Case Report

Intradural spinal seeding metastasis of clival chordoma: a case report

Xiaohai Liu, Mingchu Li, Ge Chen

Department of Neurosurgery, Xuanwu Hospital Capital Medical University, Beijing, China
Correspondence to: Ge Chen. Department of Neurosurgery, Xuanwu Hospital Capital Medical University, Beijing 100053, China.
Email: chengecn@126.com.

Background: Clival chordoma is a locally aggressive tumor originating from remnants of the embryologic notochord. Although clival chordomas account for only 0.2% of all central nervous system tumors, they are characterized by local invasion and destruction, dural invasion, bone erosion, and cranial nerve palsy, and even metastasis.

Case Description: We report a case of a 49-year-old female with an intradural spinal seeding metastasis 16 months after the initial endoscopic endonasal surgery (EES) for a clival chordoma. Gross total resection of tumor in upper clival region was achieved after initial EES and pathology revealed a classic chordoma. After 10 months, follow-up magnetic resonance (MR) showed a recurrence *in situ* and gamma knife was applied. After 16 months, the patient complained of neck pain and MR showed a new lesion in the spinal canal at C1 to C2 level. After craniotomy, the lesion in the spinal canal was totally removed, and pathology confirmed a chordoma with increased proliferative potential. The spinal chordoma might have occurred as a result of intradural spinal seeding metastasis through cerebral spinal fluid the during the initial operation.

Conclusions: Chordomas are not only locally aggressive but also unpredictable and may metastasize through cerebral spinal fluid. Intensive follow-up is of great importance in the long term postoperatively time for clival chordoma patients.

Keywords: Clival chordoma; endoscopic transsphenoidal approach; metastasis; intradural spinal seeding; case report

Submitted Jan 26, 2022. Accepted for publication Apr 12, 2022.
doi: 10.21037/tcr-22-211
View this article at: https://dx.doi.org/10.21037/tcr-22-211

Introduction

Chordomas are rare malignant bone tumors derived from the embryologic remnants of notochord with an incidence of approximately 0.1/100,000/year (1). Chordomas are most common in the sacrococcygeal region (50%), followed by skull base (35%), especially clival region and vertebra (15%) (1). Although chordomas are considered to be slow growing and low-grade malignant, they have metastatic potential with many reports of metastatic chordomas in lungs, bones, and lymph nodes, resulting in challenges in surgical removal of these lesions. Cerebrospinal fluid seeding metastasis is very rare. Here, we report the rare case of a 49-year-old female patient with a recurrence *in situ* and an intradural spinal seeding lesion at C1 to C2 level, suggesting chordomas are unpredictable and may metastasize through cerebrospinal fluid. We present the following case in accordance with the CARE reporting checklist (available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-211/rc).

Case presentation

The patient was a 49-year-old female who presented with gait instability for 3 months. Magnetic resonance imaging (MRI) of the skull base revealed a large lesion with a high isointense signal on both T1- and T2-weighted images located in the upper clival region (*Figure 1*). The lesion was approximately 46 mm × 44 mm × 37 mm, encroaching on

