Cervical chordoma involving C3/C4: A case report

Muhammad Imran, Atiq Ahmed Khan, Syed Muneeb Younus

Abstract

Chordoma is a rare, indolent but locally invasive, osteolytic, slow growing, low grade, primary bone malignancy, derived from the embryonic remnants of the notochord. It is a midline tumour and it predominantly emerges from the axial skeleton. The most commonly observed location of a chordoma is in the sacrococcygeal region (50 to 55%) followed by the cranio occipital region (25 to 30%) We present a case of a 30 years old lady who came to us with complaints 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. A preamble diagnosis of a chordoma at level of C3-C4 spine was made, a biopsy done to confirm the diagnosis of a chordoma and surgical excision was carried out. No recurrences or metastasis were noted in a follow up at 6 month post operatively.

Keywords: Chordoma, Notochord, Cervical spine, En bloc resection, C3-C4 spine.

Introduction

Chordomas are rare, slow-growing malignant yet locally aggressive osteolytic primary bone tumours derived from persistent notochordal remnants that principally arise in the axial skeleton.[1,2] Cases of extra-axial chordomas have also been reported.[3] They make up about 1% of all intracranial, 4% of primary bone and 2% of spinal tumours[4,5] They are the most common primary bone tumours of spine and have a male to female ratio of 2:1. The most common site is cranium followed by spine and sacrum.[6] Cervical spine is affected in 6% of all cases of which upper cervical segments have a higher predilection.[1,7] The usual complaints in patients with cervical chordomas include neck pain, shoulder weakness, dysphagia, dysphonia and symptoms due to neck mass or spinal cord compression.[6] Although MRI plays a pivotal role in establishing the extent of the tumour and involvement of the surrounding structures,[8] CT scan guided biopsy performed by a trocar serves as the mainstay to confirm the diagnosis.[3] The treatment of choice is surgical resection.[3,8] However to resect the tumour completely is difficult due to its close proximity to the spinal cord.Median survival in these patients irrespective of age, race and gender was found to be 6.3 years.[9]

Here we present the case of a 30 years old lady who was diagnosed with 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 us via the out patient clinic at Department of Neurosurgery at Civil Hospital Karachi, Pakistan, in the month of April 2013. Her presenting complaints were, neck pain, vertigo, difficulty in walking and an inability to hold objects (both hands) for a period of 1 month. There was no dysphagia, dysphonia or swelling in the neck. Physical examination revealed decreased sensation and power in all four limbs.

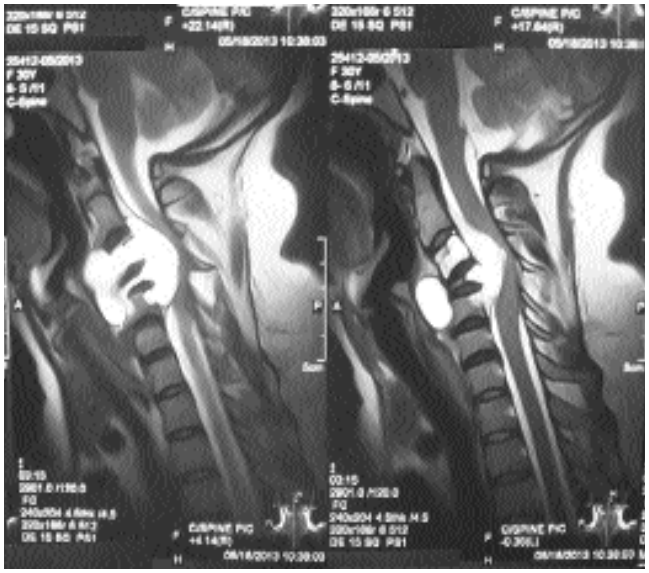

Figure-1: MRI Cervical spine sagittal view showing collapsed C4 vertebral body and the adjacent paravertebral soft tissue mass.

Department of Neurosurgery Dow University, Civil Hospital Karachi.
Correspondence: Syed Muneeb Younus. Email: muneebkazi@gmail.com

