

Intradural Invasion of Extradural Clival Chordoma

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Most chordomas show extradural extension and bone destruction. A 32-year-old man presented with neck pain and progressive paraparesis. He had been diagnosed a clival chordoma and underwent an operation seven years ago. Radiological studies revealed that the tumor was recurred in a retroclival area and invaded into intradural region. We removed the tumor by two staged operations. After surgery, satisfactory results were achieved.

KEY WORDS : Chordoma · Clivus · Intradural · Extradural.

Introduction

Chordomas are rare neoplasm that arise from the remnants of the primitive notochord^{1-8,10-12}. Their incidence among primary intracranial brain tumors is 0.5%⁴. Peak prevalence of chordomas is in the 4th decade of life, but less than 5% of patients diagnosed in the age of less than 20 years². Approximately, 50% of all chordomas arise in the sacrococcygeal region, and 35% arise in the skull base, where they typically involve the clivus. The remaining 15% occur in the midline along the path of notochord, primarily involving the cervical vertebrae^{8,10}. Generally chordoma is thought to be essentially extradural in nature and is associated with extradural extension and bone destruction⁸. However, if left to grow for long periods, it may invade the dura and extend intradurally as well as extradurally^{4,8}.

We report a patient with intradural invasion of extradural clival chordoma with special emphasis of its local invasiveness.

Case Report

History

A 32-year-old man was admitted to our hospital with the complaints

of progressive paraparesis and posterior neck pain. In the past history, the patient had been diagnosed as the clival chordoma and had undergone a subtotal removal seven years ago at other hospital. After operation the patient was lost on follow-up visit. Recently symptom was developed then he visited to our hospital.

Preoperative examination

Neurologic examination revealed that motor power of lower extremities were four-fifth strength. The power of shoulder elevation and elbow flexion were slightly decreased. Mild spasticity of lower extremities, increased deep tendon reflex, and ankle clonus were evident. Magnetic resonance image(MRI)



Fig. 1. Initial sagittal T1 and T2-weighted magnetic resonance images (A, B) show a well capsulated retroclival mass. Brain stem and upper cervical cord are compressed. Gadolinium enhanced axial (C) T1 weighted image reveals heterogeneous enhancement and intradural invasion of mass (arrow).

• Received : January 17, 2005 • Accepted : May 6, 2005

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showed a huge retroclival mass, which was located from foramen magnum to C4 level, and upper cervical cord was compressed severely. Noticeably, the mass invaded intradural region from C2 to C4 level. The mass showed homogenous and isointense signal intensity on T1 and T2 weighted sagittal images (Fig. 1A, B), and heterogeneously enhanced after intravenous infusion of gadolinium (Fig. 1C).

Operation

We have planned two staged operations. The right far lateral transcondylar approach was chosen in the first operation for removal extradural located chordoma. In a park-bench position, skin incision and muscle dissection were done down to the C4 level with a caution of possible injury to vertebral



Fig. 2. Sagittal T1 weighted gadolinium enhanced magnetic resonance image demonstrates a intradural mass (arrow), which remained after the first operation. Upper cervical cord is compressed.



Fig. 3. Tongue and mandible are cut off, and C3 body is removed during second operation.

artery. The mastoid process was removed until whole sigmoid sinus was exposed. Then C1 and C2 arch were removed. Occipital condyle was removed partially. The tumor extended from the anterior portion of foramen magnum to C4 body level. The mass was soft, friable, and grayish, which was found to compress the brain stem and easily

aspirated by suction. On the right side of the patient, the 5th to 11th cranial nerves could be identified by this approach. A small area of the tumor was attached to the dura in the C2 and C4 area. We could remove extradural chordoma. Second operation was performed one month later. We chose transoral approach for the removal of intradural chordoma (Fig. 2). The operation was done with cooperation of an otolaryngologist. After tracheostomy, tongue and mandible was split. Inspection and palpitation of oral cavity identified the body of C2-4. A longitudinal midline incision was made in the posterior pharyngeal mucosa. Corpectomy of the C3 body was done (Fig. 3). Dura was incised. The mass was located from the cervicomedullary junction and extending down to the upper margin of the C4 level, which was resected totally.

Histological finding

Histological examination of the lesion revealed a lobular pattern caused by the insertion of thick strands of fibrous connective