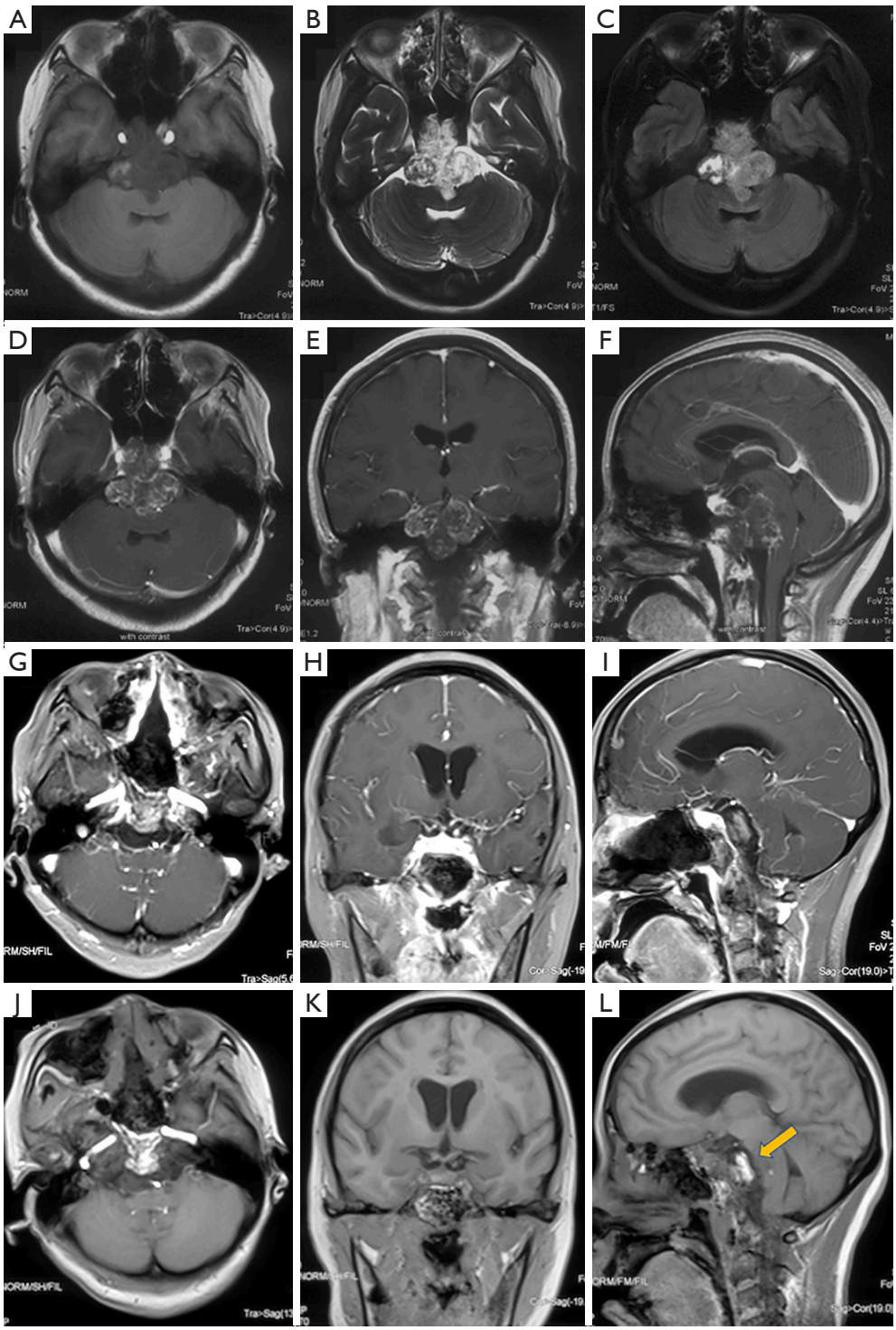

Figure 1 MRI scans of the clival lesion. (A-F) MRI showed a lesion of approximately 46 mm × 44 mm × 37 mm located in the upper clival region. The mass was uniformly enhanced after contrast administration. (G-I) Postoperative enhanced MRI showed total resection of the lesion. (J-L) Postoperative non-enhanced MRI showed fat-packing anterior to the brain stem (arrow). MRI, magnetic resonance imaging.

