SÃO PAULO MEDICAL JOURNAL

CASE REPORT

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# Chordoma: retrospective analysis of 24 cases

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**Introduction:** Chordoma is a rare and slow-growing tumor, with local aggressiveness and preferential localization in the vertebral column. **Objective:** The main objective of this study is to evaluate natural history and results of treatment of chordomas. **Methodology:** This is a retrospective study from 1953 to 1993. **Material and methods:** The age ranged from 2 to 86 years (mean=34.5). Twelve patients were male and 12 female. The localization of the tumor was: 20 in the sacral region, 3 in head and neck and one out of the spine. **Results:** The treatment, alone or combined, was surgery, radiation therapy and chemotherapy. The survival rate for patients with lesions in the sacrum ranged from 4 to 119 months, since the date of the symptoms. The 5-year overall survival was 4.2%. **Conclusion:** Chordoma is a rare and slow growing tumor, with a very difficult approach by surgery due to its preferential location in the sacrum and poor therapeutic results with radiation therapy or chemotherapy, mainly in patients with advanced disease.

**UNITERMS:** Chordoma, sacral tumor, notochord, bone tumor, treatment.

## INTRODUCTION

Chordoma was first described by Virchow in 1857 under the name of "ecchordosis physaliphora".[17] In 1894, Ribbert defined chordoma as a malignant tumor arising from embryonic remains of the notochord.[12]

The preferential localization of chordoma is in the vertebral column, from the spheno-occipital synchondrosis to the coccyx, more frequently in the sacrococcygeal region.[9]

The majority of chordomas are slow growing with local proliferation and high aggressiveness and infiltration of the adjacent organs and structures (Fig. 1). Surgery is the only curative treatment [7,8,9,13] when adequate margins are obtained. There is a low rate of metastases for this type of tumor.[7]

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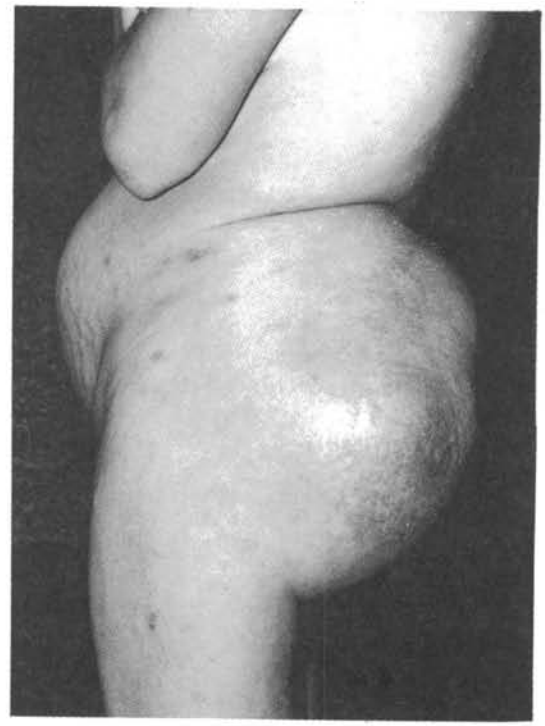

**Figure 1** - Female patient with a large tumor bulging the posterior region of the pelvis.

## MATERIALS AND METHODS

A series of 24 patients with chordomas was identified in the Hospital files (1953-1993), with ages ranging from 2 to 86 years (mean=34.5), 12 males and 12 females; 22 were white. The anatomical location was: sacrum-20 cases (83.4%), 8 with extension to the coccyx and 5 invading to the lumbar column; head and neck-3 cases (12.4%) in the clivus region; retroperitoneum-1 case (4.2%). A summary of clinical data, treatment and follow-up is shown in Table I.

When ultrasonography, computerized tomography and magnetic resonance were not available, the diagnosis of tumors in the sacral region were made through digital rectal examination and barium enema. In the absence of imaging exams, the diagnosis of chordomas of other locations were very difficult due to the lack of other trustworthy methods.

