



Giant Cell Tumor of Soft Tissues of Neck

Yin-Jei Chan¹, Tai-Kuang Chao², Yueng-Hsiang Chu^{1*}, and Hsing-Won Wang¹

¹Department of Otorhinolaryngology - Head and Neck Surgery;

²Department of Pathology, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan, Republic of China

Conflict of Interest: The authors declare that they have no competing interests. We report a case of giant cell tumor of soft tissues of the neck in a 32-year-old woman with a 2-year history of painless neck mass. MRI showed an enhancing mass about 2.1 × 1.7 cm in size in the left cervical posterior paraspinal space. Intraoperatively, a well-encapsulated tumor was found adjacent to the spinous process of the third cervical spine. Histological features showed characteristic dispersed multinucleated giant cells admixed with mononuclear cells. Immunohistochemical staining was positive for CD68 and vimentin, but negative for desmin, smooth muscle actin and S-100 protein. Both mitotic activity and Ki-67 proliferation index were exceedingly low. These features were consistent with giant cell tumor of soft tissues with low-grade potential. Marginal excision was performed due to its proximity to vital structures. She was free of local recurrence after 15-month follow-up. A brief review of the literature of giant cell tumor of soft tissues of the neck was also presented.

Key words: giant cell tumor, soft tissues, neck

INTRODUCTION

Giant cell tumors (GCTs) of bone, characterized by the presence of large, osteoclast-like multinucleated giant cells, are low-grade neoplasms typically affecting the ends of long bones and demonstrating a potential for local recurrence. On the other hand, giant cell tumor of soft tissues (GCT-ST) is exceedingly uncommon and is a primary soft tissue neoplasm clinically and histologically similar to GCT of bone. GCT-ST in the head and neck region is rare and only individual case reports are documented. In the literature, most GCTs that arise in the head and neck region occur in the larynx,^{1,2} pharynx,³ and salivary glands.⁴ GCT-ST confined to the neck region is extremely rare. Only two cases of malignant GCT-ST confined to the neck have been reported so far.^{5,6} Both GCT-ST and GCT of bone may recur locally, but rarely metastasize. However, in some reports, GCT-ST has displayed a wide spectrum of clinical behaviors ranging from local benign tumors to highly aggressive malignan-

cies.⁷⁻⁹ It is difficult to predict the clinical behavior of GCT-ST on the basis of histological features and depth of tumor alone. Here we presented a case of GCT confined to the extralaryngeal soft tissues of the neck. In addition, a brief review of the literature on GCT-ST, its treatment and the pathological features is presented.

CASE REPORT

A previously healthy 32-year-old woman with a 2-year history of neck mass was initially evaluated. Medical history was unremarkable and no trauma history was told. On physical examination, there was a fixed, very hard, nontender neck mass of about 2 cm in diameter located posterior and inferior to the left mastoid process tip. Magnetic resonance imaging (MRI) of the head and neck showed an enhancing well-encapsulated mass about 2.1 × 1.7 cm in size with eccentric tumor necrosis in the left posterior paraspinal space embedded between the semispinalis cervicalis muscle and capitis muscle (Figure 1). The radiological differential diagnoses include neurogenic tumor such as schwannoma, or less likely other soft tissue tumor such as malignant fibrous histiocytoma. Result of fine needle aspiration cytology was nondiagnostic. The patient underwent excision biopsy under general anesthesia. Intraoperatively, a well-encapsulated tumor was found adjacent to the spinous process of the third cervical spine. This mass was not associated with the cervical spine or its ligaments and was excised en bloc.

Received: August 4, 2009; Revised: October 1, 2009;
Accepted: October 7, 2009

*Corresponding author: Yueng-Hsiang Chu, Department of Otolaryngology - Head and Neck Surgery, Tri-Service General Hospital, National Defense Medical Center, No.325, Sec. 2, Cheng-gong Rd, Taipei 114, Taiwan, Republic of China. Tel: +886-2-8792-7192; Fax: +886-2-8792-7193; E-mail: chufascia@pchome.com.tw

