

# MANAGEMENT OF PRIMARY SPINAL CHONDROSARCOMA

## Report of two cases causing cord compression

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**ABSTRACT** - Chondrosarcomas are malignant tumors that rarely grow inside the spinal canal. Prognosis depends on histological features, patient's age and surgical margins free from tumor. Response to radio and chemotherapy is poor. Ideal treatment consists of total "en-block" resection, not always achievable due to limitation of location, compromise of stability and risk of inducing neurological deficits. Two cases of spinal chondrosarcoma causing cord compression are reported, located in the cervical and thoracic spine. Microsurgical technique consisted of initial debulking followed by removal of margins until limits free from tumor were obtained. Total resection was accomplished and neurological function improved in both cases. Follow-up has been seven and one year respectively, with no evidence of recurrence and preserved neurological functions. Association between chondrosarcoma and estrogen-dependent tumor has been confirmed in this report. Although "en-block" resection of a chondrosarcoma should be tried whenever possible, tumor fragmentation should be considered in difficult cases, as in the present report, in which a long period free from recurrence with good quality of life can be obtained.

**KEY WORDS:** chondrosarcoma, spinal neoplasms, spinal cord compression, microsurgery.

### **Manejo dos condrossarcomas espinhais primários: relato de dois casos causando compressão medular**

**RESUMO** - Os condrossarcomas são tumores malignos, raramente localizados no interior do canal espinhal, com prognóstico dependente do grau histológico do tumor, idade do paciente e margens cirúrgicas livres. Esses tumores apresentam pouca resposta à radio e quimioterapia. O tratamento ideal consiste em ressecção tumoral em bloco, condição particularmente difícil em se tratando de tumores causando compressão medular, devido à localização da lesão, comprometimento da estabilidade axial e necessidade da manutenção ou recuperação da integridade da função neurológica do paciente. Relatamos dois casos de condrossarcomas causando compressão medular, um na coluna cervical e outro na torácica, submetidos a cirurgia com esvaziamento tumoral seguido de remoção das margens, atingindo-se ressecção total e melhora das funções neurológicas em ambos os casos. Um paciente se encontra há 7 anos e outro há 1 ano livre de recorrências, ambos com funções neurológicas preservadas. Associação entre condrossarcoma e tumor dependente de estrógeno é confirmada clinicamente pelo presente estudo. Embora uma ressecção em bloco dos condrossarcomas deva ser tentada quando possível, deve-se considerar uma ressecção com fragmentação tumoral em casos específicos, podendo-se atingir, como demonstrado nos presentes casos, períodos livres de recorrência e longo tempo de sobrevida.

**PALAVRAS-CHAVE:** condrossarcoma, neoplasias medulares, compressão da medula espinhal, microcirurgia.

Chondrosarcomas are malignant lesions, which represent the second most common tumor of the skeleton. However, presentation as primary tumor of the spine is extremely rare. These tumors can originate from healthy bone or develop from a cartilaginous lesion with sarcomatous degeneration<sup>1</sup>. Primary malignant tumors should be resected with wide safety margins. However, limitations to total resection may

involve risk of causing spinal instability and/or inflicting new deficits. Isolated cases of spinal chondrosarcomas have been reported<sup>12-18</sup> as well as a few series with limited number of patients<sup>19-22</sup>.

Specific surgical strategies are delineated in the present report emphasizing the necessity of total removal, particularly considering tumor resistance to adjuvant therapy<sup>19,20</sup>.

