

Case Report

Chordomas Involving Multiple Neuraxial Bones

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We present a patient with multifocal symptomatic osseous chordomas having unusual growth patterns with review of the pertinent literature. The patient was 62-year-old male and had multiple osseous chordomas located in sacral, thoracic, and paraclival jugular foramen areas. There was no metastasis in other organs. All affected sites were osseous. The multicentric chordomas are extremely rare. This case could be considered as a chordoma involving multiple neuraxial bones. But, the possibility of multicentricity could also be thought. In such cases radical resection should be performed for each lesion at the initial diagnosis. If complete surgical resections are infeasible or impossible, preoperative or postoperative radiation therapy should be planned for the highest possibility of successful treatment.

KEY WORDS : Chordoma · Multiple · Thoracic · Sacral · Paraclival · Jugular foramen.

INTRODUCTION

Chordomas are tumors of the axial skeleton, arising from primitive notochord remnants, and constitute 2% to 4% of all primary bone tumors^{3,4,7}. They are slow-growing, locally invasive, rare tumors of the spine. Multicentric chordomas are especially rare. To our knowledge, only one case of multicentric chordomas has been reported¹. We present a patient with multifocal symptomatic osseous chordomas having unusual growth patterns with review of the pertinent literatures. The possibility of multicentricity is also discussed.

CASE REPORT

A 62-year-old male patient had a palpable mass on the sacrococcygeal area. The protruded part of the palpable mass on the skin was surgically removed at the private hospital and it was pathologically diagnosed as a chordoma. We could not get the data about the pathology and image study. Postoperatively, the patient was given a course of radiation therapy, with a total dose of 6400 cGy. The

patient made a full recovery without any symptoms following the radiation therapy. In January 2004, 20 months later, the patient developed persistent headaches and left-side tongue atrophy. Brain magnetic resonance imaging (MRI) showed a tumorous lesion in left side of the clivus, extending to the left jugular foramen. T1WI images showed the tumor to be hypointense. Following intravenous administration of gadolinium, moderate to marked heterogeneous enhancement was observed. Subsequent T2WI images showed the tumor to appear hyperintense (Fig. 1). It was diagnosed as a glomus jugulare tumor and surgery was performed with a Gamma Knife without pathologic confirmation. We designed conformal 50% isodose line to cover 99% of the tumor. The prescription dose was 26 Gy and the mean margin isodose was 13 Gy. Tongue atrophy persisted and the patient began to experience radiating leg pain in the right dermatome, progressively worsening one year later. The initial X-rays of the lumbosacral spine showed an osteolytic change in the right sacroiliac area. MRI showed multiple tumorous lesions in the thoracic and lumbosacral areas. The patient was then evaluated with abdominopelvic computed tomography (CT), chest CT, whole body bone scan, and positron emission tomography (18-FDG) whole body scan for possibility of metastasis. There was no metastatic evidence in other regions. The lumbosacral tumor involved the iliac bones on both sides, forming a soft mass which

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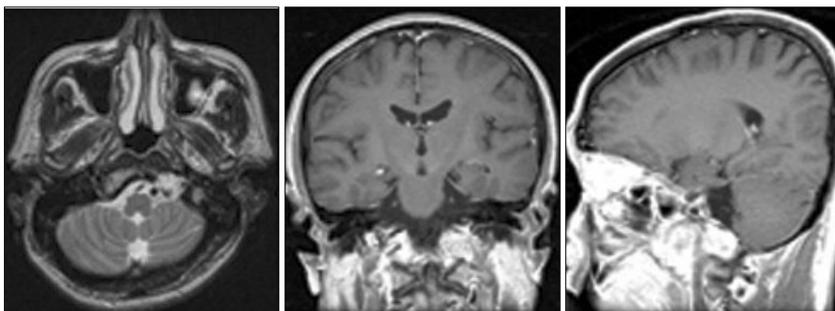


Fig. 1. Chordoma in the left jugular foramen, extending to the left side of the clivus.

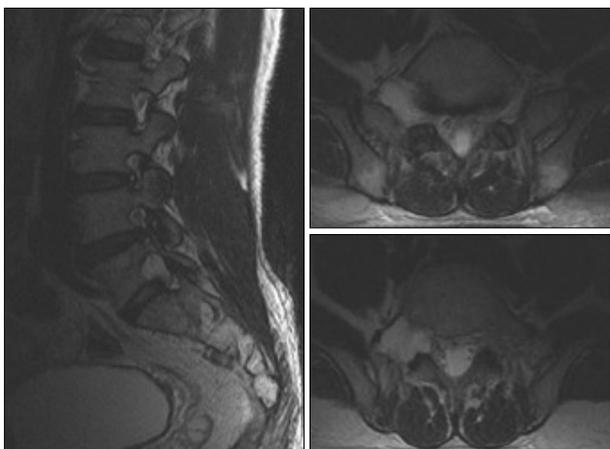


Fig. 2. Chordoma in the sacral area involving iliac bones on both sides, forming a soft mass pinching the spinal nerve root.

