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CASE REPORT

Long-term survival case of malignant glomus tumor mimicking "dumbbell-shaped" neurogenic tumor

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Abstract
Purpose We report a very rare case of long-term survival case of malignant glomus tumor (MGT) without widespread metastasis.
Study participants Sixty-three-year-old woman who experienced multiple relapses over a 40-year period beginning with a scapular lesion since she was at age 21.
Results After four local recurrences, the patient underwent wide resection of the scapula at age 36. Thirteen years later, the patient had a neck tumor at the same side with the scapulectomy. The tumor was originated from the C6 spinal nerve and was resected. Twelve years after the surgery, the cervical tumor relapsed with myelopathy at age 61. The tumor infiltrated from the extraspinal canal to the intraspinal canal at the C5/C6 foramen and appeared as a dumbbell-like tumor. Furthermore, the local recurrence occurred 2 years after the operation at age 63. The tumors that were detected at age 36 and age 63 were confirmed to be histologically identical. Those were more than 2 cm in size and arose from a deep location. The basement membrane stained positively for collagen type IV and α-smooth muscle actin on immunohistochemistry. Based on these aspects, the tumor was diagnosed as an MGT.
Conclusions This is the first case report of MGT which lesions at two time points were confirmed to have common histological features and which confirmed the long-term survival over a 40-year period.

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Keywords Malignant glomus tumor (MGT) · Long-term follow-up · Histology

Introduction

Glomus tumors are uncommon and account for approximately 1.6 % of the 500 consecutive soft tissue tumors reported by the Mayo Clinic [1]. Although glomus tumors are considered to be benign, some may present unusual clinical features such as large size, deep soft tissue or visceral location, infiltrative growth pattern, or multicentricity [2–5]. Malignancy is a very rare feature of glomus tumors. To our knowledge, this report is the first to describe the long-term follow-up of a malignant glomus tumor (MGT) based on histological findings over 40 years. No reports have described cases of MGT wherein the lesions were confirmed as having common histological features at two time points and the patients exhibited long-term survival.

Case report

A 49-year-old woman presented with a cervical tumor on the right side. At 21 years of age, she had developed right scapular hemangioma, which was finally treated with scapulectomy at 36 years of age, after 4 recurrences. The cervical tumor was found to have originated from the C6 spinal nerve and was resected.

At 12 years after the resection, the patient developed cervical myelopathy. Magnetic resonance imaging indicated that spinal cord was compressed by a tumor (Fig. 1), which had relapsed at the C6 spinal nerve root and entered the spinal canal via the C5/6 intervertebral foramen.

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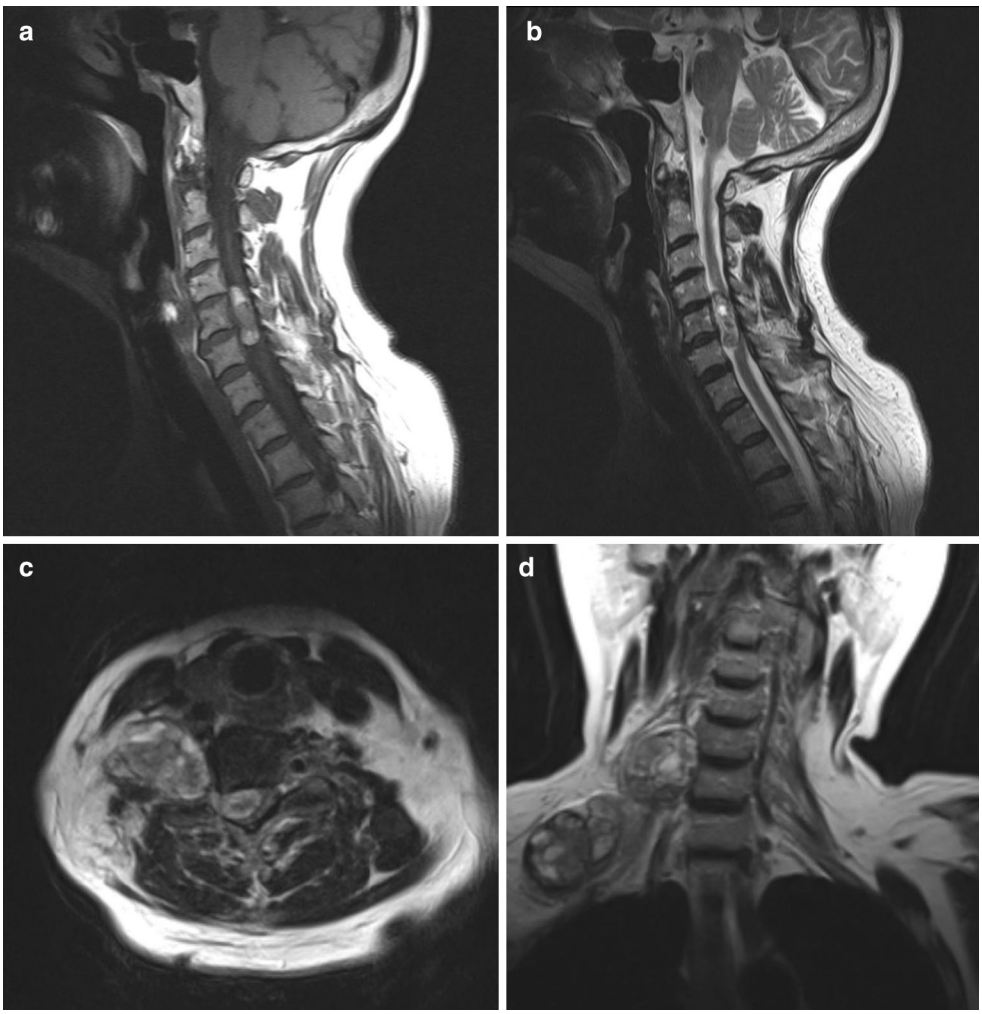