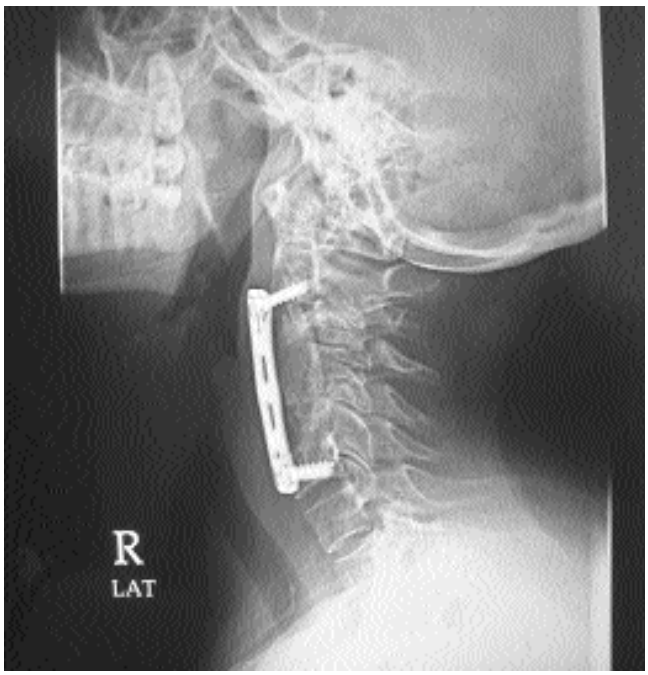

Figure-2: X-ray spine after en bloc resection and anterior cervical fixation.

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.

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 tumour 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 tumour through an anterior approach and then anterior cervical fixation was performed (Figure-2). Her power improved to normal after surgery. For the residual tumour 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 tumours derived from intraosseous remnants of embryonic notochord principally arising in the midline.[1,2] 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 tumour forming remnants of notochord called ecchondrosis physaliphora are found in clivus and nucleus pulposus, but ectopic remnants have also been documented in the literature.[4,8]

Chordomas are the most frequent among primary tumours of the mobile spine,[6] 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.[1,9] 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.[1,3,9,10] Of all chordomas, only 6% affect the cervical spine and within these, upper cervical segments have a higher predilection.[6,7]

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.[6-8]

MRI is fundamental in assessing the extent of tumour.

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.[7,8] 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 tumour cells show markers of both epithelial as well as mesenchymal differentiation such as vimentin, cytokeratin 5/6 and S-100 protein on immunohistochemical analysis.[1,7]

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 tumour.[2,10] Chordomas of the mobile spine metastasize more often than sacrococcygeal lesions.[7]

Surgical resection with adjuvant radiotherapy is the treatment of choice in cases of cervical chordoma as was done in this case.[1,6] Amongst surgical procedures, En-bloc radical dissection, if feasible, along with stabilisation is the optimal treatment and allows long term disease-free survival.[2,8] If not favourable, piecemeal removal of tumour can be followed by radiotherapy.[6]

Traditionally, chordomas are considered radioresistant.[3,7] 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 tumour during biopsy, reconstruction or primary tumour 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 tumour 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.

References

1. Shamekh R, Altiok S, Leon ME.A woman with swelling of the posterior pharyngeal wall. JAMA Otolaryngol Head Neck Surg. 2014; 140:671-2.
2. Kotnis N, Goepel J.Paraspinal chordoma mimicking a neurofibroma: a rare but important radiological pitfall. Skeletal Radiol. 2013;42:443-6.
3. Jiang L, Liu ZJ, Liu XG, Ma QJ, Wei F, Lv Y, et al. Upper cervical spine chordoma of C2-C3 Eur Spine J. 2009; 18:293-8.
4. Lang SS, Thawani JP, Sayah A, Venneti S, Brewington D, Zager E, et al. Chordomas and Their Consideration in the Radiographic Differential of Extra-Axial Lesions of the Central Nervous. Open J Modern Neurosurg 2012; 2:58-62.
5. Khong P, Milross J, Cherukuri RK. Lumbar chordoma mimicking a neurogenic tumour. J Clin Neurosci. 2011; 18:302-4.
6. Ahsan F, Inglis T, Allison R, Inglis GS. Cervical chordoma managed with multidisciplinary surgical approach. ANZ J Surg. 2011; 81:331-5.
7. D'Haen B, De Jaegere T, Goffin J, Dom R, Demaerel P, Plets C.Chordoma of the lower cervical spine.Clin Neurol Neurosurg. 1995; 97:245-8.
8. Bannur U, Chawda SJ, O'Donovan DG, Kaddour H, David KM. Paravertebral cervical chordoma--a case report.Br J Neurosurg. 2011; 25:527-9.
9. McMaster ML, Goldstein AM, Bromley CM, Ishibe N, Parry DM. Chordoma: incidence and survival patterns in the United States, 1973-1995. Cancer Causes Control. 2001; 12:1-11.
10. Erkmen CP, Barth RJ Jr, Raman V. Case report: Successful treatment of recurrent chordoma and bilateral pulmonary metastases following an 11-year disease-free period. Int J Surg Case Rep. 2014; 5:424-7.