Recurrent Axis Chordoma after Carbon ion Therapy Necessitating Reconstructive Surgery with Osteocutaneous Radial Forearm Free Flap: A Case Report

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Abstract

Chordomas originate from remnant tissue of the notochord during embryonic development, with a relatively low incidence rate. Furthermore, chordomas, being resistant to radiotherapy, are primarily treated by resection; however, in some cases, particularly in those that involve the skull base or upper cervical spine, chordomas are unresectable. In recent years, carbon ion/proton beam therapy has shown significant efficacy in such cases. However, it is not sufficiently curative and is commonly associated with recurrence. Moreover, there is no consensus regarding the treatment of recurrent cases, resulting in several uncertainties pertaining to it. Here, we present the case of a 55-year-old male patient with axial chordoma who experienced recurrence after carbon ion therapy and underwent tumor reduction surgery for a longer life span. Two months postoperatively, dehiscence was found in the posterior pharyngeal wall, probably due to heavy ion therapy. Salvage surgery was performed using an osteocutaneous radial forearm free flap, and the patient's postoperative course was uneventful.

Keywords
- recurrent chordoma
- axis chordoma
- carbon ion therapy

Chordomas originate from remnant tissue of the notochord during embryonic development, and has an incidence rate of approximately 0.088 cases per 100,000 persons.[1–3] Chordomas are resistant to radiotherapy,[4] and resection is the mainstay of treatment.[5] However, they occur in approximately equal proportions in the skull base, spine, and sacroiliac region, and not a few cases are unresectable.[1,2,6,7] In recent years, carbon ion/proton beam therapy has been employed to treat unresectable chordomas with partial efficacy.[8,9] Despite its potential as a new treatment option for such cases, recurrence still occurs, and there is no consensus regarding its usage. Reduction surgery is an option; however, its associated risks remain unknown. Previous reports on spine surgery after radiotherapy[10,11] have stated that increased complications can be expected with particle therapy, thus complicating decision-making processes.

We present a case of an unresectable axial chordoma that was initially treated with carbon ion therapy, but later recurred. The tumor grew slowly and compressed the

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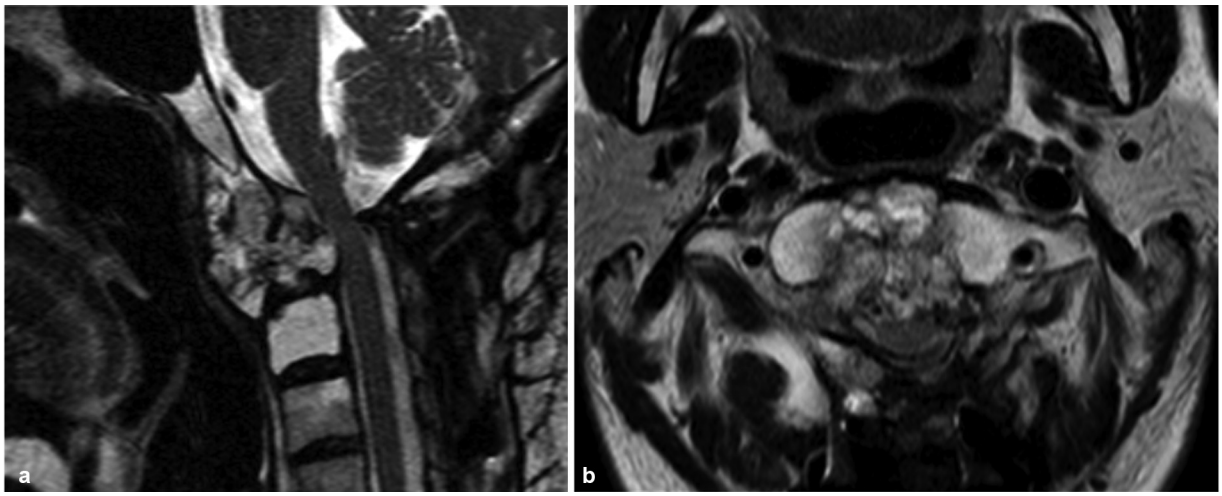