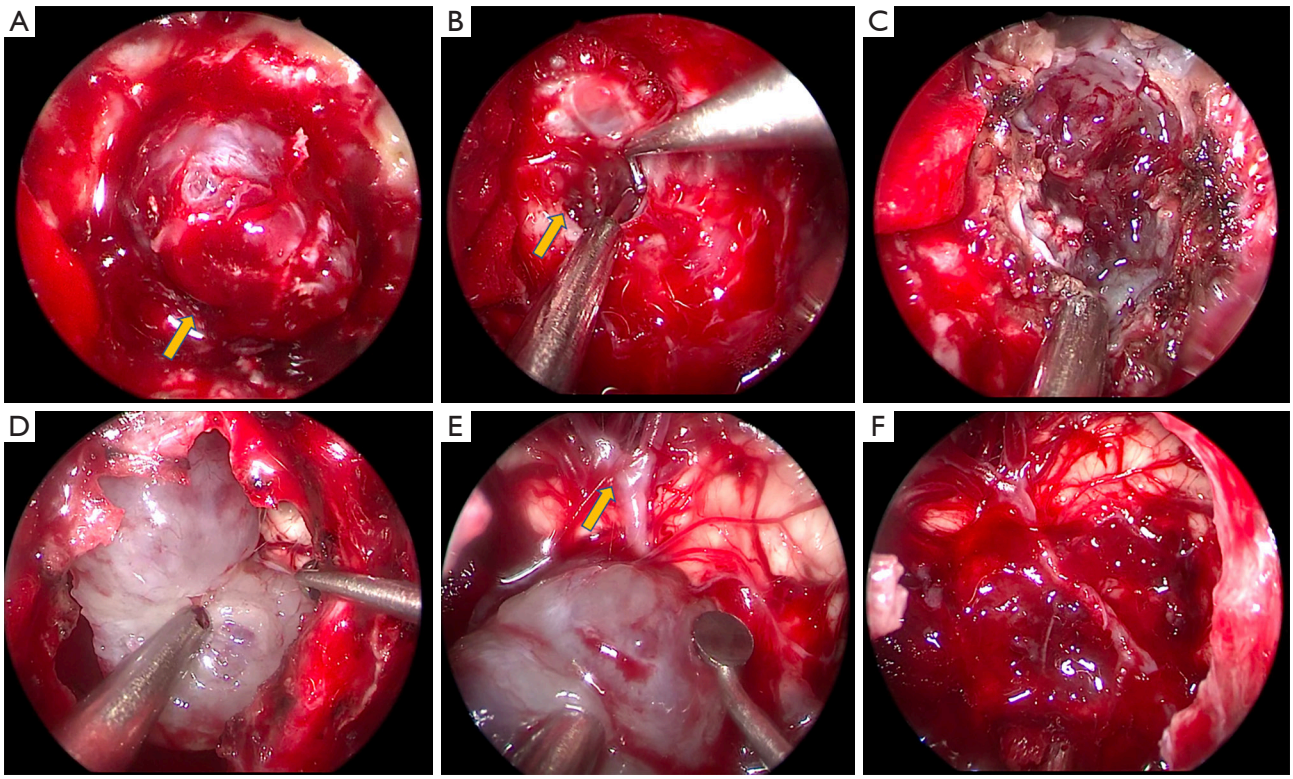

Figure 2 Intraoperative conditions of the lesion. (A) The bone of the clivus was invaded by the tumor (the arrow). (B) The inner layer of the dura was invaded by the tumor (the arrow). (C) After the inner layer of the dura was opened, the tumor was resected. (D) The lesion that adhered to the brain stem was slightly separated. (E) The lesion adhered to the basilar artery (the arrow). (F) The lesion was completely resected and the brain stem and the basilar artery were exposed.

the brain stem. Visual fields of both eyes were relatively normal, and no past cancer history was reported. Pituitary hormonal evaluation and other routine laboratory tests did not show abnormality. Therefore, a clival chordoma was highly suspected, and the endoscopic transsphenoidal surgery was applied. During the surgical operation, the bone of the clivus was found to be invaded by the tumor (*Figure 2A*, the arrow) and the lesion was found to be soft in texture, infiltrating through a dura defect, extending into the intradural space and attaching to the brain stem and the basilar artery. The inner layer of the dura of the clivus was also invaded by the tumor (*Figure 2B*, the arrow). After the inner layer of the dura was opened, a part of the tumor was resected (*Figure 2C*). The tumor adhering to the brain stem and the basilar artery was slightly separated (*Figure 2D,2E*). The lesion was completely resected and the brain stem and the basilar artery were exposed (*Figure 2F*). Postoperative MRI showed the lesion had been totally resected (*Figure 1G-1I*). Postoperative non-enhanced MRI showed fat-packing anterior to the brain stem (*Figure 1J-1L*, arrow). Histopathological results confirmed a chordoma (*Figure 3*), and immunohistochemistry revealed positivity for CK (*Figure 3B*), EMA (*Figure 3C*) and BRACHYURY (*Figure 3D*). The Ki-67 index was 10% (*Figure 3E*). The patient's gait instability was improved after endoscopic surgery.

