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Partial Sacrectomy in the Management of Sacral Chordoma: Case Report and Literature Review

Sacrectomia parcial no manejo do cordoma sacral: Relato de caso e revisão da literatura

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Abstract

Chordomas are rare neoplasms of low to intermediate grades, which arise from ectopic remnants of notochordal tissue, presenting a slow growth pattern and locally aggressive behavior. Due to their insidious course, the diagnosis is late, requiring immediate therapeutic intervention. The main prognostic factor is total surgical resection with wide margins. This therapeutic objective is only achieved in 40% to 55.6% of the cases, since chordoma tends to present an aggressive behavior, invading adjacent tissues and neurovascular structures. Currently, the main challenge of sacrectomy is to balance a wide resection with the preservation of the neurological function of the patient. Despite cases of successful gross total resection, local recurrence is an inevitable reality, and the overall survival is relatively low. The indication of adjuvant therapies is not well stablished in the literature, since the response to radiotherapy is not satisfactory for these tumors. The aim of the present study is to present a report the case of a patient with sacral chordoma (SC) who underwent partial sacrectomy and to carry out a brief review of the literature on sacrococcygeal chordomas.

Keywords

- sacrectomy
- sacral chordomas
- partial sacrectomy

Resumo

Cordomas são neoplasias raras de graus baixo a intermediário, que surgem de remanescentes ectópicos de tecido notocordal, e apresentam padrão de crescimento lento e comportamento localmente agressivo. Devido ao seu curso insidioso, o diagnóstico é tardio, e necessita de intervenção terapêutica imediata. O principal fator prognóstico é a ressecção cirúrgica total com margens amplas. Esse objetivo terapêutico somente é atingido em 40% a 55,6% dos casos, pois o cordoma tende a apresentar um comportamento agressivo, pois invade tecidos adjacentes e estruturas neurovasculares. Atualmente, um dos principais desafios da sacrectomia é equilibrar uma ampla ressecção com a preservação da função neurológica do paciente. Apesar

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Palavras-chave
- sacrectomia
- cordomas sacrais
- sacrectomia parcial

dos casos de ressecção total bem-sucedida, a recorrência local é uma realidade inevitável, e a sobrevida global é relativamente baixa. A indicação de terapias adjuvantes não está bem estabelecida na literatura, uma vez que a resposta à radioterapia não é satisfatória para esses tumores. O objetivo deste trabalho é relatar o caso de um paciente com cordoma sacral (CS) submetido à sacrectomia parcial e fazer uma breve revisão da literatura sobre cordomas sacrococcígeos.

Introduction

Chordomas are rare malignant neoplasms of low to intermediate grades, which arise from ectopic remnants of notochord tissue, presenting a slow growth pattern and locally aggressive behavior.[1] They affect the midline along the neuroaxis, with preferential involvement of the sacrum (50% of the cases), followed by the spheno-occipital region (25% to 30%), the cervical vertebrae (10%), and the thoracolumbar vertebrae (5%).[2–4] Due to its invasive behavior, the lesion may infiltrate adjacent muscles, sacral nerve roots, pelvic viscera, and sacroiliac joints.[5]

They correspond to 1% to 4% of primary bone malignancies, representing the most common primary malignant bone tumor of the spine,[2,5] with male predominance in a ratio of 2:1.[1] The peak incidence is between 50 and 60 years of age,[2,4,5] and children and adolescents are rarely affected ($<$ 5% of the cases).[2] Chordoma has a relatively high mortality rate, with an average life expectancy of 5 to 7 years after the diagnosis, abbreviated to 1 year in individuals not submitted to adequate treatment.[6]

Case Report

We herein report the case of an 80-years-old female patient who, 3 years before, had started to feel severe sacrococcygeal pain (8/10 on the Visual Analog Scale, VAS), without irradiation and refractory to analgesia. The pain worsened progressively, reaching 10/10 on the VAS 3 months prior to hospital admission. Upon admission, the patient presented without neurological deficits, but with mild constipation.

A computed tomography (CT) scan showed an expansive lesion affecting the sacrococcygeal region with cortical rupture, associated with a large soft tissue component with an expansive infiltrative aspect, measuring $6.2 \times 6.6 \times 9.1$ cm (►**Figure 1**), with compression of the upper and middle thirds of the rectum, but without intestinal obstruction. The patient underwent an open biopsy of the sacral lesion, with the diagnosis of chordoma. During hospitalization, the patient evolved with a neurogenic bladder.

A partial sacrectomy (►**Figure 2**) was performed below S3 and the lesion was removed en bloc after retroperitoneal dissection in an area previously approached via the anterior route by general surgery. There was no injury to the sacral nerves. The immediate postoperative period was uneventful. At the last follow-up, two months after surgery, the patient maintained fecal incontinence and a neurogenic bladder, without neurological deficits. Adjuvant therapies were not indicated.

