



CASE REPORT

## Tenosynovial giant cell tumor of the cervical spine: a case report

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### Abstract

**Introduction** Tenosynovial giant cell tumors (TSGCTs) generally occur in the limb joints, and only rarely in the spine. This case report describes a patient with TSGCT of the spine at C1–C2, which was treated surgically and diagnosed as TSGCT. **Case presentation** A 32-year-old woman with a 4-month history of neck pain and numbness in both upper extremities was referred to our department. Magnetic resonance imaging (MRI) revealed a neoplastic lesion extending from the left epidural space to the erector spinae muscles at the C1–C2 vertebral level, which was isointense on T1-weighted images, heterogeneously hypointense on T2-weighted images, and showed heterogeneous enhancement on gadopentetate dimeglumine (Gd-DTPA)-enhanced T1-weighted images. Computed tomography showed no findings suggestive of bone destruction of the vertebral body. Because the neurological symptoms were progressive, total macroscopic resection of the tumor was performed via a posterior approach. Histopathological examination of the resected specimen revealed the diagnosis of TSGCT. Improvement of the both the neck pain and upper-extremity numbness was noted postoperatively. An MRI obtained 6 months after the surgery revealed no evidence of tumor recurrence and the postoperative course was uneventful. **Discussion** TSGCT of the upper cervical spine (C1–C2) is rare, and this is the tenth reported case. If a tumor is heterogeneously hypointense on T2-weighted MRI, which reflects hemosiderosis, the possibility of this tumor should be considered in the differential diagnosis.

### Introduction

Tenosynovial giant cell tumor (TSGCT) is a fibrohistiocytic tumor arising from the tendon sheath, synovium, or bursa and is pathologically characterized by proliferation of mononuclear synovial-like cells and the presence of osteoclast-like multinucleated giant cells and hemosiderin-laden macrophages [1]. TSGCT usually occurs in the limb joints and rarely in the spine. This case report describes a patient with TSGCT of the spine at C1–C2.

### Case presentation

A 32-year-old woman with a 4-month history of neck pain and numbness in the upper extremities were referred to our department. She had no history of trauma. On physical examination, she had impaired dexterity and weakness of the left wrist extensor muscles.

Magnetic resonance imaging (MRI) revealed a neoplastic lesion extending from the left epidural space to the paravertebral muscles at C1–C2, which was isointense on T1-weighted images (T1WI) and heterogeneously hypointense on T2-weighted images (T2WI) and showed heterogeneous enhancement on gadopentetate dimeglumine (Gd-DTPA)-enhanced T1WI (Fig. 1a–e). The cervical spinal cord was displaced to the left laterally by the tumor and T2WI showed changes in intramedullary signal intensity (Fig. 1f). No bone erosion was seen on computed tomography (CT) (Fig. 2).

