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Review

A review of the surgical management of sacral chordoma

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Abstract

Background: Sacral chordomas are rare low-to-intermediate grade malignant tumours, which arise from remnants of the embryonic notochord. This review explores prognostic factors in the management of sacral chordomas and provides guidance on the optimal treatment regimens based on the current literature.

Patients and methods: Electronic searches were performed using MEDLINE, Embase and the Cochrane library to identify studies on prognostic factors in the management of sacral chordomas published between January 1970 and December 2013. The literature search and review process identified 100 articles that were included in the review article. This included both surgical and non-surgical studies on the management of sacral chordomas.

Results: Sacrectomy with wide resection margins forms the mainstay of treatment but is associated with high risk of disease recurrence and reduced long-term survival. Adequate resection margins may require sacrifice of adjacent nerve roots, musculature and ligaments leading to functional compromise and mechanical instability. Large tumour size (greater than 5–10 cm in diameter), dedifferentiation and greater cephalad tumour extension are associated with increased risk of disease recurrence and reduced survival. Chordomas are poorly responsive to conventional radiotherapy and chemotherapy.

Conclusion: Operative resection with wide resection margins offers the best long-term prognosis. Inadequate resection margins, large tumour size, dedifferentiation, and greater cephalad chordoma extension are associated with poor oncological outcomes. Routine long-term follow-up is essential to enable early detection and treatment of recurrent disease.

Keywords: Chordoma; Surgical management; Prognostic factors; Recurrence; Survival; Metastasis

Introduction

Chordomas are the fourth most common malignant neoplasms originating from bone and have an incidence of less than 0.1 per 100,000 people per year.[1] They arise from remnants of the embryonic notochord and occur exclusively within the axial skeleton.[2,3] The tumours occur most commonly within the sacrum (50–60%), followed by the spheno-occipital vertebrae (25–30%), cervical region (10%) and thoracolumbar vertebrae (5%).[3,4] Patients often present with advanced disease owing to the vague and indolent symptoms produced by these slow growing tumours deep within the pelvis.[5] Chordomas are poorly sensitive to conventional radiotherapy[6–8] and chemotherapy[9,10] and surgical excision forms the mainstay of treatment. Sacrectomy with wide resection margins offers the best long-term prognosis[1,7–9,11] but may require extensive nerve root and ligamentous excision leading to ambulatory, sexual, bowel and urinary incontinence.[5,10] There is a high risk of disease recurrence[9,12,13] and metastases occur in 5–40% of patients.[11,14–18] The overall 5 and 10-year survival rates following sacrectomy are 45–77% and 28–50% respectively.[1,8,19,20]

This review explores prognostic factors in the management of sacral chordomas and provides guidance on the optimal treatment regimens based on the current literature. The following outcomes of interest are discussed: Duration of presenting symptoms, adequacy of resection margins,

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surgical approach, chordoma size, chordoma location, dedifferentiation, muscular and sacroiliac joint infiltration, radiotherapy, chemotherapy, functional outcome, disease recurrence, metastases and survival.

Patients and methods

Electronic searches were performed using MEDLINE, Embase and the Cochrane library to identify studies on prognostic factors in the management of sacral chordomas. The following Medical Subject Headings (MESH) were used to carry out a systematic search of the literature: "Sacral chordoma," "Sacrectomy," "Radiotherapy," "Chemotherapy," "Functional outcome," "Disease recurrence," "Metastases," and "Survival." The initial search was further stratified using the individual prognostic variables: Duration of pre-operative symptoms, resection margins, surgical approach, sacroiliac and muscular infiltration, chordoma size, chordoma location, and dedifferentiation. Articles were selected based on pre-defined inclusion criteria and included literature published between January 1970 and December 2013 (Fig. 1). The reference lists from the retrieved articles were also reviewed for additional articles. In total, 100 articles were included in this study.

Results

Of the 100 studies included, 84 were retrospective studies, six were prospective cohort studies, and two articles reported on randomised controlled trials. A further five studies were review articles reporting on bone tumour classification and oncological behaviour. Three studies were descriptive reports of radiological and pathological findings in sacral chordomas. Studies on both operative and non-operative management of sacral chordomas were included. The review article is up to date with studies published up until 31st July 2013. Due to the rarity of the tumour, there was a lack of well-designed comparative studies. We therefore present an evidence-based review of current management.

Discussion

Presenting symptoms

Sacral chordomas most commonly present with poorly localised lumbar or gluteal pain.[15,21] This is caused by pressure of the tumour mass on the presacral fascia and anterior soft tissues within the rigid pelvis. Localised tumour infiltration into the vertebral foramina and pressure on adjacent sacral nerves may also produce urinary incontinence, bowel dysfunction and lower limb neurological compromise.[22] Approximately 30% of patients have radiculopathy involving the lower limb at presentation due to compression of the ipsilateral sacral nerve or iliolumbar trunk.[15,23]

Sacral chordomas are twice as common in men compared to women and are uncommon in individuals under 40 years of age.[19,24] The average duration of pre-

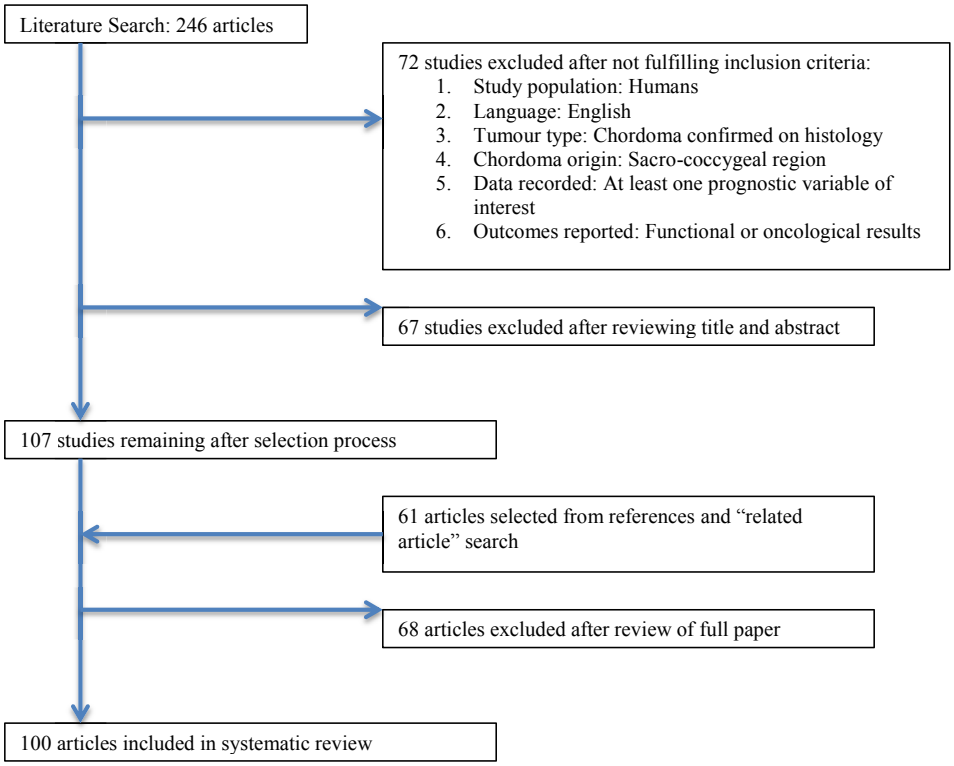

Figure 1. A flow chart showing the selection process of studies into the review article on management of sacral chordomas.