Fig. 1 Magnetic resonance imaging (MRI) of the cervical spine. **a**, **b** T1- and T2-weighted image of sagittal section showing intraspinal canal mass lesion. **c**, **d** Axial and coronal section of C5/6 level showed that the tumor had entered the dura mater of the spinal cord via the C5/6 intervertebral foramina and compressed the spinal cord

Moreover, another tumor had arisen from the C5 spinal nerve. The tumors were marginally resected via two-step operation.

After 2 years (63 years of age), local recurrence was observed at the residual C6 spinal nerves. The tumor was simultaneously resected via an anterior–posterior spinal procedure. First, the anterior procedure, which involved wide resection of the tumor and ligation of the vertebral artery, was performed. Thereafter, anterior spinal C5–7 fusion was performed for stabilization (Fig. 2).

Second, a dorsal procedure was performed to resect the residual tumor at the intraspinal canal. On histological examination, the tumor exhibited polygonal or dense spindle cells, whereas hematoxylin–eosin staining indicated hyperplasia around the blood vessels (Fig. 3).

Furthermore, immunohistochemical analysis indicated positive staining of the basement membrane for collagen type IV and α-smooth muscle actin (Fig. 4).

Thus, the specimens obtained during the last two operations were histologically diagnosed as glomus tumors.

We obtained specimens from the scapular region from the oncology center where the patient was treated 27 years previously (Fig. 5). The tumors, which were resected at ages 36 and 63 years, were confirmed to be histologically identical.

We have followed up the patient for 3 years since the previous operation, and no recurrence has been observed.

Discussion

Glomus tumors are generally benign, solitary tumors that can be treated via simple excision. Although a few MGT cases have been reported thus far, they have primarily involved locally invasive tumors, and the occurrence of metastases is rare [6, 7]. Through a retrospective analysis

