CASE REPORT

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Plasmacytoma to the Axis Mimicking Metastatic Paraganglioma: Circumferential Reconstruction via Posterior Approach

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Plasmacytoma is a malignant plasma cell tumor growing within soft tissue or the axial skeleton. Here, we present the case of a patient with plasmacytoma of the axis vertebra who underwent decompressive surgery with reconstruction via a posterior approach. The patient was referred because of quadriparesis with severe neck pain. Magnetic resonance imaging revealed a relatively demarcated, highly enhanced mass lesion in a destructed axis, with spinal cord compression. Computed tomography revealed a 5.6×4.3 cm adrenal mass at the left retroperitoneal space. We suspected the axis lesion to be a metastatic paraganglioma from the adrenal mass. The patient underwent total excision of the tumor under an operative microscope with occipitocervical fixation. Histopathologically, the tumor was shown to be a plasmacytoma. Following the operation, the patient recovered without significant complications. This was a rare case of plasmacytoma in the axis, mimicking metastatic paraganglioma.

Key Words: Plasmacytoma · Axis · Paraganglioma · Posterior surgery · Fusion

INTRODUCTION

Plasmacytoma is a clonal proliferative disorder of neoplastic plasma cells and is biologically malignant[4]. Plasmacytoma is rare that responsible for only 5% of plasma cell neoplasms. Plasmacytoma is further classified into solitary plasmacytoma of bone (SPB), extramedullary plasmacytoma (EP), and multiple solitary plasmacytomas that are either primary or recurrent. Among the three, SPB is the most common and occurs as lytic lesions within the axial skeleton[3]. EP mostly occurs in the upper respiratory tract (85%), as do the multiple solitary plasmacytomas. In the latter case, either multiple solitary bone or soft tissue lesions must also be present. In almost all cases, radiotherapy is the main choice for curative treatment because plasmacytomas are radiosensitive. However, in some cases, like with this patient, surgical intervention is required. We have discovered that out of the multiple vertebrae affected by plasmacytoma, only the axis presented a symptomatic compressive myelopathy. Here, we describe this case of plasmacytoma at the axis, which was treated by decompressive tumor removal and reconstruction via a posterior approach.

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CASE REPORT

A 72-year-old man was admitted to our institute complaining of severe neck pain and quadriparesis without any significant history of trauma. Plain radiographs showed atlantoaxial instability with osteolytic lesions on the C2 (Fig. 1). Magnetic resonance imaging (MRI) showed a well-enhanced soft tissue mass (5.9×5.5×5.8 cm) involving the whole compartment of C2 with cortical destruction. The epidural mass extended from the C1 to C4 vertebra at the ventral portion of the dura and from the C2 to C3 vertebra at the posterior portion (Fig. 2). The spinal cord was compressed at the C2 vertebra by the mass and showed slightly increased T2 signal change, suggesting compressive myelopathy. The bilateral vertebral arteries were involved at the C2 level, but retained intact distal flow. Computed tomography (CT) revealed a 5.6×4.3 cm adrenal mass at the left retroperitoneal space (Fig. 3). Subsequently, the patient underwent an F-18 FDG whole body PET/CT examination, which revealed a hypermetabolic soft tissue mass (SUVmax=5.4) involving the C2 body, lamina, and spinous process. The adrenal mass was mildly hypermetabolic (SUVmax=2.8), which ordinarily suggests a benign tumor;

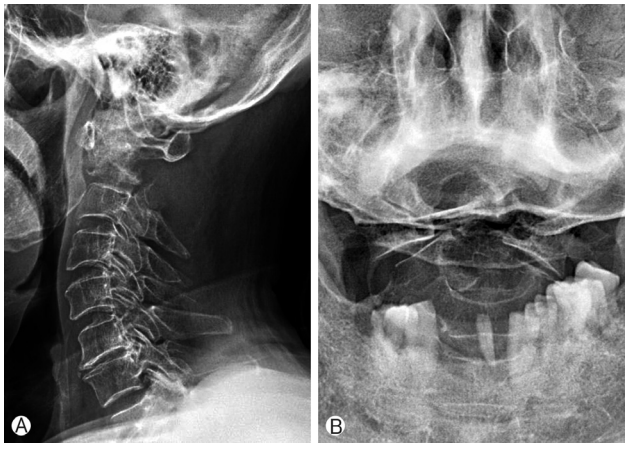

Fig. 1. Plain radiograph showing osteolysis of the axis with malaligment. (A) A lateral cervical plain radiograph, (B) An open mouth radiograph.