Discussion

Due to slow and indolent growth, sacral chordomas (SCs) generally remain clinically silent for long periods, delaying

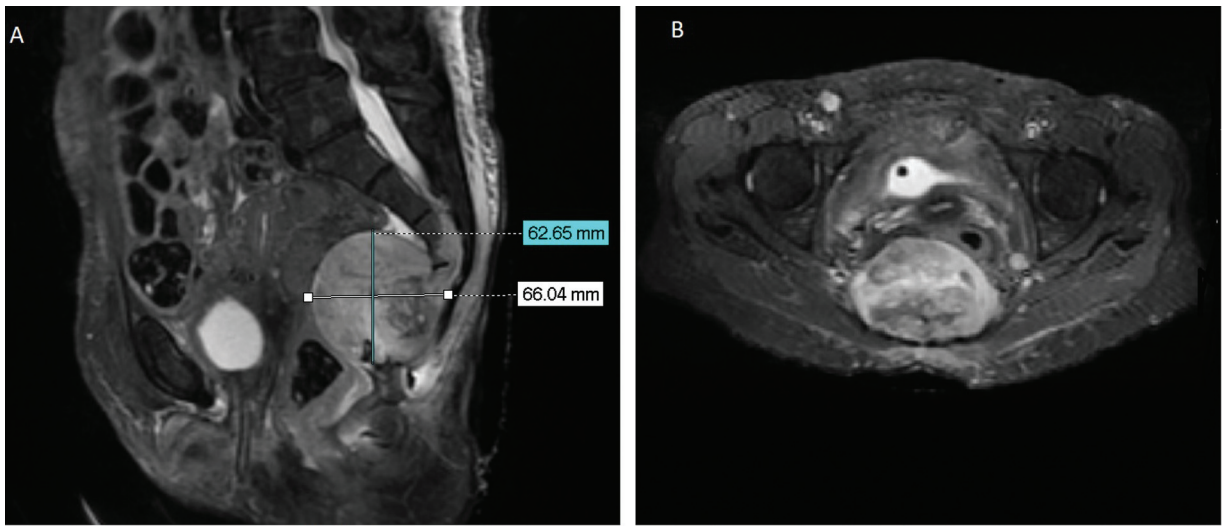

Fig. 1 Magnetic ressonance imaging scan in short tau inversion recovery (STIR) sequence showing a regular hypodense mass ventral to the sacrum (sacral chordoma), in (**A**) sagittal incidence and (**B**) axial incidence.

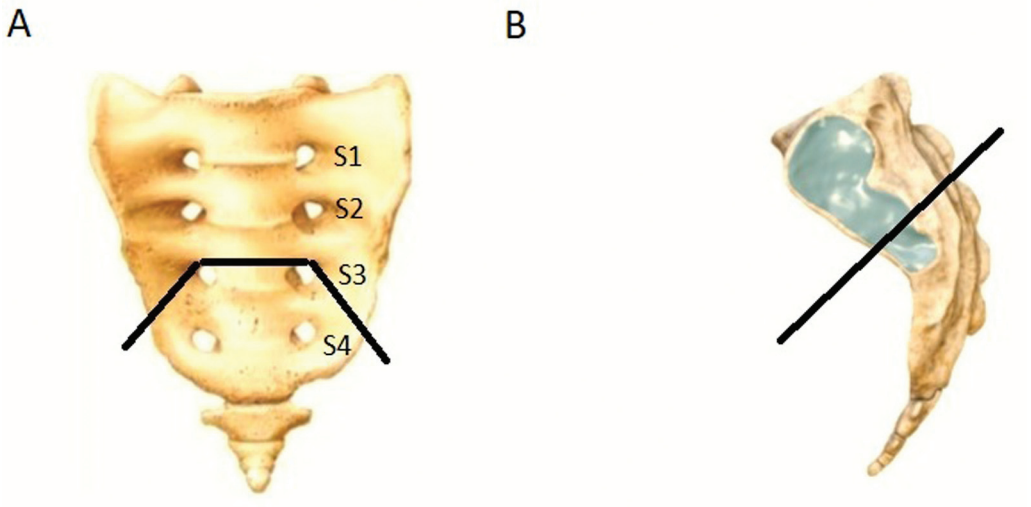

Fig. 2 Schematic representation of a partial sacrectomy in (**A**) coronal and (**B**) sagittal views.

diagnosis until 12 to 14 months from first manifestation.[1] Therefore, these tumors are often diagnosed incidentally or at an advanced stage, with destruction of adjacent structures and soft tissue invasion.[4] The main differential diagnosis is chondrosarcoma, a similar neoplasia both in terms of radiology and histology.