Our therapeutic approach for chordomas is curative surgery followed by adjuvant radiation therapy and/or chemotherapy. This conduct was not always possible in our series because 13 out 24 cases were previously submitted to various types of treatment with residual or recurrent disease, and the majority of the intact cases had advanced tumors. In two cases, due to this reason, no treatment was possible.

Radiation therapy, combined or alone, was the treatment in 16 (66.7%) of the cases, being 9 (37.5%) associated with surgery and 5 (20.9%) with chemotherapy. Chemotherapy was the exclusive treatment in the case of the retroperitoneal tumor.

## RESULTS

In the 20 cases with tumor in the sacral region, the treatment and overall survival were: a) radiation therapy alone (5 cases) - 5 to 50 months (mean-18.8); b) radiation therapy and surgery (4 cases) - 1 to 88 months (mean-27.8); c) radiation therapy, chemotherapy and surgery (4 cases) - 6 to 45 months (mean-24.8); d) surgery alone (3 cases) - 11 to 17 months (mean-14); e) chemotherapy and surgery (1 case) - 27 months; f) chemotherapy and radiation therapy (1 case) - 31 months. Two patients did not receive any treatment due to the advanced stage of the disease.

In the 3 cases with tumors in the head and neck region (ages 10,11 and 21 years old), the first was treated with surgery and radiation therapy and has been disease free for a 21-month period; the second was submitted exclusively to radiation therapy, dying due to the disease after 11 months, and the third was treated with surgery alone and has been disease free for a 3-month period.

The patient with tumor in the retroperitoneum was submitted exclusively to chemotherapy, dying 11 months after admission.

From the total of 24 cases, eight (33.4%) have developed metastases in the following sites: lung- 5; liver, lymphonodus and bone- one case each. The 5-year overall survival is shown in Figure 2.

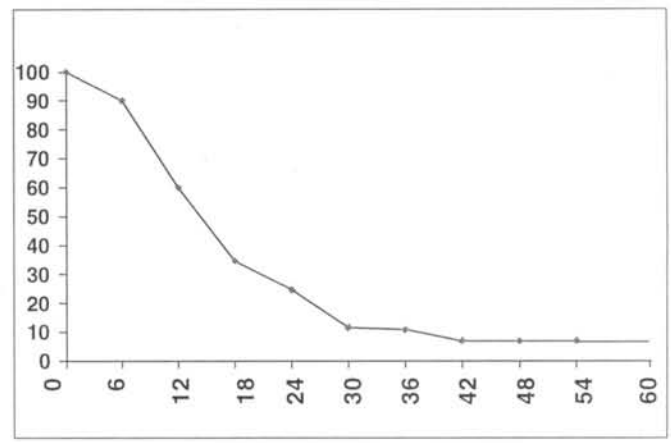


**Figure 2 -** Five-year overall survival

Chordoma is a rare neoplasm even in a cancer center. The present series of 24 cases represented 0.02% of all 79,917 new cancer cases seen at the A.C.Camargo Hospital (1953-1993). Among the 1,030 cases of bone tumors in the same period, chordomas corresponded only to 2.2%.

The chordoma can arise from all axial skeleton, but various authors show that 50% begin in the sacrum, 35% in the spheno-occipital region and 15% in the vertebrae.[3,6,7,19] The localization out of the spine is rare.[9] In this series, the majority of the tumors were in the sacral region, but we must remember that before the introduction of computerized tomography, it was difficult to obtain the diagnosis of spheno-occipital lesions.

The time elapsed between the initial symptoms and the treatment is usually very long, which is a characteristic of a slow-growing tumor. Presence of mass and sacral pain, with rectal and urinary bladder dysfunction are the main symptoms of the disease in the sacrococcygeal region.[15] The digital rectal examination is a very important propedeutic method for diagnosis of sacral tumors.

