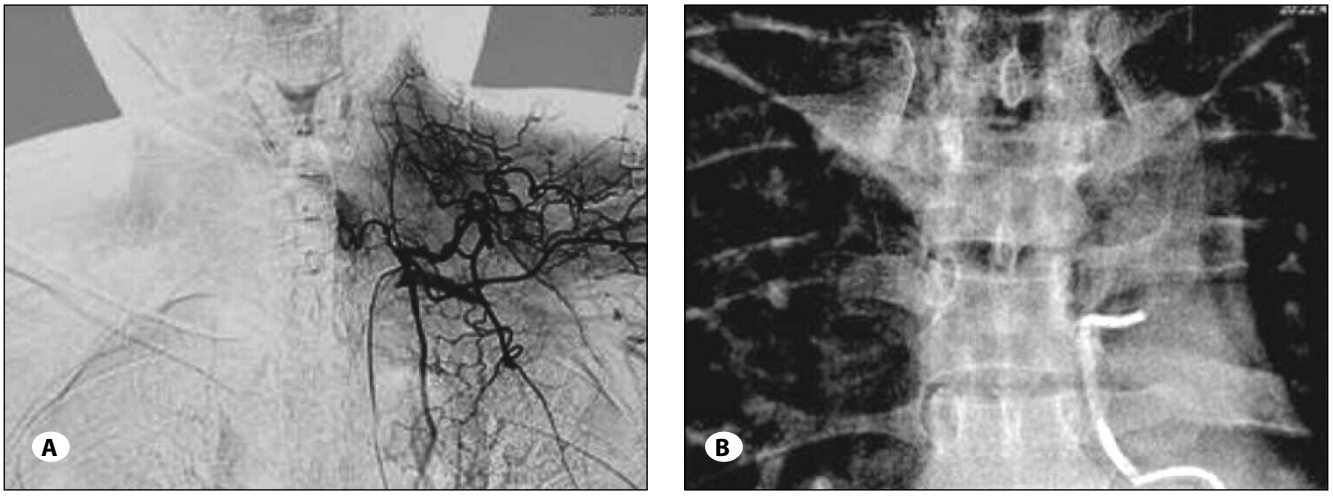

Figure 2: In order to decrease the bleeding during the operation, embolization of the segmental arteries at T2-T4 was performed before operation. **A)** pre- embolization. **B)** post-embolization.