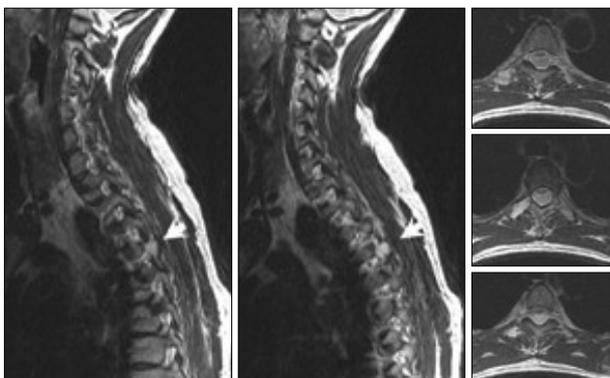


Fig. 3. Chordoma in the thoracic area involving the inferior articular process and lamina of T3 vertebra on the right side, with a soft mass in the paraspinal area extending to an adjacent proximal part of the 3rd rib (arrow).

was pinching a spinal nerve root (Fig. 2). The thoracic tumor involved the inferior articular process and lamina of T3 vertebra on the right side with a soft mass in the paraspinal area and extended to an adjacent proximal part of the 3rd rib (Fig. 3). We performed surgery for the sacral and thoracic tumors to confirm pathology. The thoracic lesion was totally removed and the soft mass of the sacral lesion pinching the spinal nerve root was also removed. The masses were grossly lobulated, grayish, and soft with a

gelatinous consistency. Microscopic examination showed physaliphorous cells with myxoid stroma. The immunohistochemical staining for cytokeratin (DAKO, AE1/AE3, 1/300) and epithelial membrane antigen (DAKO, E29, 1/100) showed positive reaction. However, S-100 protein (DAKO, polyclonal, 1/1000) and carcinoembryonic antigen (DAKO, II-7, 1/50) showed negative

reaction. Both lesions were pathologically confirmed as chordomas (Fig. 4). Immediately following surgery, the radiating leg pain disappeared and the patient had fully recovered without any evidence of local recurrence. The patient had been doing well until post-operative 2 years follow-up.

DISCUSSION

Chordomas are rare bone tumors that occur in the midline of the axial skeleton. They account for only 1% to 4% of primary bone tumors^{3,7}. More than 50% of these lesions are located in the sacrococcygeal region, 35% are located in the clival and cervical area, and the remainders are spread throughout the vertebral column, with a distribution of 61.1% in the lumbar, 11% in the thoracic and 27.8% in the cervical spine^{2,3,5-7}.

This case involved three discrete chordomas located in the clival, thoracic, and sacral areas respectively. However, the clival tumor was not pathologically confirmed. Because the clival lesion was identified prior to the sacral and thoracic lesions, it was thought to be a glomus jugulare tumor. In light of the subsequent pathologic confirmations, the clival lesion was much more likely to be a chordoma rather than a glomus jugulare tumor.

The radiographic differential diagnoses include metastatic disease, and primary or secondary spine tumor, such as multiple myeloma, chondrosarcoma, neurogenic tumor, tumor-like condition such as Paget's disease. Chordomas behave as malignant entities because of their local invasiveness, high recurrence rate, and potential for metastasis. Regions of metastasis include the lymph nodes, lung, liver, brain, or bone, and the reported incidence of metastasis varies from 5% to 43%. To detect the metastasis, whole body bone scan, chest CT, abdominopelvic CT, PET scan should be done. In this case, the patient did not show any evidence of metastasis from these studies.

