ECRONICON OPEN ACCESS

NEUROLOGY
Case Report

Cervical Chordoma Involving C3/C4: A Case Report

Syed MuneebYounus[1]*, Muhammad Imran[2], Ateeq Ahmed Khan[3] and Junaid Ashraf[4]

[1]Resident Department of Neurosurgery, DUHS, CHK, Pakistan

[2]Associate Professor, Department of Neurosurgery, DUHS, CHK, Pakistan

[3]Associate Professor, Department of Neurosurgery, DUHS, Civil Hospital Pakistan

[4]Head of Department, Department of Neurosurgery, Civil Hospital Pakistan

***Corresponding Author:** Syed MuneebYounus, Resident Department of Neurosurgery, DUHS, CHK, Karachi, Pakistan.

Received: November 01, 2015; **Published:** November 06, 2015

Abstract

Chordoma is a rare, indolent, osteolytic, primary bone malignancy typically arising in the midline and derived from notochordal remnants. It predominantly emerges from the axial skeleton. We present a case of C3/C4 chordoma after taken informed written and oral consent in 30 years old lady who came to us with complains of difficulty in walking and inability to hold objects in both hands. MRI showed collapse of C3 and C4 vertebral bodies with a large mass extending into paravertebral tissues and causing spinal cord compression. Biopsy confirmed the diagnosis of a chordoma and surgical excision was carried out. No recurrences or metastasis were noted in a 6 months' follow-up.

Keywords: *Chordoma; Notochord; Cervical spine; En bloc resection*

Introduction

Chordomas are rare, slow-growing malignant yet locally aggressive osteolytic primary bone tumor derived from persistent notochordal remnants that principally arise in the midline [1,2]. Predominantly, chordomas arise in the axial skeleton; however, cases of extra-axial chordomas have also been reported [3]. They are the most common primary bone tumors of spine and have a male to female ratio of 2:1. The most common site is cranium followed by spine and sacrum [4]. Cervical spine is affected in 6% of all cases of which upper cervical segments have a higher predilection [1,5]. The usual complains in patients with cervical chordomas include neck pain, dysphasia, dysphonia and symptoms due to neck mass or spinal cord compression [4]. Although MRI plays a pivotal role in establishing the extent of tumor and involvement of the surrounding structure [6], CT scan guided biopsy performed by a trocar serves as the mainstay to confirm the diagnosis [3]. The treatment of choice is surgical resection [3,6]. Median survival in these patients irrespective of age, race and gender was found to be 6.3 years [7].

Here we present a case of a 30 years old lady after taken oral and written informed consent, which was diagnosed as a case of C3/C4 chordoma, a rare location for this entity. To our knowledge, this is the first case of a cervical chordoma reported from Pakistan. We have discussed the clinical presentation, etiology, radiological and histological features, and treatment of the case with references to previously reported cases.

Case Report

We report the case of a 30 year old female who presented to our hospital in April 2014 with complains of neck pain and vertigo along with difficulty in walking and inability to hold objects in both hands for 1 month. There was no dysphagia, dysphonia or swelling in the neck. Physical examination revealed decreased sensation and power in all four limbs.

MRI cervical spine with and without intra venous contrast administration was performed, revealing almost complete collapse of C4 vertebral body (Figure 1). Abnormal signal was seen involving collapsed C4 vertebral body as well as adjacent body of C3 Vertebra,

showing patchy enhancement on post contrast study. The abnormal signal was also seen to involve C3/C4 disc. There was extensive paravertebral soft tissue mass which appeared hypo intense on T1 weighted images while hyper intense on T2 weighted images and showing thick peripheral enhancement. Right paravertebral component measured 1.2 cm in maximum thickness and left paravertebral component measured 0.8 mm in maximum thickness. The craniocaudal dimension of the mass was 3.8 cm extending from C2 till C5 vertebral level. The paravertebral soft tissue mass was shown gaining access to epidural space through neural foramina resulting in severe compression of spinal cord and displacing it posteriorly. Rest of the vertebrae and intervertebral disc had normal height and signal characteristics. Loss of normal cervical curvature was noted likely due to muscular spasm.