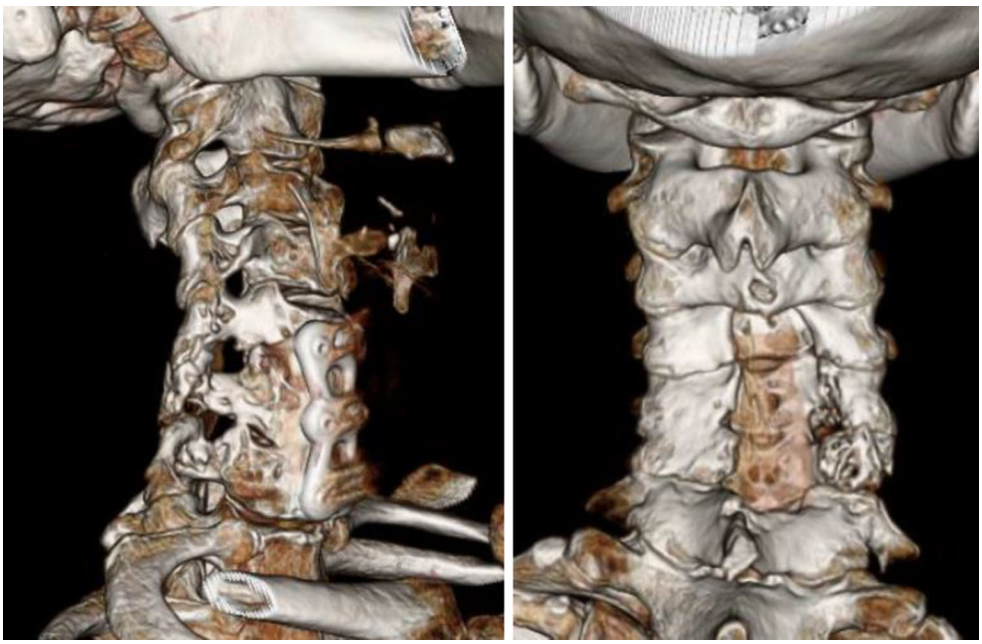
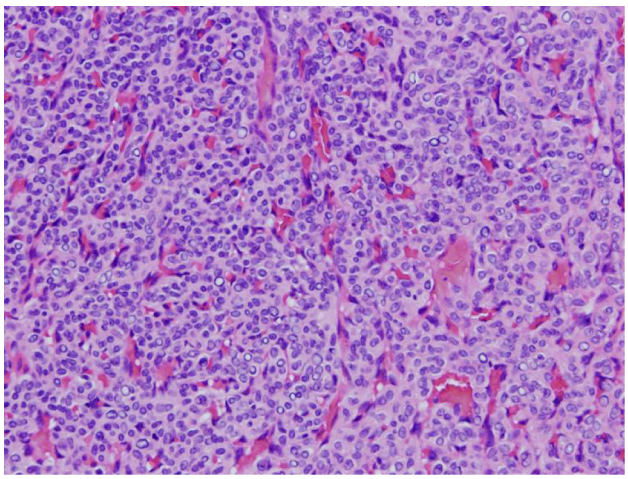


Fig. 2 The three-dimensional computed tomography after tumor resection with anteroposterior spinal procedure simultaneously and anterospinal fusion



Hematoxylin-Eosine (HE) staining OM ×20

Fig. 3 The tumor was composed of polygonal or spindle, rounded cells and hyperplasia around the blood vessels (HE stain)

of 52 cases of atypical glomus tumor of the peripheral soft tissues to establish criteria for malignancy, Folpe et al. observed that malignancy developed in benign glomus tumors [8]. The authors proposed that a deep location and a size of >2 cm, atypical mitotic figures, or moderate- to high-grade nuclear atypia and ≥5 mitoses per 50 high-powered fields are criteria for malignancy. Moreover, cytoplasmic staining for actin and at least focal staining for collagen type IV are highly suggestive of MGT [8]. The features of the present case are consistent with these criteria for malignancy. MGTs are known to exhibit an aggressive clinical course and are believed to be low-grade sarcomas [9, 10]. In the present report, we describe a rare case of scapular MGT that exhibited repeated local relapse, without any distant widespread metastasis to other organs for many years. Moreover, infiltration from the scapular region into the intraspinal canal led to the formation of a dumbbell-shaped tumor, which is also a rare phenomenon. The probable underlying mechanism may become evident when the course of the suprascapular nerve is considered. The tumor was derived from the C5 and C6 nerves that are associated with the supra- and infraspinatus muscles. We speculate that the tumor arose from the suprascapular nerve and infiltrated through the nerve root into the intraspinal canal after many years.

In conclusion, we report a rare case wherein long-term survival was observed despite the development of MGT. Physicians should carefully consider the treatment of large glomus tumors that arise in non-acral sites. A long-term follow-up of at least >10 years may be required after the resection. Nevertheless, further studies are warranted to determine the appropriate treatment for MGTs, given its significant potential for aggressive behavior.