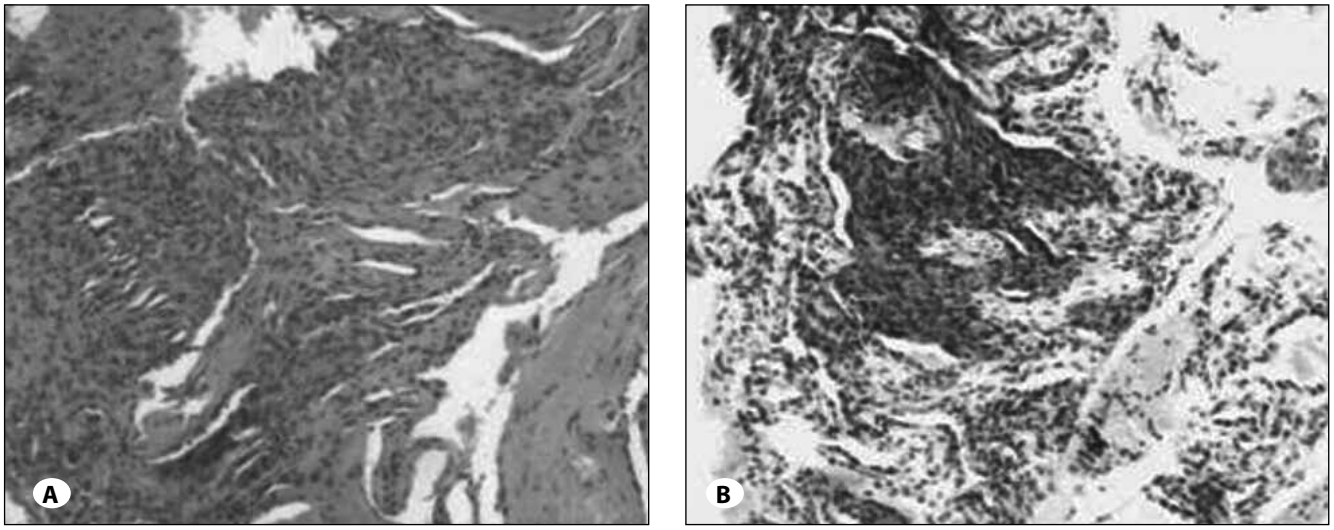

Figure 3: A) microscopic examination showing a proliferation of glomus cells with round monotonous nuclei (hematoxylin and eosin; original magnification, 200). **B)** microscopic examination showing that glomus cells are immunoreactive for smooth muscle actin. (Envision microscopic examination, 200).

necrosis were not in appearance. S-100, chromogranin, epithelial membrane antigen, and CD34 were immunonegative in the neoplastic cells (Figure 3B). Smooth muscle was immunoreactive. Therefore, the diagnosis of a glomus tumor was established by both the staining pattern and the histological features.

At the 5-year follow-up examination, the patient had recovered completely. There was no evidence to demonstrate residual or recurrent tumor in the postoperative MRI (Figure 4A,B).

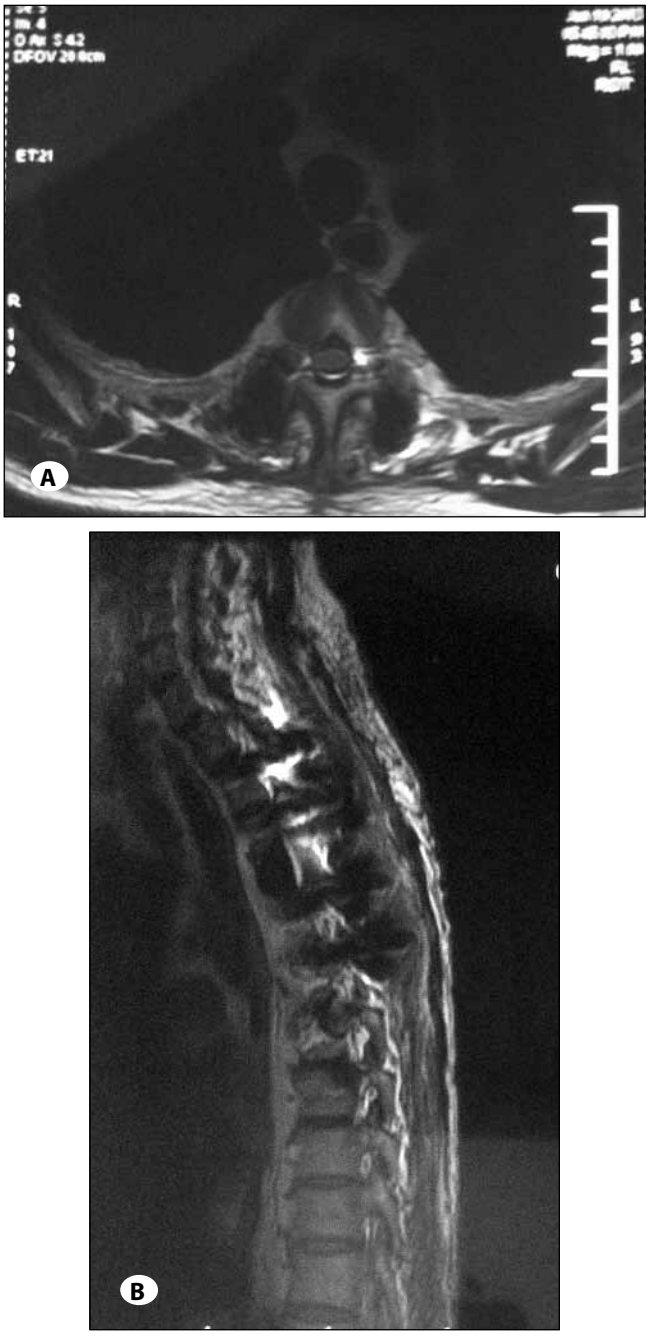

Figure 4: A) It showed there was no paravertebral recurrence of the glomus tumors in the transverse section of MRI. **B)** The inner fixation of the patient was maintained properly without loosening in the sagittal section of MRI.