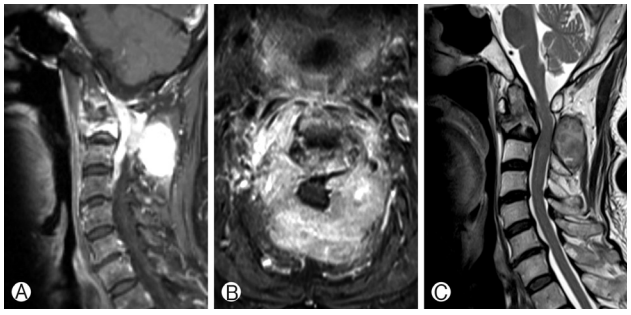

Fig. 2. A mass lesion extending along the anterior epidural space with cord compression at preoperative MRI. (A) Contrast-enhanced sagittal MRI, (B) Contrast-enhanced axial MRI, (C) T2 sagittal MRI.

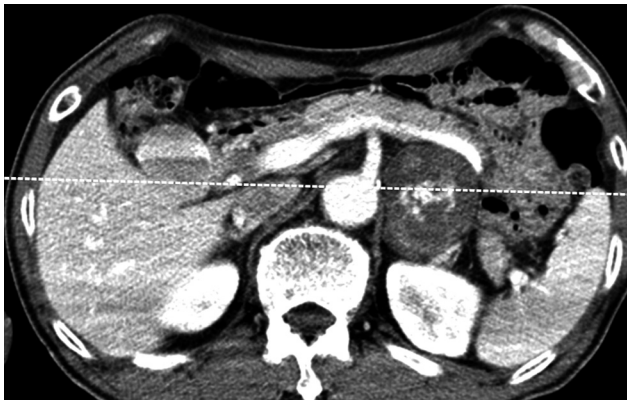

Fig. 3. An abdominal PET/CT image showing a 5.6×4.3 cm size mass at the left retroperitoneum.

however, the possibility of malignancy remains.

At first, we suspected the axis lesion to be a metastatic paraganglioma from the adrenal mass. We requested a needle biopsy from a skeletal radiologist; however, the radiologist recommended an open biopsy and surgery because of the hypervascularity of the tumor and the possibility of neural injury by the needling. On preoperative day 1, trans-femoral spinal angiography was performed, which revealed that the axis mass was fed from both the vertebral and deep cervical arteries. Feeder embolization was done by embolization particles. Under intra-operative neurophysiological monitoring, removal of the tumor was performed with a Cavitron Ultrasonic Surgical Aspirator (CUSA). After C1 lateral mass screw (posterior arch entry) and lateral mass screw fixation at C3 to C5 was applied, the axis tumor's lateral mass and pedicle was removed by CUSA. The tumor compressing spinal cord was grossly removed from the posterior to anterior poles; however, the bony structure around the left C2 pedicle was severely damaged. Therefore, we reconstructed the atlantoaxial joint with a Harms Cage, a surgical titanium mesh cage, filled with allograft cancellous bone chips. After the occipital plate placement, occipitocervical fixation was performed using a polyaxial screw-rod system with multiple rods (Fig. 4).

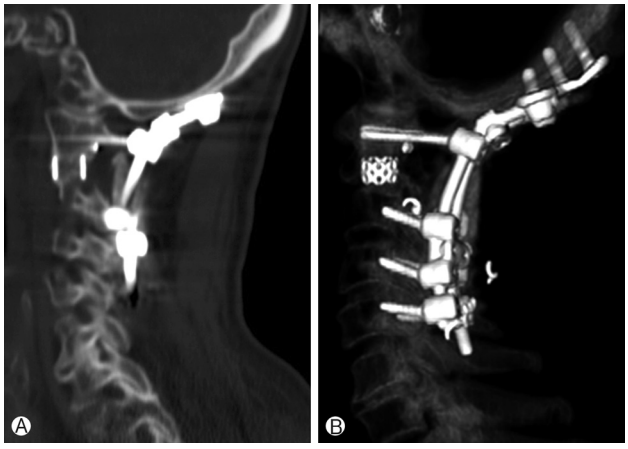

Fig. 4. Postoperative 12 months later, a CT image showing bony bridge formation through the implanted mesh. (A) Sagittal CT image, (B) Postoperative CT 3D reconstruction image.

