ABSTRACT

Objectives: chordomas of cervical spine are rare tumors but are the primary malignant tumors of the spine. As Chordomas has poor sensitivity to radiotherapy and chemotherapy it requires surgical resection. The purpose of this study was to determine the suitable methods for removal of the tumors completely and outcome over time in patients undergoing complete en bloc excision. Methods: a retrospective study of twelve patients from October 2003 to August 2007 between the ages of 45-75 years who presented with gradual onset of neck pain and upper extremity weakness. All patients were clinically evaluated and preoperatively underwent Digital X-Rays, CT scans and contrast enhanced MR imaging. All patients underwent tumors resection and spinal instrumentation. All cases were followed up clinically and radiographically for determination of their status. Results: eleven patients were available for follow-up. All patients underwent an anterior

RESUMO

Objetivos: os tumores da coluna cervical são raros, mas são tumores malignos primários da coluna. Como os cordomas apresentam baixa sensibilidade à radioterapia e à quimioterapia requerem cirurgia para sua ressecção. O objetivo deste estudo é determinar os métodos adequados para a remoção total dos tumores e conhecer os resultados para o paciente no período posterior à sua retirada. Métodos: foi realizado um estudo retrospectivo de 20 pacientes, de Outubro de 2003 a Agosto de 2007, com idade entre 45 e 75 anos, os quais apresentavam uma gradual dor no pescoço e uma importante fraqueza nas extremidades superiores. Todos os pacientes foram avaliados clinicamente e no período pré-operatório por meio de radiografia digital, tomografia computadorizada e por imagem de ressonância magnética com contraste. Todos os pacientes foram submetidos à ressecção dos tumores e instrumentação da coluna vertebral. Todos os casos

RESUMEN

Objetivos: los tumores de la columna cervical son raros, siendo tumores primarios malignos de la columna. Como los cordomas presentan baja sensibilidad a la radioterapia y a la quimioterapia requieren cirugía para su resección. El objetivo de este estudio es determinar los métodos adecuados para la remoción total del tumores y conocer los resultados de los pacientes en el periodo posterior a su retirada. Métodos: fue realizado un estudio retrospectivo de 20 pacientes, de Octubre de 2003 a Agosto de 2007, con edad entre 45 y 75 años, que presentaron un dolor gradual en el cuello y una debilidad importante en las extremidades superiores. Todos los pacientes fueron evaluados clínicamente y en el preoperatorio fue realizada radiografía digital, tomografía computarizada y resonancia magnética de contraste. Todos los pacientes fueron sometidos a la resección del tumores y a la instrumentación de la columna vertebral. Todos los casos...
stabilization procedure combined with interbody fusion (seven with iliac crest and five with Interbody cages) whereas one required an occipitocervical fusion. At the time of this submission, there were two signs of recurrence. Discussion: the uncontrolled growth of chordoma is commonly the cause of death. En bloc excision of the lesion, sometimes combined with radiation therapy as an adjuvant, obtained the best results. Conclusion: chordoma occur in most cases directly from the vertebral body. En bloc excision of these tumors even if marginal seems to be the most effective treatment combined with megavoltage radiation which can be administered as an adjuvant.

KEYWORDS: Chordoma/surgery; Orthopedic procedures/methods; Spinal neoplasms/surgery

INTRODUCTION
Chordoma is a low grade malignant tumors accounting for about 1% to 4% of all malignant bone tumors. It originates from the remnants of the notochord. It is predominantly found in the clivus (50%), followed by the sacral spine (15%). Only rarely is this tumors seen in cervical spine accounting for 6% of all chordoma. Cervical spine Chordomas present significant surgical challenges because of the important anatomical structures present in this area. The tumors grow slowly by infiltrating cancellous bone, is seen typically in adults and the elderly. At the time of initial presentation, the cervical chordoma usually shows infiltration into the paravertebral and epidural compartments. It has a marked tendency toward recurrence following intraslesional excision or biopsy. Metastasis is usually seen in the brain, skin, lungs, bone or internal organs. Rate of metastasis varies from 0% to 5%. Survival seems to be more affected by the local spread rather than by metastasis. Magnetic resonance imaging and computed tomography have made the diagnosis of the size of the tumors faster and easier, they also play an important role in improving the prognosis of chordoma by discovering small tumors, which can be submitted to en bloc resection.

In this study we present twelve cases with isolated cervical spine Chordomas. All patients at the time of presentation had evidence of tumors extension. In this study we determine the suitable methods for removal of the tumors completely and outcome over time in patients undergoing complete en bloc excision.