After 10 months, follow-up MRI showed a recurrence *in situ* (*Figure 4A-4C*) and gamma-knife radiosurgery was applied. After 16 months, the patients complained of neck pain and follow-up imaging showed growth of the recurrent tumor despite treatment by gamma-knife radiosurgery (*Figure 5A-5D*). Moreover, a new lesion was found in the spinal canal at C1 to C2 level in the MR, and a metastatic chordoma with intradural spinal seeding was highly suspected (*Figure 2B-2E*). With midline suboccipital approach, the lesion in the spinal canal was totally removed (*Figure 5E,5F*) and the pathology confirmed a metastatic chordoma (*Figure 3F*), with increasing proliferative potential

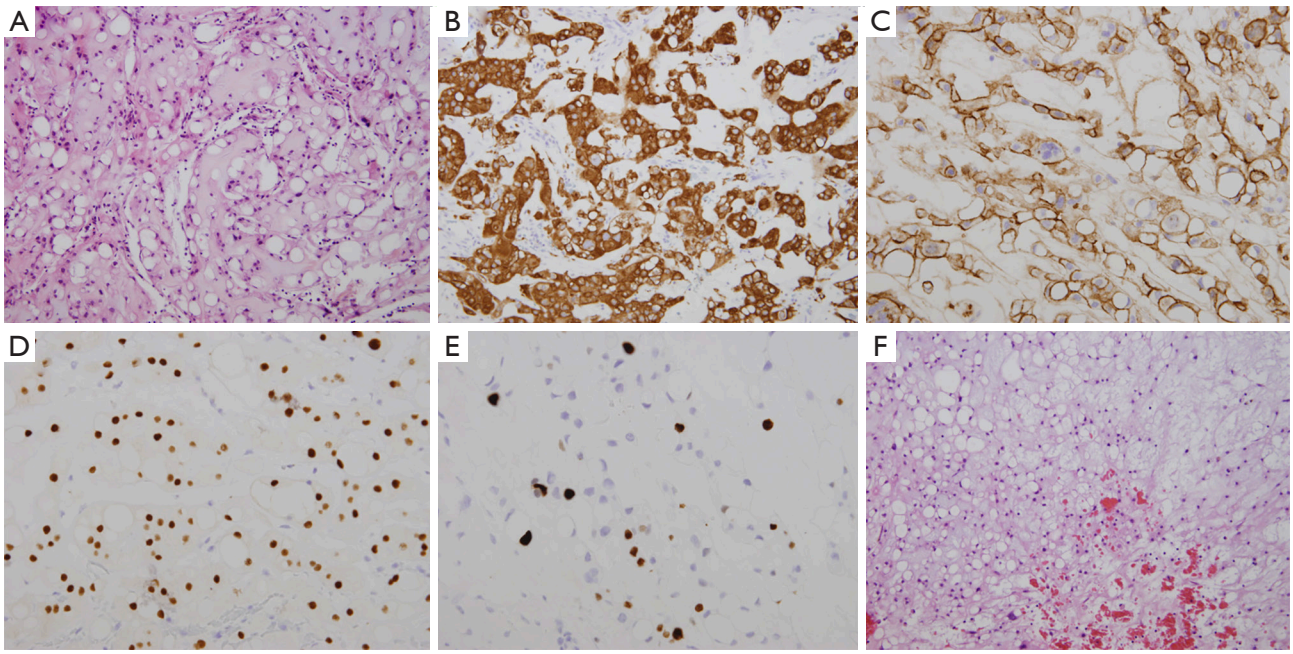

Figure 3 Histological examination revealed the same origin of the two different lesions. (A,F) (×200) H&E staining revealed a chordoma. (B-E) (×400) Immunohistochemistry revealed positivity for CK (B), EMA (C), BRACHYURY (D) and Ki-67 (E). H&E, hematoxylin and eosin.

with a Ki-67 labeling index of 10–15%. As the anlanto-odontoid joint was intact through midline suboccipital approach, the stability of the spine was not destroyed and occipito-cervical fusion was not needed. And there was no new deficits or complications after surgery. The symptom of neck pain disappeared and postoperative MR showed total tumor resection. A 6 months follow-up MR showed total resection and no recurrence in the C1 to C2 level and stable lesion in the clival region (*Figure 5G-5I*). For the pontine lesion that did not respond to gamma knife, the patient refused to have operation and chose to have an MRI scan every 6 months.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

Chordomas arise from the remnants of the notochord, which is formed in the embryonic development process. Therefore, chordomas can occur along the spinal column and most commonly occur in the sacrococcygeal region (50%), followed by the skull base (35%) especially the clival and vertebral regions (15%) (1). Although chordomas are well known for their ability to recur at the primary site, they are thought to have a low metastatic potential, and metastatic chordomas from the clival region are uncommon. According to the literature, metastases is most common to the lungs and in younger patients (2). Other metastatic sites include the liver, lymph nodes, bone, cerebrospinal fluid seeding and surgical pathway seeding (3). To date, very few cases of intraspinal seeding of clival chordoma have been described in the literature. Uggowitzer *et al.* (4) reported a metastatic chordoma from a clival chordoma thorough cerebrospinal fluid seeding and the dura remained intact during the first operation.