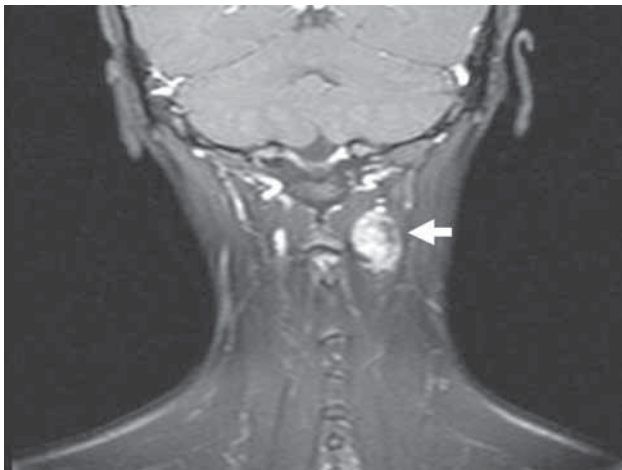


Figure 1. Coronal T1-weighted magnetic resonance imaging scans with gadolinium enhancement showed an enhancing well-encapsulated mass (arrow) about 2.1×1.7 cm in size with eccentric tumor necrosis in the left posterior paraspinal space embedded between the semispinalis cervicalis muscle and capitis muscle.

The postoperative course was uneventful, and the patient had no evidence of local recurrence or distal spread after 15-month follow-up.

Gross examination of the surgical specimen revealed a well-circumscribed mass, measuring approximately $2.5 \times 2 \times 2$ cm in size. The tissue was entirely firm and appeared grayish-brown. Microscopically, the cellular nodule was separated by fibroconnective tissue septae of varying thickness and contained numerous hemosiderin-laden macrophages. These septae were relatively hypocellular and devoid of osteoclast-like giant cell. The nodule comprised a mixture and uniform distribution of round-to-oval mononuclear cells as well as characteristic multinucleated osteoclast-like giant cells (Figure 2). Both types of cell were immersed in a richly vascularized stroma and the giant cells contained a variable number of nuclei, occasionally in excess of 20 nuclei per cell. The mitotic activity without atypical figures was occasionally present; on average at one mitotic figure per 10 high-power fields. Atypia, pleomorphism and tumor giant cells were absent, and necrosis was not found. No neoplastic osteoid or cartilage matrix was observed. Immunohistochemical studies revealed that the mononuclear and multinucleated tumor cells were positive for vimentin and CD68 but negative for desmin, smooth muscle actin, HMB-45, CD1a, ALK and S-100 protein. CD68, the histiocytic marker for further confirmation of the diagnosis

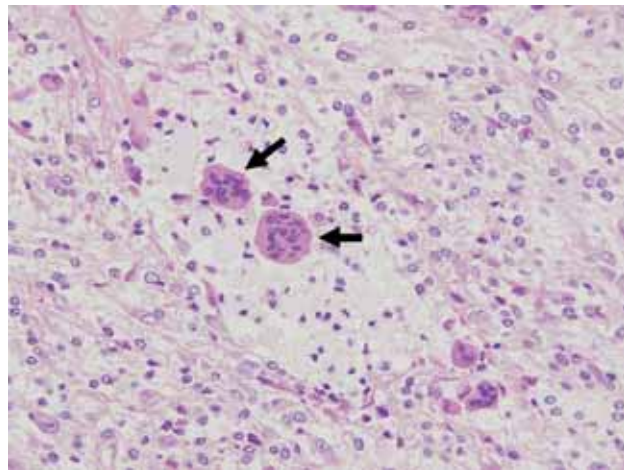


Figure 2. Histopathology section showing the characteristic multinucleated giant cells with bland-looking nuclei (arrows) (H & E stain, original magnification $400 \times$).

of GCT-ST, were strongly positive in the multinucleated giant cells. The immunostaining of Ki-67 proliferation index was exceedingly low. According to the histological and immunohistological findings, the final diagnosis is a giant cell tumor of soft tissues.

DISCUSSION

In 1972, GCT-ST as an uncommon but distinct primary soft tissue tumor was first described.⁸ Later, Folpe *et al.*¹⁰ also proposed a subset of malignant giant cell tumors of soft tissues with low malignant potential. These tumors were termed as “giant cell tumors of low malignant potential.” It was suggested that the term “malignant giant cell tumors” should be restricted to histologically high-grade soft tissue sarcomas. In the current WHO classification of tumors of soft tissue and bone,¹¹ giant cell tumors of soft parts are represented by two different entities. The high-grade spectrum is determined by the diagnosis of undifferentiated pleomorphic sarcoma with giant cells (also known as malignant giant cell tumor of soft parts or giant cell malignant fibrous histiocytoma) and the low-malignant end is covered by GCT-ST (also known as giant cell tumor of low malignant potential). Primary GCT-ST is also known as osteoclastoma and the extremities are the most common sites involved. It is very rarely found in the head and neck region.¹¹ To our knowledge, only two cases of malignant GCT-ST confined to the soft tissues of neck have been reported thus far.^{5,6} We believe that our patient is the first documented

case of GCT-ST confined to the extralaryngeal soft tissues of the neck.