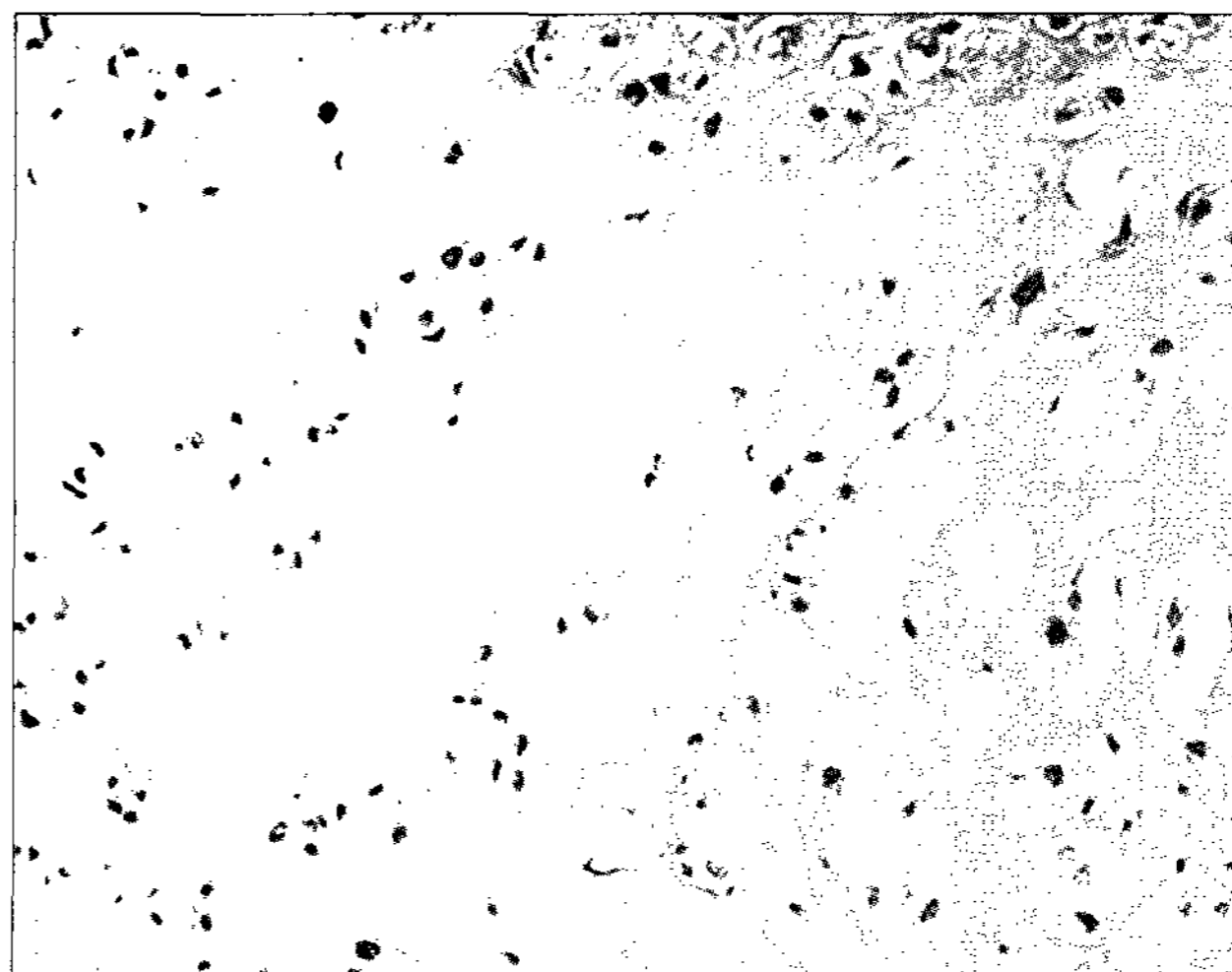


Fig. 4. Photomicrograph of the surgical specimen shows lobules composed of physaliphorous cells on a chondroid background. (H&E×200 original magnification).