En-bloc resection is the recommended treatment which is unfortunately very difficult for clival and spine chordomas, while sacral chordoma instead are those chordoma amenable for *en bloc* resection (5). Currently, the gold standard for chordoma care is complete resection, combined with radiation administered after surgery (6). In the present case, the patient refused to take radiotherapy or proton beam therapy after surgery for some insurance reasons. So, gamma-knife radiosurgery was given when the recurrent

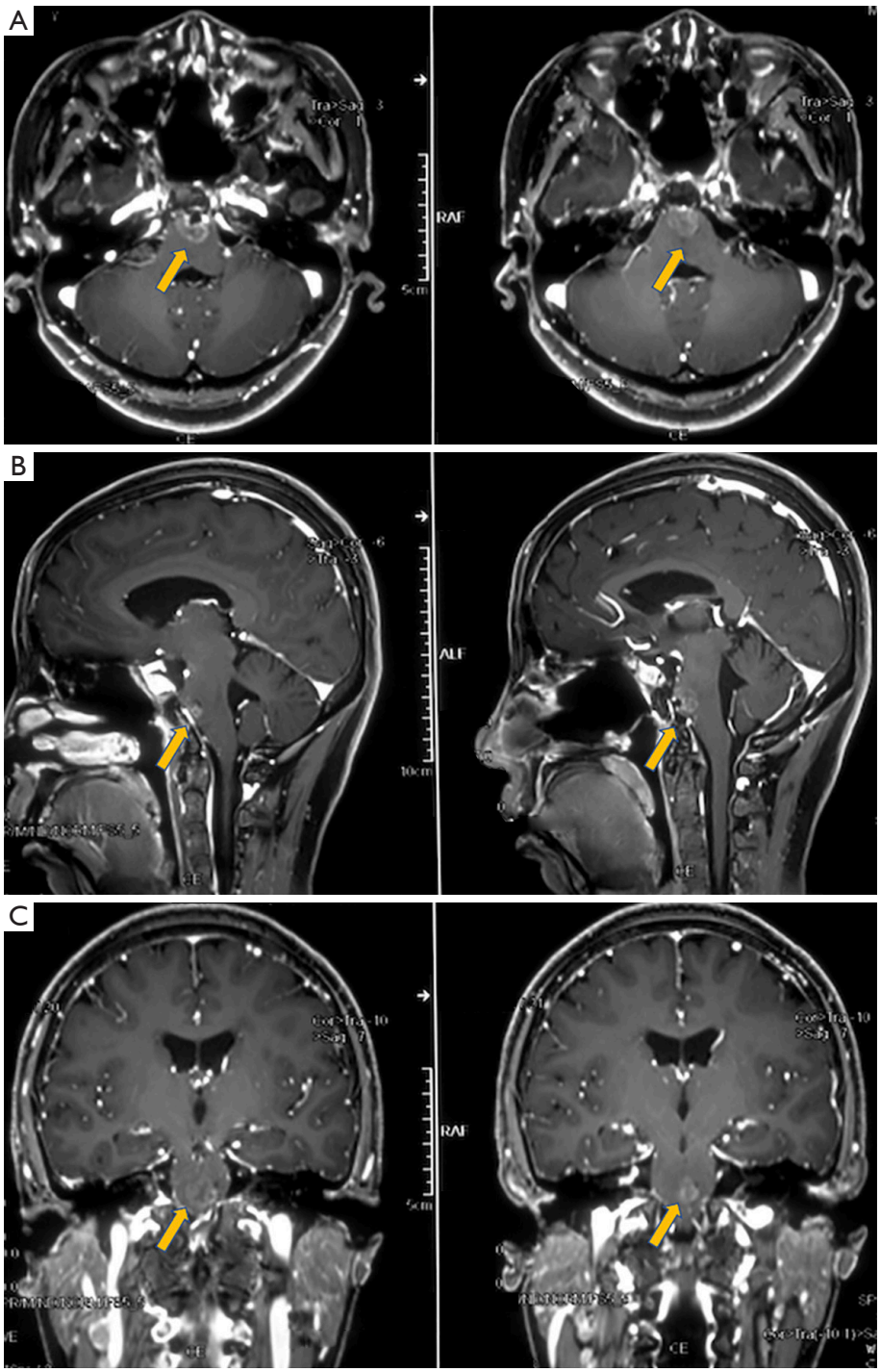

Figure 4 Follow-up MRI of the clival chordoma recurrence *in situ* 10 months after endoscopic surgery. MRI showed a recurrent lesion of approximately 6 mm × 7 mm × 8 mm located *in situ* (arrows) and no lesion was seen in the spinal canal at C1 to C2 level (A, axial plane; B, sagittal plane; C, coronal plane). MRI, magnetic resonance imaging.