Most cases of GCT of bone and GCT-ST have a benign clinical course, but can be locally aggressive. It is interesting that metastasis of GCT of bone is not uncommon despite of its benign histomorphology. Lung metastasis of osseous GCT was reported in around 3% of cases.¹² It seemed that the risk of metastasis in osseous GCT did not correlate with the proliferation index, increase in mitotic figures and vascular invasion¹³. On the other hand, some controversy about GCT-ST does exist. Guccion *et al.*⁸ suggested that GCT-ST beneath a fascial plane was more likely to show an aggressively clinical course with a higher metastasis rate, whereas only a few superficial tumors developed metastasis. In contrast, Folpe *et al.*¹⁰ did not find any correlation between the depth of tumor involvement and clinical outcome. Moreover, Grabellus *et al.*¹⁴ and May *et al.*¹⁵ reported the unusual cases of aggressive clinical course of GCT-ST with Ki-67 proliferation index of 25% and 35%, respectively. It needs further investigation to specifically correlate the Ki-67 proliferation index as a reliable prognostic indicator with the clinical behavior of GCT-ST. Nevertheless, when malignancy is proved histologically and the proximity of critical anatomic structures prevents wide excision of tumor, radiation therapy can be considered. The use of chemotherapy on malignant GCT-ST is still under debate.⁵ In conclusion, it seemed that the prognosis of GCT-ST varies and the biological course for its local recurrence and metastasis cannot be predicted. Proper surgical excision and long-term follow-up are still essential in the case of GCT-ST.

REFERENCES

1. Wong KK, Seikaly H. Giant cell tumour of the larynx: case report and review of the literature. *J Otolaryngol* 2004;33:195-197.
2. Wieneke JA, Gannon FH, Heffner DK, Thompson LD. Giant cell tumor of the larynx: a clinicopathologic series of eight cases and a review of the literature. *Mod Pathol* 2001;14:1209-1215.
3. Paulino AF, Spiro RH, O'Malley B, Huvos AG. Giant cell tumour of the retropharynx. *Histopathology* 1998;33:344-348.
4. Itoh Y, Taniguti Y, Arai K. A case of giant cell tumor of the parotid gland. *Ann Plast Surg* 1992;28:183-186.
5. Shah DR, Gaw F, Balderrama DC, Dev VR, Liu F, Brown CA. Malignant giant cell tumor of the soft tissues of the neck. *Hospital Physician* 2001;52:55.
6. Angervall L, Hagmar B, Kindblom LG, Merck C. Malignant giant cell tumor of soft tissues: a clinicopathologic, cytologic, ultrastructural, angiographic, and microangiographic study. *Cancer* 1981;47:736-747.
7. O'Connell JX, Wehrli BM, Nielsen GP, Rosenberg AE. Giant cell tumors of soft tissue: a clinicopathologic study of 18 benign and malignant tumors. *Am J Surg Pathol* 2000;24:386-395.
8. Guccion JG, Enzinger FM. Malignant giant cell tumor of soft parts: an analysis of 32 cases. *Cancer* 1972;29:1518-1529.
9. Oliveira AM, Dei Tos AP, Fletcher CD, Nascimento AG. Primary giant cell tumor of soft tissues: a study of 22 cases. *Am J Surg Pathol* 2000;24:248-256.
10. Folpe AL, Morris RJ, Weiss SW. Soft tissue giant cell tumor of low malignant potential: a proposal for the reclassification of malignant giant cell tumor of soft parts. *Mod Pathol* 1999;12:894-902.
11. Fletcher CDM, Unni KK, Mertens F, eds. World Health Organization Classification of Tumours: Pathology and genetics of tumours of soft tissue and bone. 2002 Lyon, France: IARC Press.
12. Tubbs WS, Brown LR, Beabout JW, Rock MG, Unni KK. Benign giant cell tumor of bone with pulmonary metastases: clinical findings and radiologic appearance of metastases in 13 cases. *Am J Roentgenol* 1992;158:331-334.
13. Sulh MA, Greco MA, Jiang T, Goswami SB, Present D, Steiner G. Proliferation index and vascular density of giant cell tumors of bone: are they prognostic markers? *Cancer* 1996;77:2044-2051.
14. Grabellus F, von Winterfeld F, Sheu SY, Metz AK, Jahnke K, Schmid KW. Unusual aggressive course of a giant cell tumor of soft tissue during immunosuppressive therapy. *Virchows Arch* 2006;448:847-851.
15. May SA, Deavers MT, Resetskova E, Johnson D, Albarracin CT. Giant cell tumor of soft tissue arising in breast. *Annals of Diagno Pathol* 2007; 11:345-349.