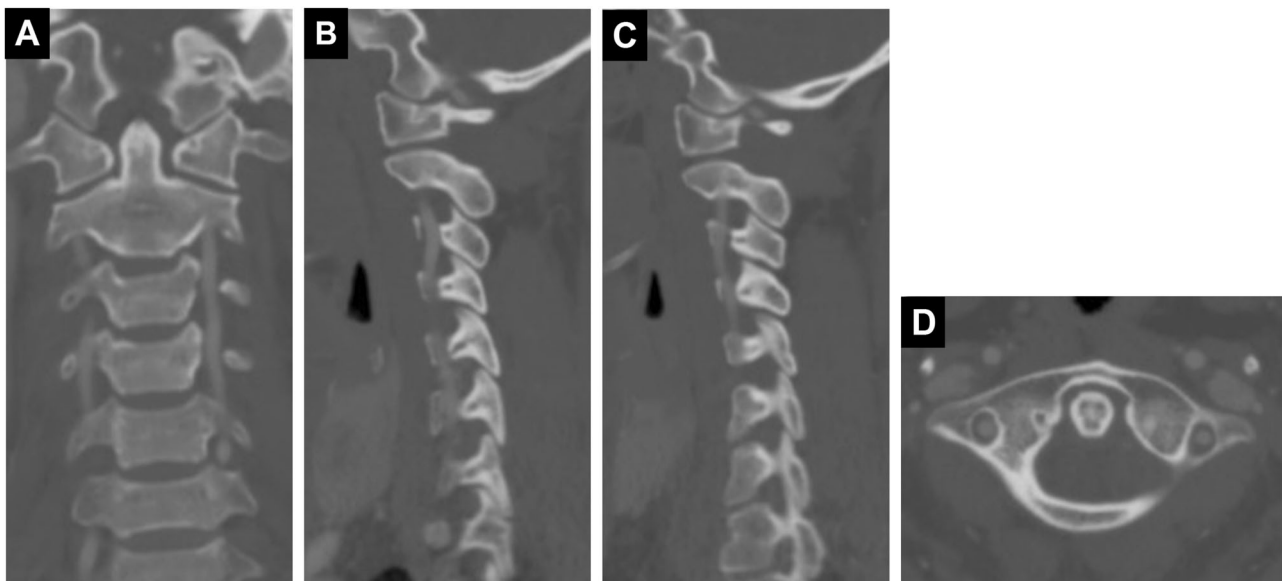
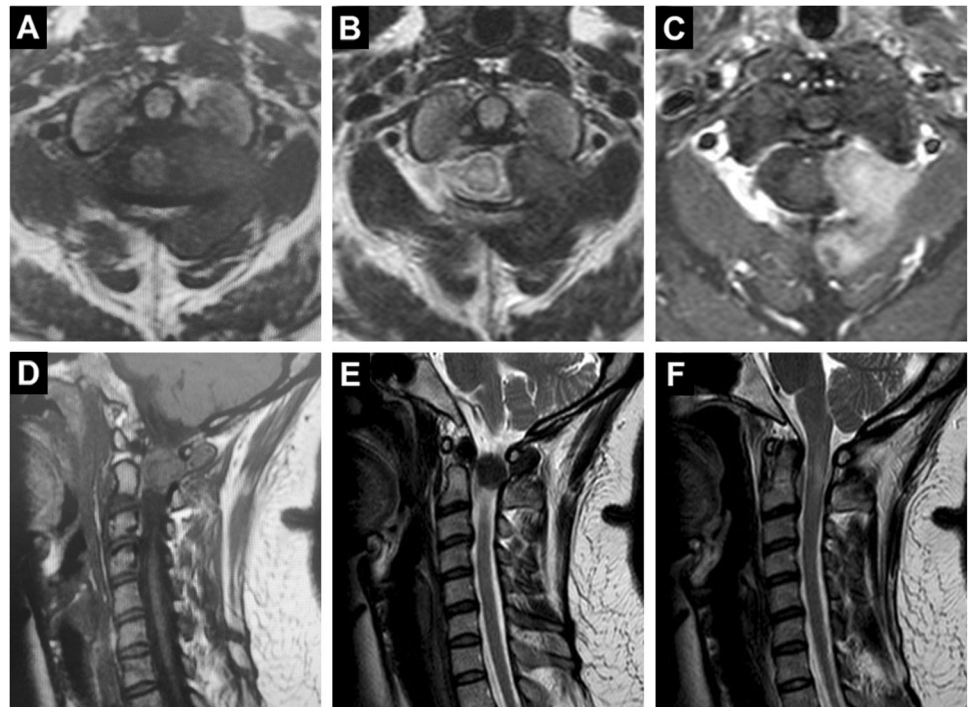
From these findings, the tumor was diagnosed as a cervical spine tumor at C1–C2. Neurinoma, meningioma, neurofibroma, and other tumors were considered in the differential diagnosis.

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**Fig. 1** Magnetic resonance images of the cervical spine showing a mass extending from the left epidural space to the paravertebral muscles at the C1/C2 level. **a** Axial T1-weighted image showing a lesion with heterogeneous isointensity. **b** Axial T2-weighted image showing a lesion with heterogeneous low signal intensity. **c** Axial T1-weighted image after administration of intravenous gadolinium-DTPA (dimeglumine) showing obvious gadolinium-DTPA enhancement. **d** Sagittal T2-weighted image showing an isointense mass at C1/C2. **e, f** Sagittal T2-weighted image showing a mass with low signal intensity and an intramedullary high signal intensity area at C1/C2