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## CASES

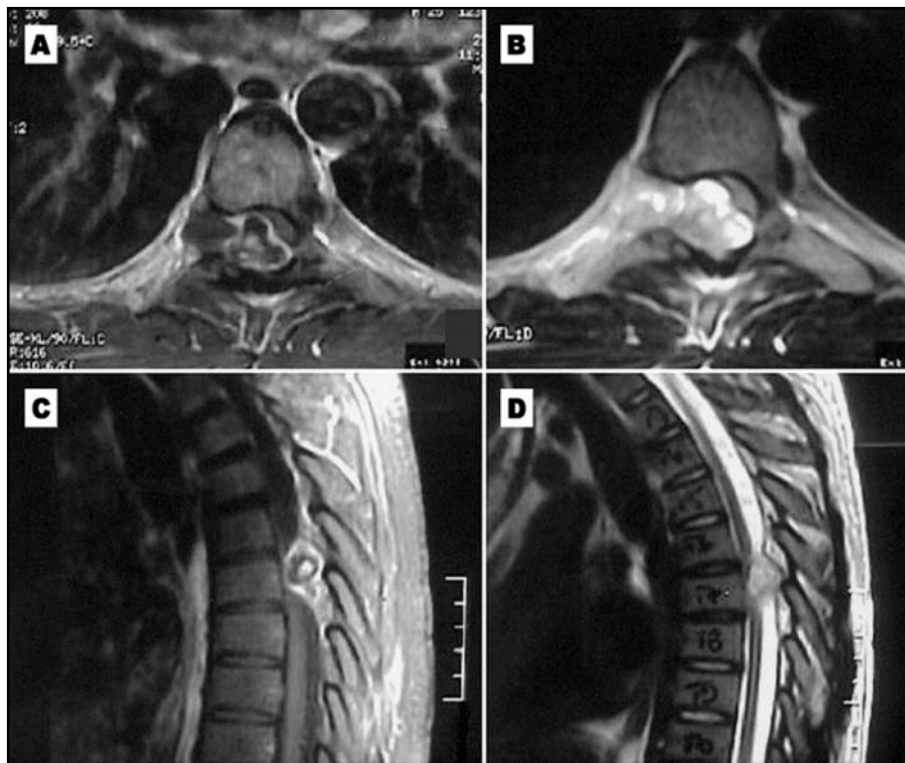
**Case 1.** An 80 year-old woman was admitted with progressive weakness in her lower limbs, which resulted in inability to walk, one week before admission. Neurological examination showed spastic paraparesis. Hypoesthesia with partial loss of pain and temperature below T4 level and preserved proprioception and vibration sense were also demonstrated. She was submitted to a cervical and thoracic spinal magnetic resonance study (MRI), which showed a posterior and lateral extradural tumor from C5 to T1, mainly on the left side, inside the spinal canal, causing significant cord compression. Laminectomies from C5 to T1 were carried out exposing an extradural mass of great proportions on the left side of the spinal canal affecting the posterior elements of C6 to T1 and causing a compressive effect on the spinal cord. The mass was completely resected in a piece-meal fashion until margins free of tumor were obtained. Post-operative period was uneventful with gradual reversal of her neurological deficits. Pathology specimen was compatible with chondrosarcoma and no adjuvant therapy was used.

Four years after the spinal surgery the patient underwent a hysterectomy due to an endometrium carcinoma. After two years, she was submitted to another procedure, a radical mastectomy, due to a breast cancer.

Seven years have elapsed since the spinal operation with no signs of tumor recurrence.

**Case 2.** A 25 year-old man was admitted with progressive right intercostal pain irradiated from the back, at a mid thoracic level, of one-year duration. Six months prior to admission he started with gait difficulties and frequent falls, progressing to a severe paraparesis and inability to walk.

His neurological examination demonstrated bilateral normal strength in upper extremities, and only minor proximal muscle function in his lower limbs. Partial loss of pain and temperature and diminished proprioception and vibration were seen below the T6-T7 level, in addition to severe spasticity. Thoracic spine MRI demonstrated a large extradural mass lesion, at T6-T7 level, 3 cm in diameter, compressing the spinal cord, growing through an enlarged right intervertebral foramen (Fig 1). The patient underwent a T6-T7 laminectomy with removal of the respective right facet, which was compromised by a whitish dense extradural tumor. The spinal cord was compressed anteriorly and shifted to the left as tumor advanced through the T6-T7 right foramen. The lesion was completely resected, in a piece-meal fashion until margins free of tumor were obtained. The T6-T7 foramen was left wide opened and the nerve root free of



*Fig 1. A) T1-weighted, gadolinium-enhanced MRI axial at the T6-T7 level; B) T2-weighted axial MRI at the same level; C) T1-weighted, gadolinium-enhanced MRI, sagittal; D) T2-weighted sagittal MRI. The images show an heterogeneous enhanced extradural lesion, compressing the spinal cord and growing through an enlarged right intervertebral foramen.*



Fig 2. T1-weighted, gadolinium-enhanced MRI obtained 1 year after the complete tumor removal. Axial at T6-T7 level (A) and sagittal (B) views are shown, disclosing no signs of recurrence.

compression. MRI in the first post-operative day confirmed total resection. Neurological deficits gradually improved. Pathology specimen was compatible with chondrosarcoma and no adjuvant therapy was used.

One year has elapsed since the operation and the patient remains asymptomatic. A new MRI was recently obtained confirming no signs of tumor recurrence (Fig 2).