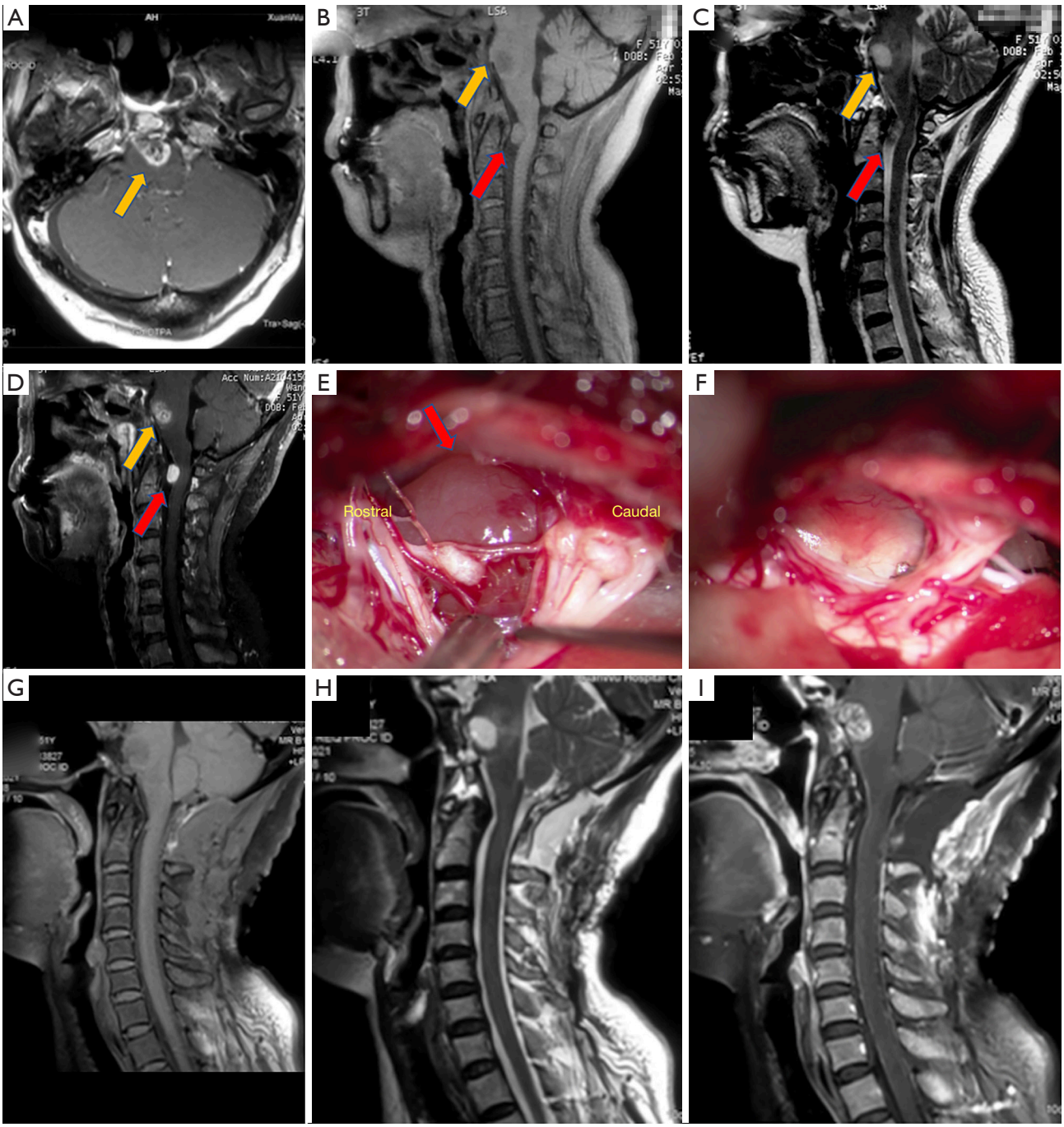

Figure 5 Follow-up MRI of the clival chordoma recurrence *in situ* and new lesion at C1 to C2 level 16 months after endoscopic surgery. (A-D) MRI showed growth of the recurrent lesion *in situ* (the yellow arrow) and a new lesion in the spinal canal at C1 to C2 level (the red arrow). (E) Intraoperative conditions of the intraspinal lesion (the red arrow). (F) The lesion was completely resected. (G-I) Six months follow-up MRI showed the intraspinal lesion was totally removed. MRI, magnetic resonance imaging.

tumor was found *in situ* 10 months after the first operation. However, the recurrent tumor still grew quickly 6 months after the gamma-knife radiosurgery. As the new lesion was found in the spinal canal at C1 to C2 level which was far away from the initial chordoma, intradural spinal seeding metastasis is more likely than intradural or arachnoidal invasion of the clival chordoma. For the intraspinal seeding metastasis in the C1-C2 region, surgical resection was the first choice and the patient still required intensive follow-up. Recently, Passeri *et al.* (7) suggested tumor growth rate (TGR), showing the percentage change in tumor size over 1 month, may be considered as a preoperative radiological indicator of tumor proliferation and seems to preoperatively identify more aggressive tumors with a higher tendency to recurrence.

As the molecular pathogenesis of clival chordomas is still unknown, no chemotherapies or targeted therapies have been developed till now. There are no specific genetic biomarkers involved in predicting the recurrence or metastatic potential of chordomas.

Conclusions

Chordomas are not only locally aggressive but also unpredictable and may metastasize through cerebral spinal fluid. Intensive follow-up is of great importance in the long term postoperatively for clival chordoma patients.

Acknowledgments