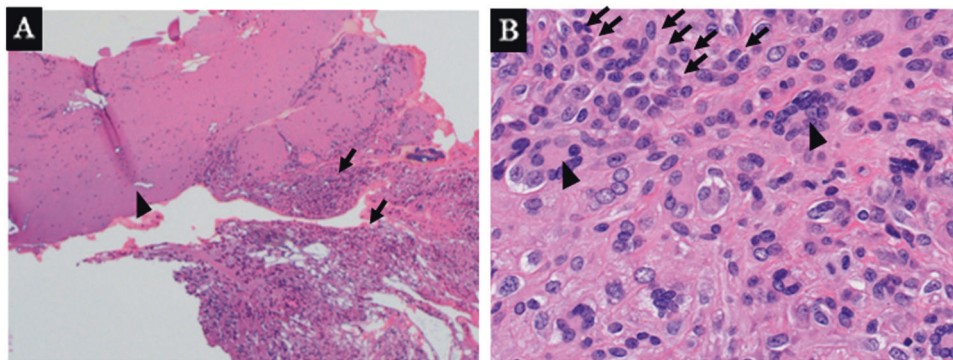
**Fig. 2** Coronal (**a**), sagittal (**b, c**), and axial (**d**) computed tomographic images of the bone window showing no bone erosion

Her neurologic symptoms worsened, and tumor resection was scheduled. We performed laminectomy of the posterior arch at C1 and hemilaminectomy of the left vertebral arch at C2, and achieved total macroscopic tumor resection.

Histopathologically, the resected specimen consisted of both tumor and normal tendon components (Fig. 3a). In the tumor component, hematoxylin and eosin staining showed

proliferation of mononuclear cells and some osteoclast-like multinucleated giant cells (Fig. 3b). Immunohistochemical staining for cluster of differentiation-68 and Factor XIIIa was positive. From these findings, the tumor was diagnosed as TSGCT.

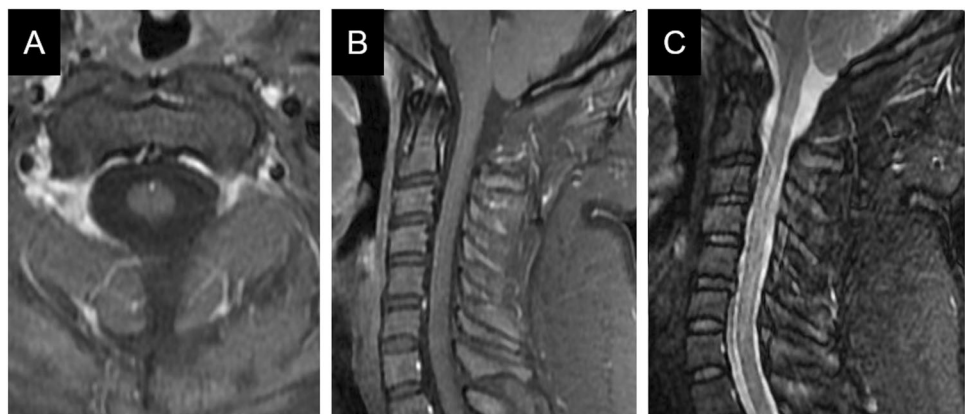
After surgery, her symptoms improved. MRI at 3 years after the surgery revealed no recurrence (Fig. 4).



**Fig. 3** Pathologic results. **a** Photomicrograph of the surgical specimen showing a cellular component composed of small mononuclear cells (arrows) and a normal tendon component (arrowheads) ( $\times 40$ ).

**b** Photomicrograph of the cellular lesion showing proliferative mononuclear cells with round to oval nuclei (arrow). Multinucleated osteoclast-type giant cells (arrowheads) are also seen ( $\times 200$ )

**Fig. 4** Magnetic resonance images of the cervical spine at 3 years after the surgery. Axial (**a**) and sagittal (**b**) T1-weighted images after administration of intravenous gadolinium-DTPA (dimeglumine), and sagittal T2-weighted image (**c**) showing no recurrence of tumor



## Discussion

TSGCT rarely occurs in the spine. The first report of TSGCT of the spine was in 1980 by Kleinman et al. [2], and ~80 cases have since been reported. The cervical spine was the most common site, followed by the lumbar and thoracic spines [3]. TSGCT of the cervical spine usually occurs in the lower cervical vertebrae, and to the best of our knowledge, there have been only 10 cases of this tumor occurring at C1–C2 (Table 1) [3–10]. TSGCT of the spine is considered to arise from the facet joint synovium [11], and often grows extra-articularly and invades the posterior elements of the vertebra [12, 13].