METHODS
We retrospectively analyzed twelve patients with cervical chordoma from October 2003 to August 2007 at the Lilavati Hospital and Research Centre, Mumbai. All data was obtained from hospital records, including preoperative and sequential postoperative clinical findings, radiological details and pictures and details of the status of the patient till the last follow-up. Neurological deficit was assessed by Frankel grading.
Out of the twelve patients studied eight were males and four were females. Mean age at diagnosis was 60 years. Follow-up duration ranged from 12 to 36 months. The slow and gradual onset of pain was the most consistent complaint; all patients presented with neck pain and upper extremity symptoms. Upper extremity symptoms ranged from hand tingling and numbness to overt weakness. One patient presented with difficulty in swallowing.

All patients preoperatively underwent standard X-rays, CT scans to assess the degree of bone destruction and cervical spine instability and contrast enhanced MRI to evaluate bone and soft tissue involvement, and nuclear bone scan to check for metastasis, we used MR angiography to evaluate the patency of vertebral artery in addition to a complete preoperative workup (Figure 1-2).

Based on the preoperative images we staged all tumors by using the oncological staging system devised by Enneking et al. and the extent of the lesion was described according to the Weinstein- Borian- Biagini (WBB) system. According to the Enneking staging system, the lesions were classified as 1A (4 cases) and 1B (8 cases). The tumors arose mostly central in the body and invaded the whole vertebrae. Chordomas appear late on standard X-Rays, the radiographic pattern is mostly radiolucent with scanty ossifications and huge masses in the surrounding soft tissues. The discs were spared. MRI and CT scan proved useful in detecting lesions which were not seen on radiographs, the MRI signal is hypo dense in T1 weighted and hyper intense in T2 weighted images. In six cases complete vertebral collapse occurred. We planned our surgical approach based on a variety of factors including tumors location, extent, area of cord compression and degree of instability. In all of the twelve surgical interventions, two were done by combined anterior and posterior approach; these had pure anterior, middle and posterior column involvement. Ten were done by anterior approach; these had anterior and middle column involvement.

The treatment performed on twelve cases included:

- En bloc resections in four cases, the margins were contaminated at some point and for this reason radiation therapy was performed as an adjuvant in these cases.
- En bloc resection of the vertebral body (vertebrectomy) could be performed in five cases (Figure 3).
- Intralesional extra capsular excision with adjuvant radiation therapy was performed in three cases.

Specimens obtained during resection were sent for histopathological analysis in all cases (Figure 4). Spinal instrumentation was performed when surgery caused instability or for reconstruction of spine after tumors removal. All patients in this series underwent spinal stabilization. Combined anterior and posterior reconstruction was performed in two cases; the vertebral body was replaced by autogenous iliac bone graft in seven cases and by interbody cages in five cases along with anterior plating (Figure 5).

Post-operative care included immobilization of the operated spine with a SOMI brace or a hard cervical collar (Philadelphia) for three months post surgery. The duration of hospital stay following surgery ranged from five to 14 days. A contrast enhanced MR imaging study was obtained at 6 weeks in all patients to evaluate the status of the tumors. Follow-up evaluation was on outpatient basis at 6 weeks post tumors excision along with a fresh digital X-ray, further follow-ups were at 3, 6, 9 and 12 months along with fresh X-Rays for assessment of spinal alignment and integrity of the instrumentation and to check for fusion. Successful fusion required the presence of bridging trabeculae across the fused levels. A contrast enhanced MR imaging study was obtained in all patients at 3, 6 and 12 months post excision to check for early tumors recurrence. Once fusion was established, the patients were weaned off the brace to a soft cervical collar.

Figure 1
58 years lady: MRI sagittal (A) and axial (B) cuts showing C6 chordoma with cord compression

Figure 2
Patient underwent C6 body excision + decompression with interbody iliac crest bone graft and stabilization

Figure 3
Microscopic picture of same patient showing large polyhedral cells with eccentric nuclei and large mucoid foci with a chondroid appearance