Fig. 1 Preoperative magnetic resonance imaging (MRI) scans revealing gradual progression of the tumor. (**a**) Sagittal section. (**b**) Axial section.

cervical spinal cord. We decided to perform tumor reduction surgery with iliac bone grafting to prolong the patient's life. Postoperatively, there was necrosis/defect in the mucosa of the posterior pharyngeal wall that required reconstructive surgery with an osteocutaneous radial forearm free flap. The patient's postoperative course was uneventful.

Our literature search revealed very few reports of surgical resection of recurrent cervical chordomas after carbon ion therapy, making this case valuable. Here, we report our experience with this case, along with a review of the literature.

Case Report

The patient was a 55-year-old man who had been diagnosed with axial chordoma 7 years previously. Because of the location of the tumor, this case was deemed unresectable, and carbon ion therapy (60.8 Gy/16 Fr) was planned and performed. The following year, he underwent posterior cervical fusion (O–C5) for cervical spine instability. Unfortunately, 4 years later, the tumor recurred. Although tumor recurrence was confirmed, owing to the difficult treatment approach, only follow-up was performed, which allowed the tumor to grow slowly and continuously, leading to compression of the cervical spinal cord (►**Fig. 1**). To extend the patient's life, tumor reduction surgery was planned.

An intraoral approach with a midline mandibulotomy was performed by a head and neck surgeon and an oral and maxillofacial surgeon. An orthopaedic surgeon incised the posterior wall of the pharynx and performed tumor reduction and free iliac bone transplantation. As the blood flow in the mucosa of the posterior wall of the pharynx appeared acceptable, and indocyanine green angiography showed rich perfusion, only a simple suture was performed at closure (►**Fig. 2**). However, the postoperative adhesion of the posterior pharyngeal wall was not favorable, leading to the development of a mucosal defect 2 months postoperatively (►**Fig. 3**). Based on these results, a reconstructive surgery was deemed necessary.

A reconstructive surgery was performed using an osteocutaneous radial forearm free flap (►**Fig. 4**). The radius was

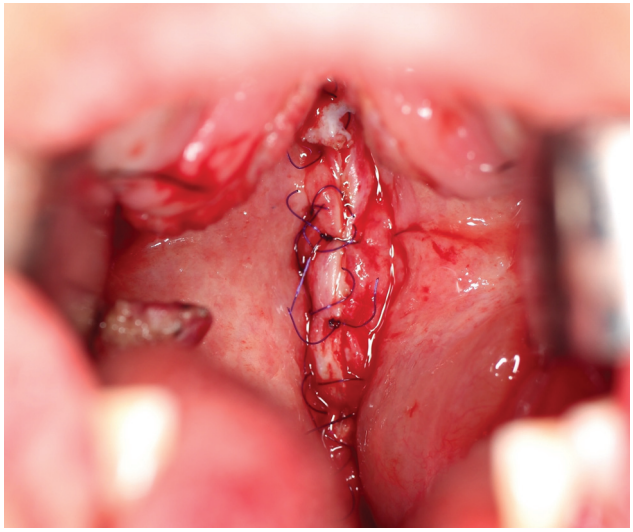

Fig. 2 The blood flow in the mucosa of the posterior wall of the pharynx appeared acceptable.

