Case report

Cervical spine chordoma of C3, C4 and C5

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Abstract

Chordoma, which is a rare bone tumor, originates from the embryonic notochord. The most common sites are the spine and skull base. It is a low grade, but malignant tumor that is located mostly in the sacrococcygeal region of the spine. Involvement of the cervical, thoracic, and lumbar spine is an unusual site for chordoma in general neurosurgical practice, occurring in only 15% of cases. Surgery should be performed as the first treatment option. Even if the entire tumor has been resected, it frequently recurs locally. Distant metastases are uncommon. We present a 53-year-old woman with retropharyngeal C3, C4 and C5 mass. She presented with long-standing swallowing difficulty and neck pain. Surgery was performed with the anterior cervical approach and the final pathologic diagnosis was chordoma. We present the case with detailed clinical course, radiological findings, and pathological findings. Complete surgical resection is the first treatment option. Although it metastasizes less, local recurrence is common. In surgical treatment, gross total resection should be applied and radiotherapy should be given if residual is considered.

Key words: cervical spine, chordoma, neurogenic tumors

Introduction

Chordoma is a rare, low-grade, malignant bone tumor. Although it is seen twice as often in men than in women, it is often seen over the age of 40, but it is observed at a younger age at the skull base. It is located in the sacrococcygeal region in 40–50% of patients. Involvement of the cervical, thoracic, and lumbar spine is uncommon, occurring in only 15% of cases [1, 2]. This locally invasive neoplasm grows slowly and tends to recur after treatment. The first treatment option in this tumor is surgery as wide excision should be made as possible and then radiotherapy should be applied. Treatment outcome depends on the size and location site of the chordoma. Although the total mass is resected, the recurrence is unfortunately frequent and increases inversely with the gross total size and location of the primary mass. Distant metastases are not very common. It is frequently seen in the lungs and spine. Median survival is 6.3 years. Early diagnosis of vertebral chordoma is difficult due to the slow growth character and nonspecific initial symptoms. Neck pain is usually the reason why patients attend the hospital and the lesion can be easily seen on computed tomography (CT) or magnetic resonance imaging (MRI), which is requested in case of suspicion. Secondary swallowing difficulties and respiratory disturbance occur when anterior pressure is present. If the

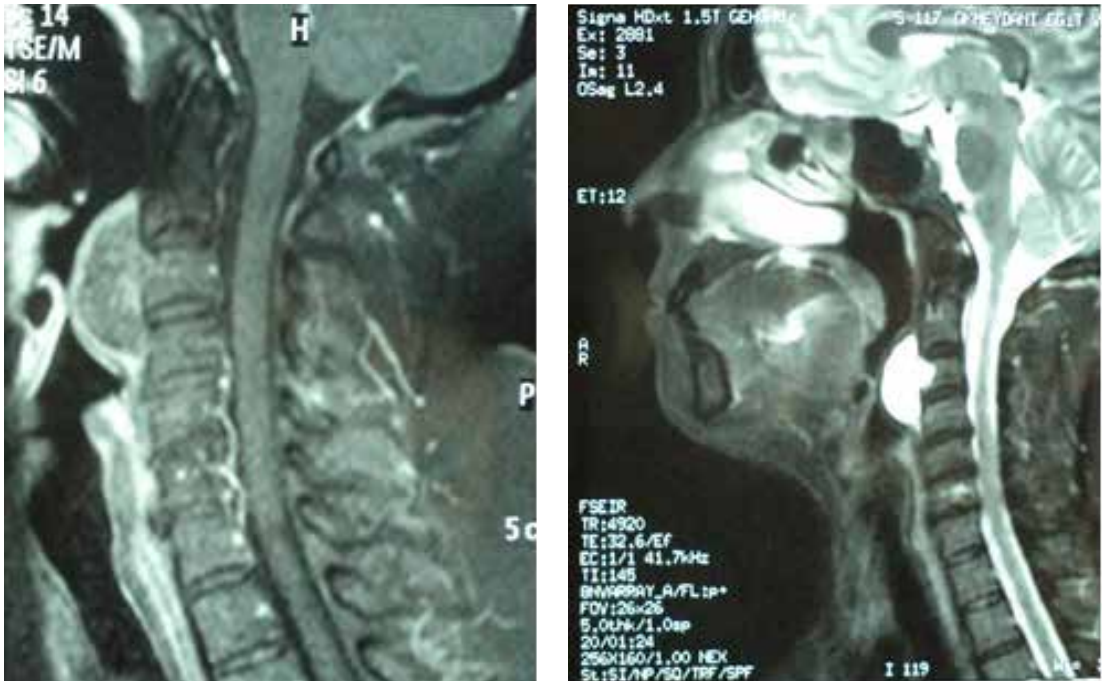

Figure 1. T1 and T2 weighted gadolinium-enhanced sagittal MRI image reveals a rim-enhancing retropharyngeal mass

posterior structures are compressed, myelopathy or radiculopathy is possible [3–7].