Funding: The financial support for this study was provided by Beijing Hospitals Authority Youth Program (code: QMS20210802 to XL). The funding institutions had no role in the design of the study, data collection and analysis, decision to publish, or preparation of the manuscript.

Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-211/rc

Peer Review File: Available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-211/prf

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tcr.amegroups.com/article/view/10.21037/tcr-22-211/coif). XL reports financial support by Beijing Hospitals Authority Youth Program (code: QMS20210802). The other authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

References

1. Erdem E, Angtuaco EC, Van Hemert R, et al. Comprehensive review of intracranial chordoma. Radiographics 2003;23:995-1009.
2. Chambers KJ, Lin DT, Meier J, et al. Incidence and survival patterns of cranial chordoma in the United States. Laryngoscope 2014;124:1097-102.
3. Walcott BP, Nahed BV, Mohyeldin A, et al. Chordoma: current concepts, management, and future directions. Lancet Oncol 2012;13:e69-76.
4. Uggowitzer MM, Kugler C, Groell R, et al. Drop metastases in a patient with a chondroid chordoma of the clivus. Neuroradiology 1999;41:504-7.
5. Yasuda M, Bresson D, Chibbaro S, et al. Chordomas of the skull base and cervical spine: clinical outcomes associated with a multimodal surgical resection combined with proton-beam radiation in 40 patients. Neurosurg Rev 2012;35:171-82; discussion 182-3.
6. Stacchiotti S, Sommer J; Chordoma Global Consensus Group. Building a global consensus approach to chordoma:

a position paper from the medical and patient community. Lancet Oncol 2015;16:e71-83.
7. Passeri T, di Russo P, Champagne PO, et al. Tumor Growth Rate as a New Predictor of Progression-Free Survival After Chordoma Surgery. Neurosurgery 2021;89:291-9.

Cite this article as: Liu X, Li M, Chen G. Intradural spinal seeding metastasis of clival chordoma: a case report. Transl Cancer Res 2022;11(9):3426-3433. doi: 10.21037/tcr-22-211