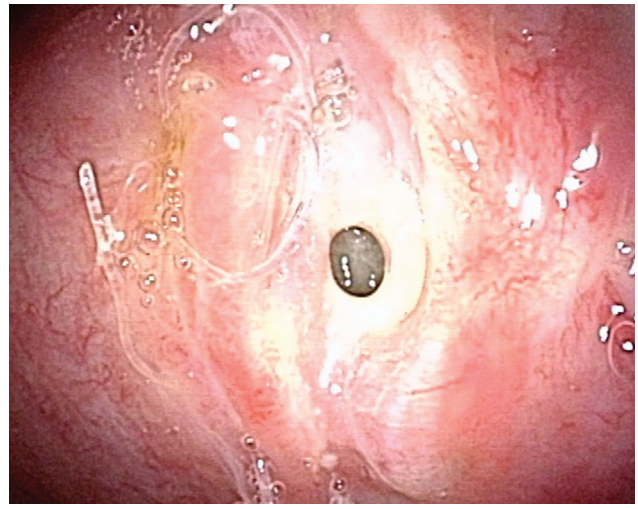

Fig. 3 Mucosal defect detected 2 months postoperatively.

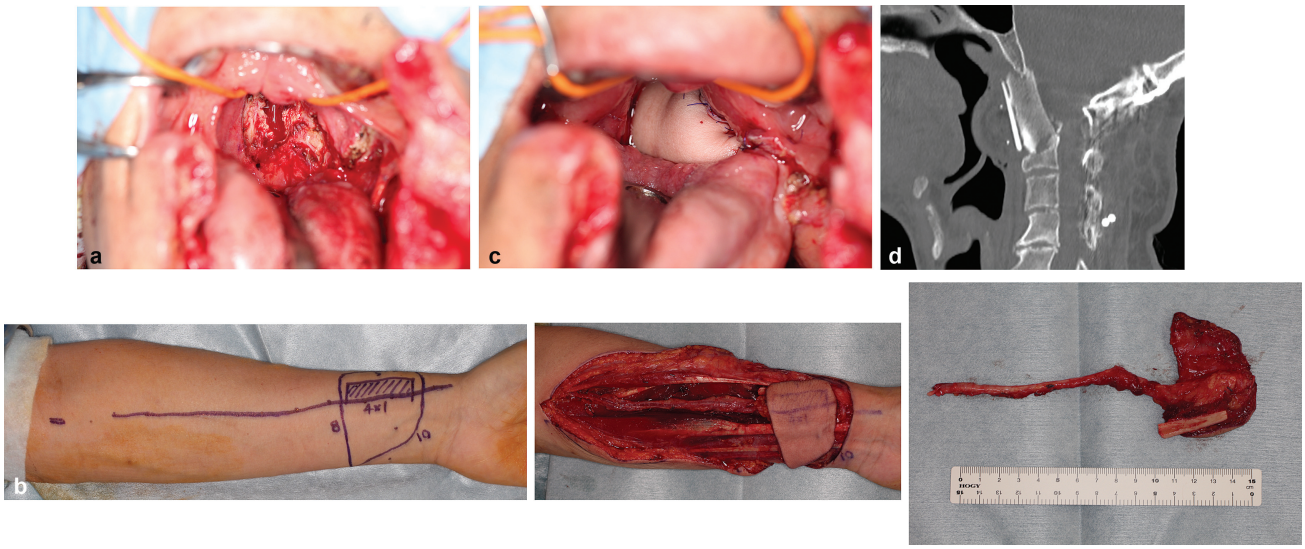

Fig. 4 (**a**) Pharyngeal posterior wall following debridement. (**b**) Radial forearm flap. (**c**) Suture of the osteocutaneous flap. (**d**) Computed tomography (CT) scan at 1 month postoperatively.