Chordomas usually show a radiating infiltrative pattern involving epidural space and prevertebral and lateral

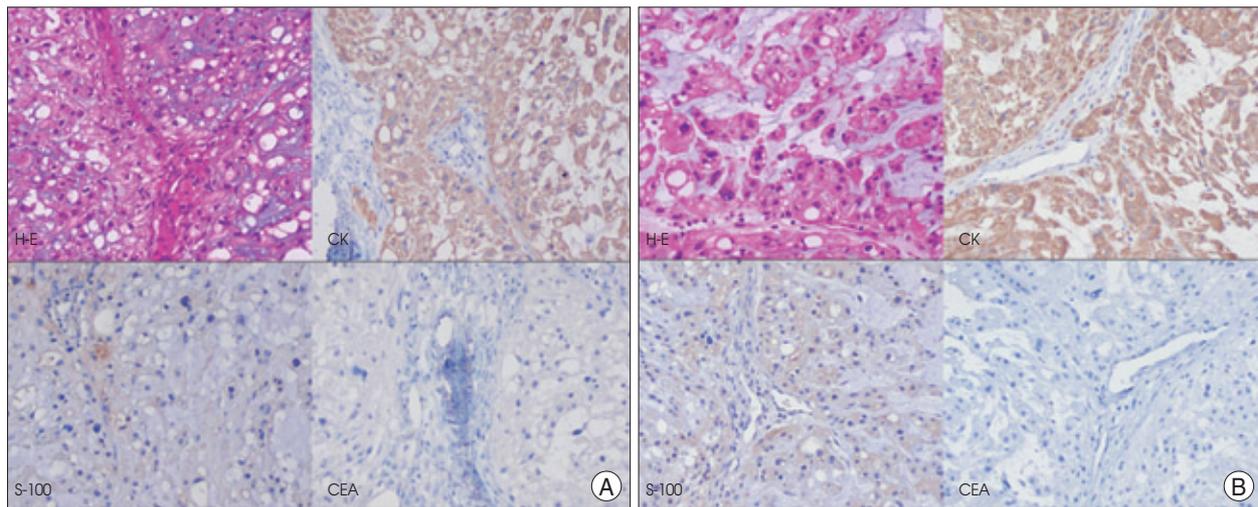


Fig. 4. Specimens had been obtained from sacral lesion (A) and thoracic lesion (B). Physaliphorous cells are seen in the tissue with small, dark, round to ovoid nuclei and abundant multivacuolated cytoplasm in both lesions. The immunohistochemical staining for Cytokeratin and EMA show positive reaction. However, S-100 protein and CEA show negative reaction. These findings are consistent with chordoma.

paravertebral regions. They can cause dural compression, meningeal sheath invasion, and cerebrospinal fluid spreading. Furthermore, metastasis along the neuraxis is extremely rare.

Chordomas sometimes occur with multicentric origins, but it is extremely rare for them to occur in different sites along the neuraxis, especially in posterior elements of spine. Vertebral body is usually affected. The disks are usually spared and the posterior elements are less involved. There are no specific criteria differentiating multicentric from metastatic tumors. In this case, the patient had tumors in both sacral and thoracic regions at presentation. All affected sites were osseous. Chordoma had been pathologically confirmed in both regions. There was no metastatic evidence that was confirmed by imaging studies. If metastatic spread of chordomas along the neuraxis occurs, there should be widely disseminated disease. Therefore, each lesion could be independent neoplasms^{2,7}.

The lesion in the left side of clivus was thought to be chordoma after surgeries for sacral and thoracic lesions. Even though the possibility of chondrosarcoma could not be excluded, there was no extensive bony erosion and soft mass formation in clival lesion. Furthermore, clival lesion showed bright signal with lobulation in T2WI and isointensity in T1WI. These findings are consistent with chordoma. Because follow-up MR had no interval change in size and the patient had no progression of symptom associated with the clival lesion, we did not try to get tissue confirmation.

Surgical “en bloc” resection with wide margins is the treatment of choice for chordomas the first time they are diagnosed. Chordomas have a well known tendency to recur locally. The recurrence rate seems to be related to incomplete resection. There is no consensus regarding the efficacy of

chemotherapy and radiotherapy to treat chordomas. Although chordomas are relatively radioresistant, adjuvant radiotherapy has been used for surgically inaccessible lesions, contaminated surgical margins, when incomplete surgical excision of the tumor occurs, and to improve the quality of life and relieve pain in patients with wide spread recurrences^{8,9}. In our case, further evaluation of the clival lesion to clarify surgical resectability may be needed, and adjuvant radiation therapy may be needed in the sacral and thoracic areas. The sacral lesion was not completely resected and radiation therapy seems to be more successful in controlling microscopic diseases.

CONCLUSION

Chordomas are slow-growing tumors that tend to recur locally and metastasize. Chordomas rarely occur multicentrically. We could not prove multicentricity in this case. This case could be regarded as a chordoma involving multiple neuraxial bones. But, the possibility of multicentricity could also be thought. In such cases, radical resection should be performed for each lesion at the initial diagnosis. If complete surgical resections are infeasible or impossible, preoperative or postoperative radiation therapy should be planned for the highest possibility of successful treatment.

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