## DISCUSSION

Surgical treatment of spinal chondrosarcoma is particularly difficult. Prognosis depends mainly on tumor removal, whereas the peculiar anatomical features of the spine practically avoids an ideal "en-block" resection. When all locations are considered, survival from chondrosarcoma is relatively high, reaching more than 87% of patients at 5 years; however, it is considerably lower when only pelvic, sacral and spinal locations are considered, varying between 25 and 54%<sup>19</sup>.

There are only a few series in the literature concerning spinal chondrosarcomas<sup>19-22</sup>. Thoracic spine is the most common location and the male sex is predominant<sup>20-21</sup>. According to Bergh et al. factors associated with worse prognosis are high histological grade, advanced age, primary surgery out of a referral center, incisional biopsy only and inadequate surgical margins<sup>19</sup>. York et al. evaluated 21 patients with spinal chondrosarcoma, during a period of 43 years, demonstrating a longer disease free interval in patients with total rather

than subtotal resection. The tumor recurred in 64% of the patients and the median disease free interval was 16 months without adjuvant therapy<sup>20</sup>. Shives et al. studied 20 patients with chondrosarcoma of the spine and found 100% of recurrence in patients submitted to tumor debulking only, half of the cases recurring among the six patients with inappropriate surgical margins and no recurrences in two patients with radical resection<sup>21</sup>.

Even though rare cases with an "en-block" resection of the lesions have been reported, some with spinal instrumentation, and the prognosis in the long run has not been clearly defined<sup>23-26</sup>. Patients submitted to radiotherapy did not demonstrate significant increase in survival in relation to those treated by surgery only<sup>19,20</sup>. There are no studies in the literature able to demonstrate effectiveness of radiotherapy, as well as of chemotherapy, in spinal chondrosarcoma.

Case 2 patient presented with enlargement of the intervertebral foramen, raising the suspicion of neurofibroma as first possible diagnosis<sup>27,28</sup>. Only one case of foraminal chondrosarcoma has been reported by Yünter et al., probably originating from a previous osteochondroma<sup>28</sup>.

Association between chondrosarcoma and adenocarcinoma of breast, without previous radiotherapy, has been reported by Mummaneni and Rosenberg<sup>29</sup>. Chondrosarcomas may suffer hormonal in-

fluence, even in the absence of detectable estrogen receptors. High estrogen levels seem to increase their growth<sup>30</sup>.

Case 1 patient developed subsequently both breast and endometrial carcinoma, representing the only case reported in the literature with such an association.

Total surgical resection is the best therapeutic option for chondrosarcomas, considering its resistance to radiotherapy and chemotherapy. Piecemeal removal is recommended when "en-bloc" resection is not feasible, but assurance of margins free from tumor must be obtained<sup>31,32</sup>.

When complete resection is obtained prognosis may be good, even in the presence of adverse factors, such as advanced age, as in case 1. Association between chondrosarcoma and estrogen-dependent tumors has been confirmed in this report.

