

Case Report

A vertebral extra dural chordoma at C5, possibly deriving from a clival chordoma

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Abstract

Background: Clival chordomas are a rare type of cancer with low metastatic potential and primary metastasize to the lung or bones.**Case Description:** This case report describes a possible metastatic, paravertebral chordoma at level C4-C5 in a patient with a past medical history of a clival chordoma.**Conclusion:** Chordomas are unpredictable and may metastasise.**Key Words:** Clivus, chordoma, metastatic, vertebral**Access this article online****Website:**www.surgicalneurologyint.com**DOI:**

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INTRODUCTION

Deriving from the embryonic remnants of the notochord, chordomas are a rare type of cancer. They account for 1-4% of all primary bone tumors.^[5,10] Chordomas are slow growing, low-grade malignancies that arise from notochord remnants in the vertebrae especially in the sacrum and the skull base. These tumors seldom arise from the cervical or thoracic vertebrae.^[5,6,10] Although chordomas are considered to have a minimal metastatic potential, there are reports of metastatic chordomas in lungs, bones, and lymph nodes.^[4,6] Regrowth or recurrence is most common in the local region or surgical pathway. In this case report, we would like to present a patient with a vertebral chordoma at C4-C5 level after initial resection of clival chordoma and proton beam therapy, 3 years ago.

CASE REPORT

We present a 52-year-old patient with a medical history of clival chordoma. In 2009 the patient underwent a

partial resection of a lower clival chordoma expanding until level C2. Six months after surgery, before proton beam could be realized, a second postoperative magnetic resonance imaging (MRI) showed a regrowth of the remnant. A second resection in 2010 and additional proton beam therapy on the postoperative remnant was performed. In 2012, an intracutaneous chordoma in the medial right neck was surgically resected. Routine MRI showed no tumor growth until 2013, when a mass at level C4-C5, suspected for a recurrent chordoma [Figure 1] appeared.

Neurologic examination revealed a slight weakness of the right deltoid muscle.

Imaging

The MRI shows an extravertebral 3 cm lobular process extending in the foramen of C4/C5. This foramen is slightly wider than normal. The right carotid artery shows no flow on MRI, suggesting an obstruction of this artery. A computed tomography (CT) scan confirmed a widening of the neural foramen.

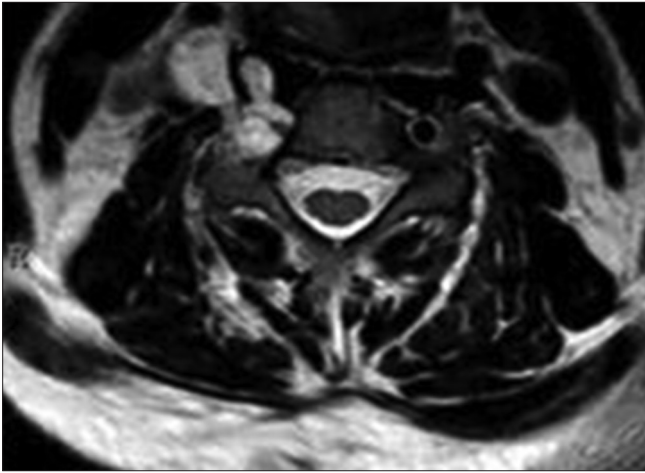


Figure 1: Axial T2-weighted images of the cervical spine showing an irregular lobular mass at C4-C5 partially inside the foramen without spinal cord compression

Surgery

At surgery, the tumor appeared as a grayish, rather solid, encapsulated mass. The tumor infiltrated the neuroforamen of C4-C5 and infiltrated the C5 nerve root. The vertebral artery had a small caliber and was compressed by the tumor. Considering the medical history, the tumor was macroscopically suspected as a chordoma, however, a schwannoma could not be excluded, intraoperative pathological examination could not confirm the diagnosis of a chordoma. Therefore the tumor was incompletely resected avoiding further damage to the C5 nerve root, thereby leaving a remnant in the C4-C5 neural foramen. This remnant was treated with proton beam therapy.

Pathology

The overall conclusion of the histological examination confirmed the diagnosis chordoma.

DISCUSSION

Chordomas arise from remnants of the notochord. The notochord forms during embryologic development, remnants can occur all along the spinal column. Chordomas are rare and account for 1–4% of all bone tumors, 25–35% of them arise from the clivus. Chordomas are known for their capacity to regrow at the primary site and seeding in the surgical pathway, however, they are considered to have a low metastatic potential.^[5,6,13]

Our case is especially rare as the patient previously had a subdermal chordoma, prior to the cervical chordoma. To our knowledge, this is the first paper reporting multiple metastatic chordomas in different tissue types out of the vicinity of the surgical field.

There are a number of possibilities for the occurrence of chordoma in the cervical vertebrae; a new primary tumor or metastatic tumor.

Primary chordomas that originate from the cervical vertebrae account for about 6–7% of all primary chordomas.^[3] A case report by Lim *et al.* describes multiple chordoma along the spinal column wandering if multiple chordomas at different sites along the neuraxis are a sign of a disseminated disease,^[9] this might suggest that our patient had a new primary cervical chordoma. However, this does not explain the appearance of the cutaneous chordoma. It is highly unlikely that the cutaneous tumor was a metastatic tumor and the cervical a new primary, thus suggesting that both of them are metastatic tumors.

Metastasis are uncommon for clival chordomas, as 73% of the metastatic chordomas originate from the sacrococcygeal region, and most frequently appear in lung, liver, lymph node, or bone.^[4-8,14] There are a number of possible pathways for tumor dissemination: Through the lymph system, hematogenous or by liquor transport.^[4,6,7,12] A drop metastasis by the liquor transportation, as described by Uggowitz *et al.*,^[12] is unlikely in this specific case, because the vertebral primary tumor did not infiltrate through the dura and the dura remained intact during the first surgery. More important, the cutaneous tumor cannot be explained by this metastatic pathway. Spreading through the lymph system as described by Jain *et al.*^[7] might explain why the tumor metastasized to the dermis and the cervical vertebrae, however, histological examination of the removed tissue did not show evidence of lymphoid tissue. Leaving hematogenous spreading as the only option, Zemmoura *et al.* explains that hematogenous dissemination by clival chordomas is rare, because the dura at the clivus is very adherent to the periosteum effectively isolating the venous sinus from the tumor and protecting it from tumor expansion toward the venous sinus.^[16] However, in our patient it is the best explanation for both the dermal and cervical chordoma.

The question remains why chordomas recur or metastasize? Can this be explained from a genetic or cellular point of view? Almefty *et al.* suggest that abnormal karyotypes are associated with a poor prognosis and more importantly tumor progression. They found that abnormalities in chromosome 3, 4, 12–14 are associated with an increased likelihood for recurrence.^[11] Sawyer *et al.* found alterations in chromosome 1 and 13 as prognostic markers for tumor progression,^[11] Walter *et al.* state that changes in chromosome 7 might play a role.^[15] A study in cancer-like stem cells by Aydemir *et al.* reports that chordomas possessing cancer stem-like cells, specifically CD133 and CD15, might be important for recurrent and metastatic chordomas.^[2] There does not seem to be a specific genetic tumor marker involved in the prediction of recurrent and metastatic potential of chordomas. Therefore we did not perform genetic research on our patient.

CONCLUSION

This case report underlines the unpredictability of clival chordomas and their possible metastatic potential.

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