On MRI, TSGCT generally appears as a mass that is isointense on T1WI and hypointense on T2WI. Hypointensity on T2WI reflects hemosiderosis, and Motamedi et al. [12] reported that 83% of TSGCT of the spine are hypointense on T2WI. However, given the variety of components in TSGCT, including hemosiderin, lipid, fibrous tissue, and cellular elements, these tumors tend to show heterogeneous signal intensity [14]. The signal intensity on T2WI is related to the amount of hemosiderin [15–18]. Heterogeneous hypointensity on T2WI may be helpful in the diagnosis of

TSGCT. However, it is difficult to diagnose TSGCT pre-operatively because of its rarity in the spine. CT shows bone erosion in at least 70% of cases of TSGCT of the spine [12]. The mechanism of bone erosion is unknown, but the following hypotheses have been proposed: in one, an increase in intra-articular pressure caused by the tumor compressing the articular cartilage and bony cortex; [19] in another, the synovium deeply invades the bone tissue through the vasculature, leading to bone atrophy [20]. In many of the reported cases occurring in C1–C2, bone erosion was seen in the facet between C1 and C2 or atlantoaxial joint, suggesting that the tumors originated from these sites. However, it has also been reported that there were no obvious bony lesions [7, 8, 10], the origins of the tumors were speculated to be the bursa [7] or vertebra membrane [10] in such cases. In our case, CT showed no bony lesion in the facet or atlantoaxial joint, suggesting that the tumor may have originated from the bursa or vertebra membrane, as in the previously reported cases [7, 10], or from the tendon sheath of the paravertebral muscles.

Gross total resection is recommended for the treatment of TSGCT. The recurrence rate of TSGCT in the spine is 18–25% [3, 21], which is equivalent to that in the limbs

**Table 1** Summary of previous reports of tenosynovial giant cell tumors of the upper cervical spine (C1/C2) [3–10]

Authors	Sex/age	Symptoms	MRI (T2)	Bone erosion	Origin (suspected)	Treatment
Pulitzer and Reed [4]	F/35	Neck pain	?	?	?	GTR
Graham et al. [5]	F/44	Neck pain	High	+	Facet joint	Radiation GTR + fixation (transarticular screw, iliac graft)
Finn et al. [6]	F/82	Quadriparesis	High	+	Atlantoaxial joint	GTR + fixation (transarticular screw, allograft)
Blankenbaker et al. [7]	M/43	No symptoms	Low	-	Bursa	GTR
Teixeira et al. [8]	?/31	Neck pain	?	-	Facet joint	GTR
Wang et al. [3]	F/23	Neck pain	High	+	Facet joint	GTR + fixation (nail plate, iliac graft)
Wang et al. [3]	F/44	Neck pain	Low	+	Facet joint	GTR + fixation (screw rod, iliac graft)
Lavrador et al. [9]	F/64	No symptoms	Low	+	Facet joint	Biopsy
Yamada et al. [10]	F/63	Photopsia	High-Low	-	Vertebra membrane	GTR
Our case	F/32	Neck pain and impaired dexterity	Low	-	Bursa, vertebra membrane, or PVM?	GTR

GTR gross total resection, MRI magnetic resonance imaging, PVM paravertebral muscles

[22, 23]. The recurrence rates after gross total resection and subtotal resection were 6.7% and 66.7%, respectively, and incomplete resection often resulted in recurrence [3]. For a recurrent case, molecularly targeted therapy using imatinib, nilotinib, emactuzumab, and PLX3397, which targets the colony-stimulating factor 1 receptor (overexpressed in TSGCT), may be a useful treatment option [24–29]. In cases of TSGCT affecting C1–C2 with marked bone destruction or intervertebral instability, fixation should be considered in addition to tumor resection (Table 1) [3, 5, 6]. However, we believe that tumor resection alone was adequate in this case because preoperative imaging showed no bone destruction or instability.

TSGCT of the upper cervical spine (C1–C2) is rare. If a tumor is heterogeneously hypointense on T2WI, TSGCT should be considered in the differential diagnosis. We performed tumor resection alone in this case because preoperative imaging showed no bone destruction or instability at C1–C2.

### Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflict of interest.

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