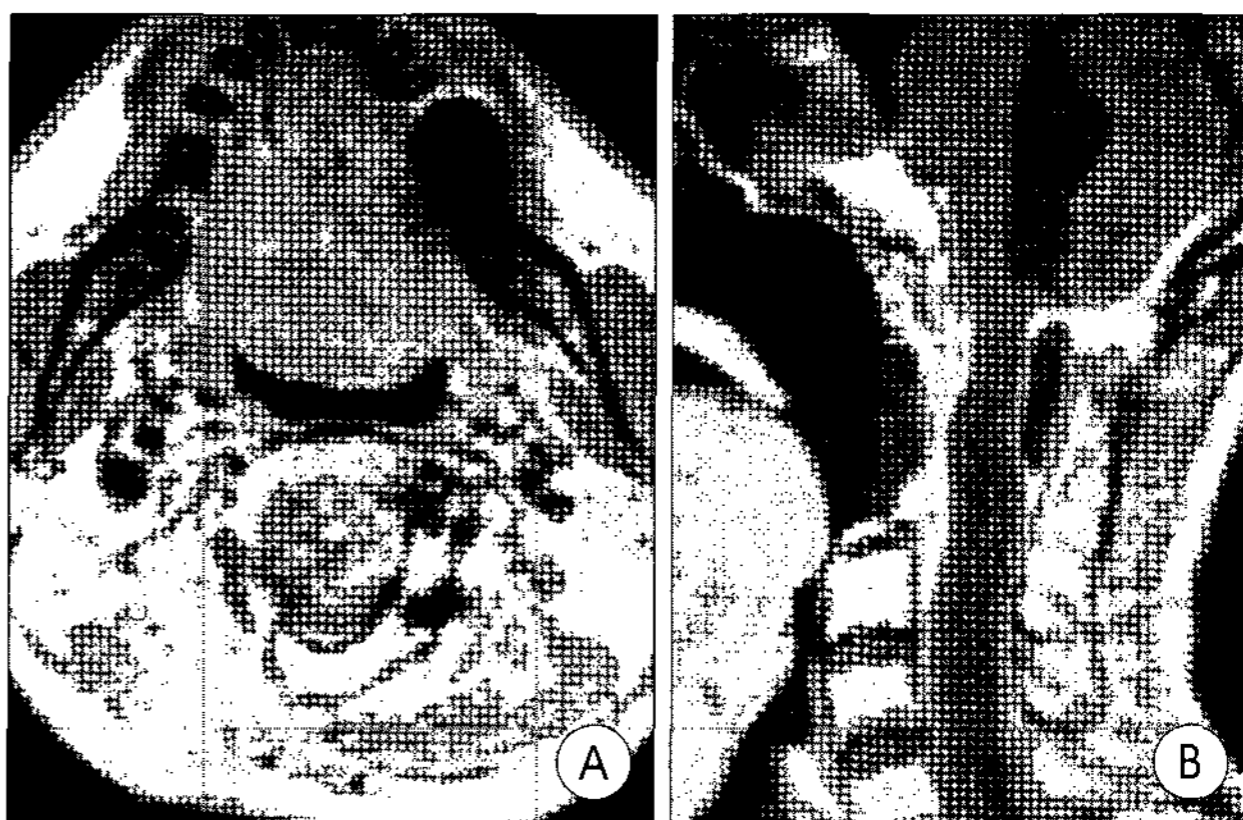


Fig. 5. Postoperative gadolinium enhanced T1 weighted axial(A) and sagittal(B) magnetic resonance images show the extent of tumor resection.

tissue. Physaliphorous cells were recognized (Fig. 4). The histological diagnosis was confirmed as a chondroid chordoma.

Post operative course

After surgery (Fig. 5), there is no evidence of CSF leakage. We perform gastrostomy for protection posterior pharyngeal mucosa. One months later, The G-tube was expelled. The patient could walk independently and discharged.

Discussion

Chordoma is believed to be originated from remnants of the primitive notochord^{1-8,10-12}, which occurs a predilection for the clivus and the sacrum. This anatomical distribution supports the generally accepted theory that these tumors arise from such notochordal remnants.

As these rests have an intraosseous localization, chordomas are usually extradural and cause local bone destruction^{3,12}. So, purely intradural chordoma is very rare^{1-8,10-12}.

The pure intradural chordoma is known to show a slower growth pattern, sharply circumscribed margins that allow total excision, and it never metastasizes¹². The prognosis of intradural chordomas may be better than extradural chordomas because there is a lack of intraosseous invasion, although long-term follow-up results have not been reported^{4,6}.

Intradural tumor extension, which is occasionally encountered, might be caused by rupture of the dura as a result of tumor compression. The dura serves as a strong barrier against tumor extension into the intradural space. So, it is important not to tear or open the dura, when the tumor is removed extradurally⁹.

Jallo et al.⁶ proposed the classification of vertebral chordoma to serve as a better predictor of the prognosis. In their system, type I chordomas are osseous extradural (the majority), type II are extraosseous extradural, type III are osseous intradural and type IV chordomas are extraosseous intradural. In their system, chordomas are separated into four categories based on the space they occupy as well as the presence/absence of an osseous connection. Type II and IV lesion are thought to have a better prognosis because complete resection can be more easily achieved^{6,10}. The extradural chordoma that invade intradural region is excluded in their system. Therefore, their system must be amended. Namely extradural and intradural chordoma should be included a new category.

The recurrence rate of clival chordomas seems to be high even using advanced microsurgical and skull base techniques. The difficulties in radical removal of clival chordomas may be mainly due to their invasiveness of the submucous layer and extension into the loose connective tissue in multilayers or multilobular fashion⁹. For radical removal, Oikawa et al.⁹

insisted that it will be necessary to removed not only the invaded bone and submucous layer but also normal bone at initial surgery. Radiologically, chordomas are best evaluated by using both computed tomography scanning(CT) and MRI⁷. CT is considered better in demonstrating tumoral calcification and defining the exact anatomy of bony destruction⁵. After intravenous injection of contrast medium, chordomas usually enhance homogenously or heterogenously. On MRI, they give slightly to moderately low signal on T1 weighted images, and heterogenous and homegenous high signal on T2 weighted images⁷.

Most patients with skull base chordomas have been treated with a combination of surgery and radiotherapy, although the efficacy of radiotherapy is controversial^{4,8}. Stereotactic radiosurgery is valuable and effective as a primary or adjuvant treatment for selected patients with chordomas. Long-term follow-up of these patients is necessary to confirm the effectiveness of this therapeutic modality^{4,8}.

Conclusion

We describe a case of intraosseous extradural chordoma that invade intradural region. Long-term follow up and research are needed to obtain accurate prognosis and effectiveness of treatment modalities of extra and intra-dural chordomas.

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