Chordoma of the cervical spine is relatively infrequent, only a few cases have been reported in the literature. We present C3-C4-C5 cervical spinal chordoma and in this case, the patient came to our clinic with dysphagia and neck pain. Accordingly, it was possible to make an early diagnosis. The findings of the patient mentioned in the study were included in the report with her knowledge and consent.

Case report

A 53-year-old woman (housewife) was admitted to our hospital with complaints of long-standing swallowing difficulty and neck pain. The retropharyngeal mass was noticed by the otolaryngologist. Preoperative contrast-enhanced MRI study confirmed the presence of cystic rim-enhancing 3 × 2 cm diameter retropharyngeal C3-C4-C5 mass, which had no pressure on the back of the spine or had no connection with the epidural or intradural area.

MRI demonstrated the tumor to be isointense on T1-weighted images and hyperintense on T2-weighted images (Figures 1, 2). These radiographic findings were indicative of a paraspinal soft tissue tumor (may be metastatic).

She was consulted in our clinic and taken over for treatment. There was no neurological deficit in the physical examination of the patient and there was no history of additional disease in her anamnesis. The patient had no complaints other than neck pain and dysphagia.

The patient underwent surgical resection. The patient was placed in the supine position and standard anterior cervical approach with longitudinal incision was used to access retropharyngeal C3-C4-C5 mass. The tumor capsule was clearly exposed and then capsulated, low vascular, gelatinous and grayish-colored tumor was totally removed microsurgically. The vertebral body was destroyed and the border of the tumor and vertebral body was not clear.

The lytic lesion on the anterior side of C4 vertebra corpus was electrocoagulated with bipolar

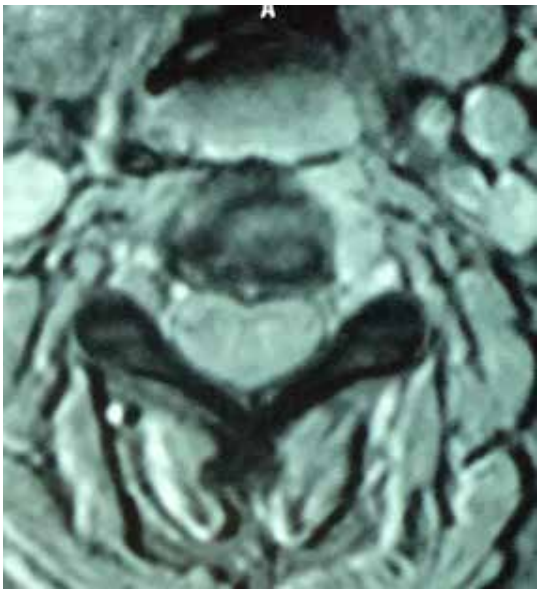

Figure 2. T1 weighted axial MRI image reveals well-circumferenced lesion localized in the anterior aspect of the C4 vertebral body

electrodes. After the surgical procedure she had no additional neurological deficit. After the operation, the stability of the spine was not impaired. We suggested that the patient use cervical collar for 30 days.

Histological examination of the tumor showed a low mitotic index, Ki 67 index was 1–2%. Most of tumor cells were stained positive for vimentin, CK, S100 and EMA, also stained negative for GFAP. The pathological diagnosis was chordoma.

Discussion

Chordoma, which is a rare bone tumor, originates from the embryonic notochord. It occurs more frequently in the midline of the axial skeleton. Although chordoma does not grow rapidly, it is a tumor with rapid postoperative recurrence. Of all reported cases, only about 15% are located in the vertebral column [2, 3]. The remaining 85% involve the sacrococcygeal area or spheno-occipital area. Chordoma in the spinal column is very rare, especially in the cervical region.

Early diagnosis of vertebral chordoma is difficult due to the slow growth character and nonspecific initial symptoms. Usually cervical spinal chordomas cause no symptoms until reaching a size large enough to cause obstructive symptoms. CT and/or MRI has detected such lesions which would previously have been subclinical. The choice of the surgical approaches depends on the location, expansion and the size of the tumor. Although surgical resection is the first treatment option, it requires total resection. The postoperative recurrence rate of the tumor is inversely proportional to the amount of resection [8–10].

It is known that the effect of chemotherapy is low in chordomas, radiotherapy is frequently applied. Radiotherapy should be given, especially in resections that cannot be gross total.

Conclusions

Chordomas are relatively rare tumors and cervical spinal location is an unusual site for them. They are slow growing tumors and have nonspecific initial symptoms. Neck pain is the most common reason for admission and it is that group of patients we encounter frequently in neurosurgery outpatient clinics. A possible cervical tumor should always be in our minds and the diagnosis and treatment of the patient should be in this direction. In cases with radiological findings suggestive of chordoma, surgical treatment should be the first choice and total resection should be done.

Conflict of interest

The authors declare no conflict of interest.

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