DISCUSSION

Glomus tumors are, with the rare exception of glomangiosarcomas, benign neoplasms that are derived from the neuromyoarterial plexus (11). Clinically, this tumor can be seen as a small, red-blue, superficial nodule with symptoms of paroxysms of spreading pain due to changes in temperature or pressure. The classic clinical triad of dull pain, intensive point of tenderness, and cold sensitivity is present in approximately 30% of patients. Substance P has been associated with a painful glomus tumor, and a striking female pre-disposition (3:1) is observed in subungual lesions, although overall there is no gender-based pre-disposition. Its predilection is for adults between 20 to 40 years of age, and often affects the third to fifth decades of their lives (4). It has a tendency to be hereditary. The 1p21-1p22 codes for glomulin and the protein plays a role in the genesis (6, 7). Glomus tumors normally develop in the subungual region, but can develop at any site where the normal glomus body may exist. However, an intraosseous glomus tumor of bone is extremely rare, especially in the spine (3, 13).The origin of intraosseous glomus tumors remains controversial. They presumably can originate from soft tissue (located at the triage department of the normal arteriole and normal venule), and erode through the opening in the bony cortex, or arise within the bone. They can also originate either from normal glomera that occasionally exists in the terminal phalanges, or from pericytes in the blood vessel walls (2,4,19). Typically, in essence, the iconography feature of the tumor is a high signal intensity on T2-weighted images, and low signal intensity on T1-weighted images.

In our case, there was no evidence of an opening of the cortex during our operation, nor any microscopic evidence of continuity between the tumor and the pericyte vessel. However, CT and MRI demonstrated the continuity of the tumor lesion to the vascular groove. Therefore, we postulated that this intraosseous glomus tumor might have arisen from pericytes in the blood vessel walls. Intraosseous glomus tumors of the spine are extremely rare lesions and there is little experience in the literature for treatment (Table I). In contrast to the experience for the treatment of vertebral hemangiomas, we have a different choice of treatments for intraosseous glomus tumors. Vessel embolization, ligation or irradiation and surgery are treatments used for the glomus tumors. If there is spinal cord compression in the vertebral hemangioma, radiation treatment could be considered initially. The embolization has produced good results (7,11).

Our patient had a transient complete paraplegia. She recovered thoroughly after radiotherapy, which indicates that radiotherapy may be a useful non-invasive treatment option for removal of glomus tumors. However, it is known that radiotherapy does not result in complete eradication of the tumor. In our case, the symptoms of the patient arose again two years after radiotherapy. MRI indicated a recurrence of the tumor and it had grown bigger in size, resulting in the collapse

Table I: The Authors Reported the Operation of the Glomus Tumors in the Past

Series (ref. no.)	no.	Age (yr) /sex	Presenting symptoms	Tumor location	Operation
Bessho Y et al., 1991 (5)	1	49/M	Back pain compressive myelopathy	T2 pedicle vertebral body	T2-3 costotransversectomy
Robinson et al., 1996 (14)	2	45/F	Back pain	L1 pedicle	Posterior laminectomy and resection
Payer et al., 2002 (13)	3	55/F	Back pain, Compressive myelopathy	T4 vertebral body	Transthoracic T4 corpectomy
Bambakidis et al.2007 (3)	4	44/M	Radicular symptoms	L3 vertebral body, epidural space at L3–L4, L3 neural foramen, abdominal cavity	Posterior decompression, Angiographic embolization, transthoracic resection, 360º reconstruction and fusion
Becce F et al. 2012 (4)	5	73/M	Upper and lower back pain symptoms	The right pedicle of the eleventh thoracic vertebra (T11).	Percutaneous CT-guide radio-Frequency ablation (RFA)
Current report	6	45/F	Back pain, Compressive myelopathy	T2-T4 vertebral body , epidural space neural foramen, chest cavity	Stab biopsy, angiographic embolization, lateral posterior undermining decompression resection, 360º reconstruction and fusion