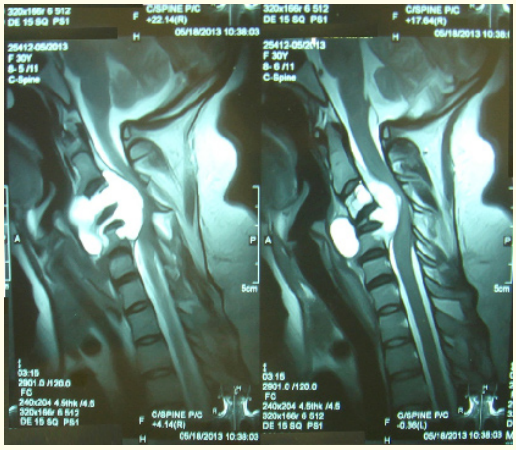

Figure 1: *MRI Cervical spine sagittal view showing collapsed C4 vertebral body and the adjacent paravertebral soft tissue mass.*

Surgical options were explained but family only gave consent for open biopsy. Formalin preserved tissue specimen, obtained from C3-C4 was sent for histopathology. The microscopic examination of the sections revealed a neoplastic lesion composed of lobules of tumor in which sheets, cords, nests of vacuolated eosinophilic to clear cells were embedded in a myxoid matrix. The neoplastic cells were shown to have clear vacuolated cytoplasm with hyperchromatic nuclei. Immunohistochemcial stains were performed which showed positive reactivity patterns for S100 and Cytokeratin AE1/AE3. These features were found to be consistent with chordoma.

Her power subsequently worsened, after initial surgery. Repeat MRI demonstrated significant vertebral deformity with cervical cord compression. At this stage, with the consent of the family, she underwent en bloc resection of the tumor through an anterior approach and then anterior cervical fixation was performed (Figure 2). Her power improved to normal after surgery. For the residual tumor which was abutting the vertebral artery, she underwent cyber knife robotic radiosurgery. The radiosurgery dose was 40 grays given in 5 fractions. During her subsequent visit and follow up MRI after 6 month the small residual did not show progression of disease.

Discussion

Chordomas are rare, slow-growing malignant yet locally aggressive osteolytic primary bone tumors derived from intraosseous remnants of embryonic notochord principally arising in the midline [1,2,9]. The vertebral column develops around the embryonic notochord. Notochord persists only as the nucleus pulposus of the intervertebral discs and degenerates where it is surrounded by vertebral bodies [2]. The tumor forming remnants of notochord called ecchondrosis physaliphora are found in clivus and nucleus pulposus, but ectopic remnants have also been documented in the literature [6].

Chordomas are the most frequent among primary tumors of the mobile spine [4,8], with an incidence rate of less than 1 per 100 000 individuals with a higher incidence in males and usually presenting in patients aged > 40 years 1, 7. Of all chordomas 32% are cranial,

32.8% spinal and 29.2% are sacral. Median survival is 6.29 years. The 5 and 10 year survival rates were 67.6% and 39.9%, respectively [1,3,7,10]. Of all chordomas, only 6% affect the cervical spine and within these, upper cervical segments have a higher predilection [4,5].

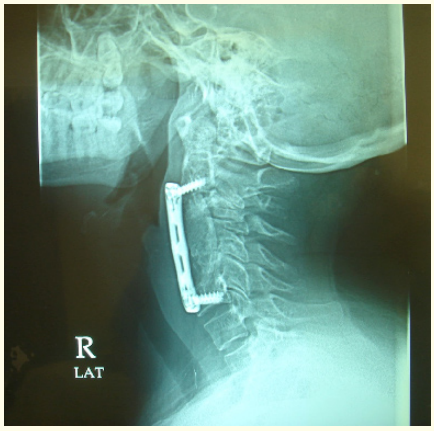

Figure 2: *X-ray spine after en bloc resection and anterior cervical fixation.*

Clinical presentation is primarily with neck pain caused by destruction of bone, compressed nerve or due to the vertebral segments becoming more mobile. Symptoms other than cervical pain can be due to neck mass or spinal cord compression, depending on the extent of intra-spinal involvement [4-6].