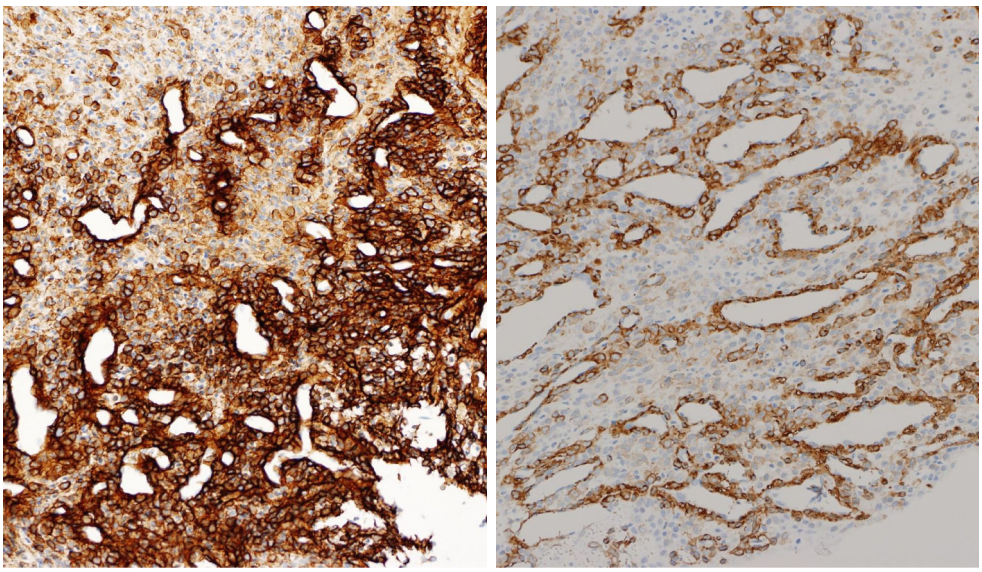


Fig. 4 Immunostaining showed that the basement membrane was positive for type IV collagen and α-smooth muscle actin

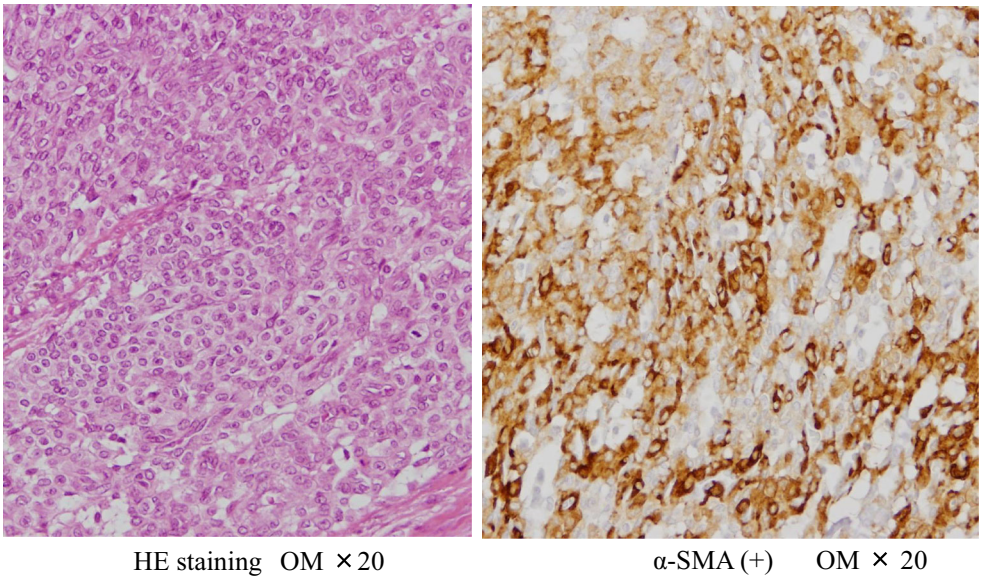


Fig. 5 The histology of tumor that was resected when she was 36 years old: tissue staining of these specimens confirmed that the tumor of the right shoulder was very similar to our histology in HE stain and was positive for α-smooth muscle actin

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Compliance with ethical standards

Conflict of interest No benefits in any form have been or will be received from a commercial party related directly or indirectly to the subject of this manuscript.

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