The most frequent clinical manifestation of CS is persistent low back pain. Compression of the sciatic nerve or the iliolumbar trunk causes radicular pain and nerve root involvement, ultimately leading to anorectal and urogenital neurological dysfunctions, with 1/3 of patients developing urinary tract infections and 10% manifesting constipation.[1,4]

In case of suspicion of SC, percutaneous needle biopsy is considered the gold standard.[1] Imaging exams are essential in the preoperative phase, for they enable the assessment of tumor location, staging, extension, and the relationship with adjacent structures. The detailed study of the local anatomy is essential for the success of the procedure and to achieve good functional outcomes.[7]

Enneking's principles define wide resection en bloc as the surgical strategy for SCs.[2,5] However, wide resection is a challenge, considering the aggressive behavior of the tumor and its infiltrative nature and poor marginalization.[8] Good preoperative planning with adequacy of imaging exams and a multidisciplinary approach regarding the surgical specialties (such as general, vascular and neurosurgery) become essential for a better outcome of wide tumor resection with maximum preservation of nearby noble structures[2]

Due to these challenges, a standardized description of the surgical technique has not yet been well established. The amplitude of sacral resection is based on the experience of the surgeon, lesion extension and nervous tissue infiltration.[3] Total sacrectomy is bilateral resection and fixation of the dural sac below S1, with resection extended to L5, L4, and to the iliac region, if necessary.[7] It normally occurs in two stages: an anterior approach and a posterior approach.[2] The anterior approach enables greater visualization and protection of visceral organs; however, in a recent multicenter study, it was a predictor of tumor recurrence. The posterior approach is single-staged, shorter in duration, and enables better handling of adjacent neural elements; however, it presents a greater risk of injury to large vessels and visceral organs.[5] The combined approach, in turn, is related to longer operative and recovery times.[6] Currently, the posterior approach has been more accepted,[4] mainly for disorders caudal to the S2 vertebra (caudal sacroiliac joints).[5] Partial sacrectomy is indicated over total sacrectomy with resection up to S1 when the printed margins below S3 can be reached.[6]

Marginal resection is achieved in only 35% to 81% of SCs, as it often involves nerve roots. Thus, there must be a balance between the chance of recurrence and the maintenance of the neurological function and the integrity of the visceral organs.[2,7]

Low sacral amputations (distal to the level of S3) tend to present a minimal deficit, with preservation of almost 100% of the permanent and intestinal function.[6] High sacral amputations and total sacrectomies cause greater spinosacral and sacropelvic instability and sexual and sphincter dysfunction, when S1 is bilaterally injured.[2] In some patients, embolization may be an alternative to reduce the extension of the tumor, minimizing the loss of additional neural function.[6]

On average, 60% of patients undergoing surgery develop complications associated with sacrectomy. Among the main complications, we can mention surgical site infection (the most common), wound dehiscence, cerebrospinal fluid (CSF) leak, sacral hernia, and failure in the musculocutaneous reconstruction process.[3,5,7] Radiotherapy (RT) may account for complications in the postoperative period.

The indication of pre-, post- or combined pre- and postoperative RT is controversial, with conflicting results in the literature, demonstrating the resistance of chordomas to adjuvant therapy and no clear association with better rates of local recurrence, metastatic disease or disease-specific survival. Radiotherapy remains reserved for unresectable patients and incomplete resection margins, considering

the surgical possibilities, the presence of associated comorbidities, and the patient's functional status.[5,7]

Sacrectomy is a procedure with high morbidity and mortality.[7] The rate of recurrence after surgery is of 40% to 50%,[9] with local recurrence 3 times higher in patients submitted to partial resection when compared to those who underwent total resection.[4] The indicators associated with poor oncological outcome are: inadequate surgical margins, history of recurrence after previous resection, infiltration affecting muscle and/or the sacroiliac joint, lesion larger than 8 cm in diameter, and locations above S3.[2,8] Despite the recurrences, tumor resection is essential for symptomatic relief of the patient and increased survival.

The risk of metastasis is relatively low, of around 5%.[1] The occurrence is mainly associated with local recurrence of the tumor, which tends to occur in later stages of the disease. It can affect adjacent areas, such as pelvic viscera and sacroiliac joints, and more distant sites, such as the lungs, liver and bone.[3,4,8] Despite the low tendency for metastasis to occur, approximately 40% to 60% of the patients develop distant metastases over the course of the disease.[9]

Currently, specific preoperative planning techniques, such as three-dimensional (3D) printed osteotomy, may be used for a better anatomical study of the region, with greater precision of the surgical procedure.[4]

Conclusion

Sacral chordomas are rare malignant neoplasms with an aggressive behavior. Total or partial sacrectomy is the main treatment of choice, being a complex procedure with high morbidity and mortality. The wide resection margin is the main prognostic factor, associated with a lower rate of local recurrence. Tumor recurrence is the most important predictor of metastasis and mortality. Preoperative planning with a multidisciplinary approach is essential to plan an accurate resection with a lower rate of complications. Chordomas are generally resistant to conventional RT treatments, which are indicated just in specific and individualized cases.

Conflict of Interests

The authors have no conflict of interests to declare.

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