MRI is fundamental in assessing the extent of tumor. Chordoma may appear hypo or isointense to muscle on T1 while hyper-intense on T2 weighted images. The lesion enhances with the injection of contrast [5,6,9]. In our case, the lesion was contrast-enhanching, hypointense on T1 and hyper intense on T2.

Microscopically, chordomas consist of nests or cords of physaliphorous cells separated by fibrous tissue septa and mucoid intercellular substance. These cells have a vacuolated eosinophilic cytoplasm. The tumor cells show markers of both epithelial as well as mesenchymal differentiation such as vimentin, cytokeratin 5/6 and S-100 protein on immunohistochemical analysis [1,5].

Chordomas only rarely metastasize. However, metastases occurring in the lungs, liver, bone, lymph nodes, and skin have been reported in 3-48% of cases and is often associated with local recurrence of the primary tumor [2,9,10]. Chordomas of the mobile spine metastasize more often than sacrococcygeal lesions [5].

Surgical resection with adjuvant radiotherapy is the treatment of choice in cases of cervical chordoma as was done in this case [4]. Amongst surgical procedures, En-bloc radical dissection, if feasible, along with stabilisation is the optimal treatment and allows long term disease-free survival [2,6,8]. If not favourable, piecemeal removal of tumor can be followed by radiotherapy [4].

Traditionally, chordomas are considered radioresistant [3,5]. Although, the curative doses of radiation therapy which should at least be in the 70 Gy range, are higher than the tolerable dose for spinal cord, still radiation is valuable particularly in cases where complete excision could not be carried out. Conventional dose of 45–60 Gy, as used in our case, is well within the radiation tolerance and has resulted in 5-year local control of 10–40% [3,10]. Seeding of the tumor during biopsy, reconstruction or primary tumor resection, all can lead to local recurrence. Even in the recurrent cases, aggressive en-bloc resection accompanying seeding precautions is preferred [10].

Conclusion

The authors present a case of cervical chordoma involving the cervical segments C3 and C4, in a 30 year old female. Diagnosis was made on histopathology and the tumor extent was determined on cervical spine MRI. En bloc resection and an anterior cervical fixation were performed. Prompt diagnosis and aggressive excision of this neoplasm are key to a good outcome.

Bibliography

1. Shamekh R., *et al*. "A woman with swelling of the posterior pharyngeal wall". *JAMA Otolaryngology Head Neck Surgery* 140.7 (2014): 671-672.
2. Kotnis N and Goepel J. "Paraspinal chordoma mimicking a neurofibroma: a rare but important radiological pitfall". *Skeletal Radiology* 42.3 (2013): 443-446.
3. Jiang L., *et al*. "Upper cervical spine chordoma of C2-C3". *European Spine Journal* 18.3 (2009): 293-298 discussion 298-300.
4. Ahsan FM., *et al*. "Cervical chordoma managed with multidisciplinary surgical approach". *ANZ Journal of Surgery* 81.5 (2011): 331-335.
5. D'Haen B., *et al*. "Chordoma of the lower cervical spine". *Clinical Neurology Neurosurgery* 97.3 (1995): 245-248.
6. Bannur U., *et al.* "Paravertebral cervical chordoma--a case report". *British Journal of Neurosurgery* 25.4 (2011): 527-529.
7. McMaster ML., *et al*. "Chordoma: incidence and survival patterns in the United States, 1973-1995". *Cancer Causes and Control* 12.1 (2001): 1-11.
8. Boriani S., *et al*. "Chordoma of the mobile spine: fifty years of experience". *Spine* (Phila Pa 1976) 31.4 (2006): 493-503.
9. Guillonnet A., *et al*. "Cervical chordoma with moderate bone impairment in a child. Answer to October E-quid". *Diagnostic and Interventional Imaging* 93.11 (2012): 903-906.
10. Erkmen CP., *et al*. "Case report: Successful treatment of recurrent chordoma and bilateral pulmonary metastases following an 11-year disease-free period". *International Journal of Surgery Case Reports* 5.7 (2014): 424-427.

Volume 2 Issue 3 November 2015