The patient's postoperative course was uneventful. A frozen biopsy examination revealed that the tumor tissue was more likely to be metastatic paraganglioma. Nevertheless, the final diagnosis of the tumor was reported as plasmacytoma, following immunohistochemistry (IHC) analysis. The IHC staining of the neoplastic plasma cells revealed immunoglobulin kappa chain and CD138 & CD56 & MUM-1 positive expression, and ki-67 positive expression in 3-5% of cells. Bone marrow aspiration biopsy was performed when the plasmacytoma was diagnosed, and normocellular marrow with a slight increase of plasma cells (3.6% of nucleated cell count) was observed. Immediate postoperative MRI showed appropriate decompression of the cervical spinal cord. Follow-up CT revealed suc-

cessful atlantoaxial joint fusion evidenced by bony bridge formation through the implanted mesh. Adjuvant chemotherapy is being administered for myelomatosis, with alkeran and prednisolone to prevent the disease from spreading and to control the remnant enhancing lesion extending to the C4 ventral epidural space.

DISCUSSION

Plasmacytoma of the cervical spine is likely to be misdiagnosed as cervical degenerative disease because of the similar clinical features and the sometimes negative findings of plain radiography. In this case, radiography showed expansile, irregular osteolytic lesions, and vertebral instability, without any periosteal reaction. If CT or MRI was conducted in the early period of the disease, the lesions can be detected early.

It is difficult to distinguish plasmacytoma from other osteolytic tumors from radiological results alone. To distinguish it from other diseases, a percutaneous biopsy of the spine is needed, if possible under the guidance of fluoroscopy or CT in consideration of this procedure's risks. A final diagnosis still depends mainly on pathological examination, especially IHC and immunofixation electrophoresis. Monoclonality and/or an aberrant plasma cell phenotype should be demonstrated. Useful markers include CD19, CD56, CD27, CD117, and cyclinD1[10,12]. In our patient, a left adrenal mass was discovered and diagnosed as either paraganglioma or pheochromocytoma. Therefore, before the operation, we considered the mass to be metastatic paraganglioma. As a result of the frozen biopsy, paraganglioma was suspected, but after the IHC examination, which was performed 7 days after surgery, plasmacytoma was diagnosed.

Although radiotherapy is the treatment of choice for plasmacytoma of the spine, the evidence of its efficacy has been mainly based on small retrospective studies[1,3,5-9,11,13,14]. Some patients suffer from tumor invasion into the spinal canal, causing extremity dysfunction or paraplegia as a result of spinal cord or nerve root compression. In these conditions, most spine surgeons believe that radiotherapy cannot be a substitute for surgery. Although surgery (partial or complete resection and radiotherapy versus radiotherapy alone) did not influence the 10-year probability of local control, it aims at relieving spinal cord and nerve root compression through excision of the tumor and reconstruction of spinal stability. We believe that the potential surgical outcome frequently outweighs the risk of surgery. In this case, the axis mass was safely removed without spinal cord or cervical root injury and with adequate stabilization using a polyaxial screw-rod system with multiple rods.

The general prognosis of SBP is comparatively better than plasmacytoma, with a 5-year survival rate of about 70% and median overall survival period of 7.5 to 12 years[2,3]. Patients, especially those with plasmacytoma of the spine, will probably eventually develop multiple myeloma (MM) in the future, with a median delay of 2 to 4 years. There is no effective method to prevent plasmacytoma from progressing to MM, and there is no consensus in the literature about these adverse prognostic features.

CONCLUSION

We presented a rare case of plasmacytoma of the axis, mimicking metastatic paraganglioma, and treated by a posterior decompressive surgery with reconstruction using a polyaxial screw-multiple rod system with a Hams cage.

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