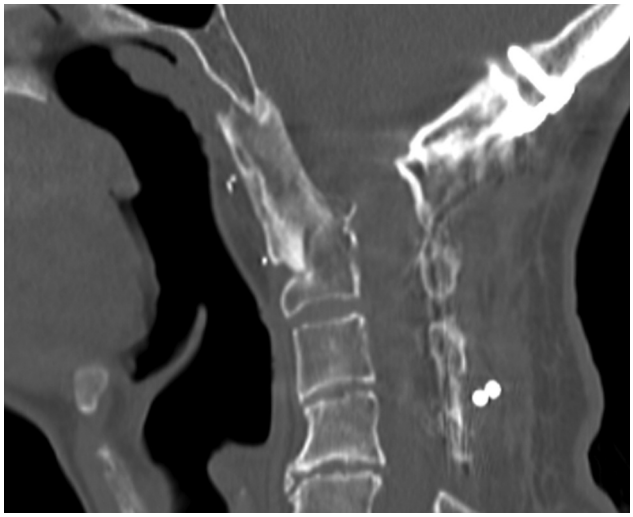

Fig. 5 Computed tomography (CT) at 1 year postoperatively shows fusion between the grafted radius, iliac bone, and surrounding bone.

placed in front of the free iliac bone, which was non-necrotic, and the posterior wall of the pharynx was reconstructed using a skin paddle. The radial artery was anastomosed to the left superior thyroid artery and the venae of the radial artery were anastomosed to the left facial vein. End-to-end anastomoses were performed in both sites. Oral intake of food was allowed after 2 weeks postoperatively, and the patient was discharged after 6 weeks. One-year postoperative computed tomography (CT) images showed fusion between the grafted radius, iliac bone, and surrounding bone (►**Fig. 5**).

Discussion

Carbon ion/proton beam therapy has been used for unresectable chordomas in recent years.[8,9] However, as it is a relatively new treatment modality, there are few reports based on its surgical use for recurrent chordomas, and many aspects are still unknown. As complications are known to increase during spinal surgery after radiotherapy,[10,11] further rise in complications can be expected with heavy ion/proton beam therapy, and we hope that more cases will be reported in future.

Baig Mirza et al[12] reviewed reports of surgery for chordomas in the spine and found that various approaches were employed depending on the tumor location. For upper cervical spine cases, Zhou et al[13] used a high retropharyngeal or transoral/transmandibular approach,[14] while Hsieh et al[15] used a submandibular or transmandibular approach. The present case involving an upper cervical tumor was treated using a transmandibular approach, which aligns with previous studies and seems to be a reasonable choice.

Since the transplanted iliac bone may have been necrotic, we used osteocutaneous flap for surgery. However, the iliac bone was not necrotic in this case, and we believe that cutaneous flap would have been sufficient. What makes this case novel is that the surgery was performed after heavy ion therapy, as, to our knowledge, there are very few reports of salvage surgery after this therapy. A meta-analysis by Akinduro et al[16] included 161 patients with cervical chordoma, with only three undergoing surgeries after proton beam therapy and none after heavy ion therapy. Matsumoto et al[17] reported three cases of recurrent high cervical spine malignancies after particle therapy, with one involving proton beam therapy and two involving carbon ion therapy. To our knowledge, the only reported case of surgery after heavy ion therapy was reported by Matsumoto et al. Among two of Matsumoto et al's cases after carbon ion therapy, one patient showed deep wound infection and cerebrospinal leakage, which developed into a defect of the mucosa of the posterior pharyngeal wall, and the other patient also showed breakdown of the posterior pharyngeal wall, which required reconstruction with a pectoralis major myocutaneous flap. Based on these cases, Matsumoto et al suggested that myocutaneous flap reconstruction should be considered during

salvage surgery after particle therapy. Our results are consistent with these findings. In addition, these results suggest that recurrence after carbon ion therapy can be salvaged by reconstructive surgery using several flaps, offering additional treatment options in the future.

Conclusion

A patient who underwent salvage surgery for recurrent axial vertebral chordoma after carbon ion therapy showed necrosis of the mucosa of the posterior wall of the pharynx, indicative of the effects of heavy ion therapy. However, the patient's life could be saved with reconstructive surgery using an osteocutaneous radial forearm free flap. These results suggest that reconstruction using a cutaneous/myocutaneous flap should be considered in salvage surgery following heavy ion therapy.

Ethical Statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflict of Interest

None declared.

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