## REFERENCES

- Gelabert-Gonzalez M, Reyes-Santias RM, Garcia-Pravos A. Dorsal medullary compression due to a primary vertebral chondrosarcoma. *Rev Neurol* 2000;30:322-324.
- Di Lorenzo N, Palatinsky E, Artico M, Palma L. Dural mesenchymal chondrosarcoma of the lumbar spine: case report. *Surg Neurol* 1989;31:470-472.
- Blaylock RL, Kempe LG. Chondrosarcoma of the cervical spine: case report. *J Neurosurg* 1976;44:500-503.
- Hirsh LF, Thanki A, Spector HB. Primary spinal chondrosarcoma with eighteen-year follow-up: case report and literature review. *Neurosurgery* 1984;14:747-749.
- Reif J, Graf N. Intraspinous mesenchymal chondrosarcoma in a three-year-old boy. *Neurosurg Rev* 1987;10:311-314.
- Lee ST, Lui TN, Tsai MD. Primary intraspinal dura mesenchymal chondrosarcoma. *Surg Neurol* 1989;31:54-57.
- Ranjan A, Chacko G, Joseph T, Chandi SM. Intraspinous mesenchymal chondrosarcoma: case report. *J Neurosurg* 1994;80:929-930.
- Herman TE, McAlister WH, Dehner LP, Kaufman BA. Dedifferentiated chondrosarcoma in childhood: report of a case. *Pediatr Radiol* 1995;25(Suppl 1):S140-S142.
- Ogose A, Motoyama T, Hotta T, et al. Clear cell chondrosarcomas arising from rare sites. *Pathol Int* 1995;45:684-690.
- Rushing EJ, Mena H, Smirniotopoulos JG. Mesenchymal chondrosarcoma of the cauda equina. *Clin Neuropathol* 1995;14:150-153.
- Ohue S, Sakaki S, Kohno K, et al. Primary spinal chondrosarcoma localized in the cervical spinal canal and intervertebral foramen: case report. *Neurol Med Chir* 1995;35:36-39.
- Vanderhooft JE, Conrad EU, Anderson PA, Richardson ML, Bruckner J. Intradural recurrence with chondrosarcoma of the spine: a case report and review of the literature. *Clin Orthop* 1993;294:90-95.
- Marchi E, Antonico A Filho, Sette AA, Sette RC. Hemithorax as a primary manifestation of chondrosarcoma of the thoracic spine. *Rev Assoc Med Bras* 1992;38:177.
- Tan SK, Seow KH, Chiang GS, Sim CS. Chondrosarcoma of the spine: report of two cases. *Ann Acad Med Singapore* 1991;20:385-388.
- Gursel B, Yalciner G. Chondrosarcoma of the cervical vertebra: a case report. *Arch Otorhinolaryngol* 1987;244:74-76.
- Chan HS, Turner-Gomes SO, Chuang SH, et al. A rare cause of spinal cord compression in childhood from intraspinal mesenchymal chondrosarcoma: a report of two cases and review of the literature. *Neuroradiology* 1984;26:323-327.
- Ozaki T, Hillmann A, Blasius TS, Winkelmann W. Skeletal metastases of intermediate grade chondrosarcoma without pulmonary involvement: a case report. *Int Orthop* 1998;22:131-133.
- Crowell RM, Wepsic JG. Thoracic cord compression due to chondrosarcoma in two cousins with hereditary multiple exostoses: report of two cases. *J Neurosurg* 1972;36:86-89.
- Bergh P, Gunterberg B, Meis-Kindblom JM, Kindblom LG. Prognostic factors and outcome of pelvic, sacral and spinal chondrosarcomas: a center-based study of 69 cases. *Cancer* 2001;91:1201-1212.
- York JE, Berk RH, Fuller GN, et al. Chondrosarcoma of the spine: 1954 to 1997. *J Neurosurg (Spine 1)* 1999;90:73-78.
- Shives TC, McLeod RA, Unni KK, et al. Chondrosarcoma of the spine. *J Bone Joint Surg* 1989;71:1158-1165.
- Camins MB, Duncan AW, Smith J, Marcove RC. Chondrosarcoma of the spine. *Spine* 1978;3:202-209.
- Mandelli C, Bernucci C, Mortini P, Tartara F, Scomazzoni F, Giovanelli M. Chondrosarcoma of the thoracic spine: total en bloc sagittal resection. A case report. *J Neurosurg Sci* 2001;45:114-119.
- Marmor E, Rhines LD, Weinberg JS, Gokaslan ZL. Total en bloc lumbar spondylectomy: case report. *J Neurosurg* 2001;95:264-269.
- Hasegawa K, Ogose A, Kobayashi H, Morita T, Hirata Y. Simultaneous anterior-posterior approach for excision of malignant paraspinous tumor and subsequent reconstruction: technical note. *J Neurosurg* 1999;91:236-240.
- Doh JW, Halliday AL, Baldwin NG, Benzel EC. Spinal stabilization by using crossed-screw anterior-posterior fixation after multisegmental total spondylectomy for thoracic chondrosarcoma: case report. *J Neurosurg* 2001;94:279-283.
- Zibis AH, Markonis A, Karantanas AH. Unusual causes of spinal foraminal widening. *Eur Radiol* 2000;10:144-148.
- Yüntün N, Calli C, Zileli M, Üstün EE, Sener RN. Chondrosarcoma causing cervical neural foramen widening: case report. *Eur Radiol* 1997;7:1028-1030.
- Mummaneni PV, Rosenberg WS. Spinal chondrosarcoma following adenocarcinoma of the breast: case report. *Surg Neurol* 2000;53:580-582.
- Mackintosh D, Mason RM. Pharmacological actions of 17 $\beta$ -oestradiol on articular cartilage chondrocytes and chondrosarcoma chondrocytes in the absence of oestrogen receptors. *Biochim Biophys Acta* 1988;964:295-302.
- Prevedello DM, Koebel A, Tatsui CE, et al. Prognostic factors in the treatment of the intradural extramedullary tumors: a study of 44 cases. *Arq Neuropsiquiatr* 2003;61:241-247.
- Koebel A, Tatsui CE, Prevedello DM, et al. Prognostic factors in the treatment of intramedullary spinal cord tumors. *Arq Neuropsiquiatr* 2002;60:818-822.