***F:** female, **M:** male, **T:** thoracic, **L:** lomber.*

of the vertebra and compression of the spinal cord. Surgical intervention may be a good choice to cure glomus tumors. Total resection or en-bloc resection can decrease the rate of recurrence of these tumors. If the glomus tumor is of large diameter (>2 cm), deep in location, and malignant, the risk of metastasis should be considered (3). Therefore, embolization before the operation should be integrated to the procedure. It should be the foundation of the treatment, and can provide assurance for total resection of glomus tumors.

With the development of catheterization techniques, embolization of the vessels to vertebral hemangiomas has become an approved therapy. However, embolization as the sole therapy is still controversial (9,14). Preoperative embolization for hypervascular spinal tumors has been proven to be effective in reducing tumor size and intraoperative bleeding (1, 9, 10). It is safe and effective, but it should primarily be used as an adjunct to definitive treatments such as surgery or radiotherapy. Bambakidis et al. (3) described a 44-year-old man with a dumbbell-shaped glomus tumor involving the L3 foramen where emergent embolization with adhesive and coil angiographic embolization was used for severe intraoperative hemorrhage that could not be readily controlled during the operation, and was satisfied with reconstruction of the vertebral column.

In order to prevent recanalization or tumor revascularization, surgery should be performed in the 24 to 48 hours after embolization. Owen argued that transarterial embolization could reduce operative hemorrhagic risks, and might also increase tumor sensitivity to chemotherapy or radiation therapy in primary (giant cell tumors, aneurysmal bone cysts, vertebral hemangioma, osteosarcoma) or metastatic bone tumors (12). For our patient, we applied preoperative embolization before total excision of the vertebra. The operation was straightforward with a clear field of view and a blood loss less than 1000 ml. A review of the literature reveals that removal of a vertebra with reconstruction can result in a blood loss of 1500-2000 ml. As reported by some authors, the mean intraoperative blood loss was 1500 ml in primary tumors, the average transfusion volume was 1520 ml, and the duration of the surgery was 225 minutes (18,20,21). We therefore think that preoperative embolization should be an essential step in the treatment of this rare lesion. Ruggieri et al. (16) reported the local recurrence rate as 91% vs. 86% with and without preoperative selective arterial embolization respectively for the sacral giant cell tumor. With the embolization procedure, the hemorrhage of the tumor was obviously decreased.

Most authors have reported that surgery is indicated when the spinal cord compression gets worse (7). The most important problem is uncontrolled intra-operative blood loss. However, the surgical method for removal of vertebral hemangiomas remains controversial. A decompressive laminectomy would have been a treatment option if the cause of compression were the glomus tumor involving only the posterior elements of the vertebra. However, if the glomus tumor involves the vertebral

body with obvious anterior spinal cord compression, partial or complete vertebral body resection and reconstruction might be indicated. With the total resection of the range of the margin and the wide reacting region, our patient has reported a satisfactory outcome without recurrence at the 5-year follow-up. We have concluded that diagnosis and surgery for decompression of this rare legion is wise to prevent progressive spinal cord compression if there are no signs for tendency of the primary tumor to become malignant. Regardless of partial or complete vertebral resection, intraoperative bleeding would be turbulent. Therefore, preoperative embolization was important in order to reduce the hemorrhage volume. There are no reports on whether the irradiation of vertebral hemangiomas before surgery could decrease the intraoperative bleeding more effectively than that of embolization. Preoperative embolization plays a role in decreasing the level of the intraoperative blood loss. Thus, it made possible the facilitation of the removal of the tumors, and maximally controlled the tumor growth. However, in patients with a posterior compressive vertebral glomus tumor, decompressive laminectomy (in order to perform local decompression and the excision of the residual tumors and the destroyed vertebra) plus irradiation can also be an effective and safe treatment.

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