Figure 4
60 year old lady with chordoma of C2 vertebrae

Figure 5
Patient underwent C2 excision with a two stage antero-posterior occipito-cervical fusion
RESULTS
All patients were analyzed using the Frankel grading. The evolution of the chordomas is known for eleven patients. All patients were submitted to clinical and imaging studies during their follow-ups to determine the relationship between treatment and outcome. In all patients in this study the tumors were resected using a standard anterior cervical approach for anterior and middle column tumours and posterior approach for involvement of posterior elements. Of the eleven patients available at follow-up their were two cases of local recurrence. The mean follow-up duration has been 24 months in this study. Three patients were treated by intraslesional extra capsular excision with adjuvant radiation therapy, at one year there was one case of local recurrence which required chemotherapy, this patient is being treated and one patient of this group was lost to follow-up. En bloc excision was performed in five cases, there was no recurrence at 48 months in this group of patients. Of the four patients who underwent En bloc resections with contaminated margins and adjuvant radiation therapy there was one patient (25%) with local recurrence at 24 months follow-up which required excision along with radiation therapy.

Chordomas are a frequent cause of cord compression because of the slow growth which expands toward the epidural space compressing the dura. In all cases a significant improvement in the neurological symptoms arose following decompression and excision of the tumors. Seven patients received postoperative radiation therapy of these two patients with local recurrence also underwent chemotherapy.

DISCUSSION
During the period of this study (2003 to 2007), twelve chordomas of the cervical spine were treated at Lilavati hospital. Chordomas of cervical spine always arise within the vertebral body, they constitute 1 to 4% of malignant bone tumors. After plasmacytomas they are the most frequent primary malignant tumors in the spine, the cervical region constitutes 6 to 7% of cases. Chordomas are locally aggressive and have a tendency to recur. Metastases are noted in approximately 30% of cases. As it is slow growing tumors it is detected late and often occupies most of the vertebrae at the time of presentation, chordomas are often destructive. Late diagnosis makes it difficult to provide appropriate treatment. According to the oncological staging proposed by Enneking, chordomas are classified as stage 1A/1B lesions. Aggressive resection is the treatment of choice.

The main problem in the treatment of chordoma is local recurrence. The uncontrolled growth of these tumors is commonly the cause of death. The treatment of chordomas in the cervical spine consists of radical excision and stabilization, followed by adjuvant radiotherapy and chemotherapy. Proton beam radiation is the modality that has been used extensively for the treatment of chordomas.

In this study five patients who underwent En bloc excisions of the tumors had no recurrence at 48 months. The results of this study thus confirms most of the conclusions of the literature and stresses the point that En bloc resection with tumors free margin seems to be the procedure of choice to allow a disease free interval. It has been reported that the margin free, en bloc tumours resection is the treatment of choice for Chordomas. This requires the tumors be confined within the vertebral body, as it is slow growing tumors it is detected late and often occupies the epidural space after penetrating the vertebral body at the time of presentation, removing such large tumors in the cervical spine in an en bloc manner when the lesion extends well beyond the spine would be associated with significant morbidity. The main disadvantage in resecting cervical spine chordoma result from the involvement of the vertebral artery and the dura mater. In this study we were able to achieve tumors eradication in three such cases with intraslesional excision and adjuvant radiation therapy, one out of three patients from this group has had a disease free interval after a follow-up of 36 months, this being a reasonable method of choice when en bloc resection is not feasible in tumors with intradural and soft tissue involvement. In 4 cases the margins were contaminated and in these cases adjuvant radiation therapy was given obtaining a disease free interval for 36 months in three out of four patients.

CONCLUSION
Chordomas in cervical spine are rare tumors. They arise directly from the vertebral body and tend to expand the whole body. At presentation, the majority of patients had tumors beyond the vertebral body, this tumors spread beyond the vertebral body makes it difficult to perform an en bloc resection of the body as the margins would never be disease free, considering the risk benefit ratio as the surgical risk presumably exceeds the benefit. In this study the patients with cervical chordoma presented with nonspecific symptoms of neck pain with radicular and myelopathic changes. The result of this study indicates that en bloc resection of chordoma in some areas seems to be the most effective treatment in local control of this disease in combination with stabilization and reconstruction of the spine. In cases when en bloc excision is not feasible that is in cases with involvement of vital neurological structures a complete excision that is piecemeal removal of the tumors is a viable technique combined with megavoltage radiation therapy or proton beam radiation.
REFERÊNCIAS


Correspondence

Dr. Amit Kohli
802-Green Blaze CHSL,
Near IDBI Bank
Juhu-Versova link road,
Andheri (West), Mumbai-400053
INDIA
Tel.: + 91-9820504676
E-mail: amitkohli99@yahoo.com

COLUNA/COLUMN. 2008;7(1):71-75