In spheno-occipital lesions the main signs and symptoms are pain, mass-invading nasopharynx,

**Table I**
**Characteristics of the patients, treatment and follw-up**

| # | Age (yr.) | Sex | Duration of symptoms | Symptoms | Site | Admission stage | Previous treatment | Institution treatment | Follow-up |
|---|---|---|---|---|---|---|---|---|---|
| 1 | 55 | m | 96 mo. | local pain narrow feces rectal / bladder dysfunction | sacral | liver metastases advanced sacral tumor | biopsy | no | dead after 13 mo. |
| 2 | 2 | f | 2 mo. | sacral tumor loss of weight | sacral | inguinal lymphonodus advanced sacral tumor | biopsy | RT(36Gy) | dead after 5 mo. |
| 3 | 2 | f | 24 mo. | sacral tumor and pain | sacral | lung metastases advanced sacral tumor | surgery | RT(36Gy) CT(act) | lost after 8 mo. with disease |
| 4 | 28 | f | 72 mo. | sacral pain with Irradiation to thighs and legs | sacral | lung metastases advanced sacral tumor | surgery | surgery RT(36Gy) CT(act) | dead after 6 mo. |
| 5 | 40 | m | 60 mo. | sacral pain with irradiation to thighs and legs | sacral | advanced sacral tumor | surgery | RT(50Gy) CT(cyc+5F) | dead after 40 mo. |
| 6 | 62 | m | 84 mo. | sacral tumor and pain rectal / bladder dysfunction | sacral | advanced sacral tumor | surgery | CT (5FU+act+ vin) | dead after 27 mo. |
| 7 | 25 | f | 5 mo. | sacral tumor and pain | sacral | advanced sacral tumor | surgery | RT(36Gy) | dead after 10 mo. |
| 8 | 44 | m | 24 mo. | sacral and leg pain | sacral | sacral tumor | surgery | RT(50Gy) | dead after 88 mo. |
| 9 | 63 | m | 76 mo. | large sacral tumor and pain | sacral | lung metastases advanced sacral tumor | surgery RT(?Gy) | CT (cyc+5FU+ MTX) | dead after 45 mo. |
| 10 | 10 | f | 36 mo. | neck pain,cough cranial nerves dysfunction (III, IV,VI,VII,XII) | clivus | large cervical tumor with respiratory obstruction | biopsy | tracheosto-my RT(70Gy) | dead after 11 mo. |
| 11 | 69 | m | 4 mo. | sacral pain and rectal disfunction | sacral | lung metastases advanced sacral tumor | no | RT(60Gy) | dead after 20 mo. |
| 12 | 42 | m | 24 mo. | sacral pain and tumor bladder isfunction | sacral | advanced sacral tumor | biopsy | RT(30Gy) | dead after 12 mo. |
| 13 | 24 | f | 5 mo. | sacral pain and loss of weigh | sacral | lung metastases advanced sacral tumor | surgery RT(50Gy) | no | dead after 1 mo. |
| 14 | 54 | m | 24 mo. | sacral tumor and pain, loss of weigh | sacral | advanced sacral tumor | biopsy | no | lost after 1 mo. with disease |
| 15 | 86 | f | 7 mo. | sacral pain | sacral | sacral tumor | no | RT(50Gy) | dead after 50 mo. |
| 16 | 39 | m | 4 mo. | sacral tumor and pain, with irradi-ation to thighs | sacral | bone metastases sacral tumor | biopsy RT(40Gy) | RT(40Gy),CT (vin+adr+cyc+ cis+dti) | dead after 31 mo. |
| 17 | 6 | f | 18 mo. | sacral tumor and pain | sacral | advanced sacral tumor | surgery | no | dead after 14 mo. |
| 18 | 37 | m | ? | sacral pain | sacral | advanced sacral tumor | no | palliative surgery | lost after 17 mo. |
| 19 | 23 | m | 168 mo. | sacral tumor and pain, leg pain, rectal / bladder dysfunction | sacral | advanced sacral tumor | biopsy RT(60Gy) | no | lost after 7 mo. with disease |
| 20 | 11 | m | 12 mo. | neck pain | clivus | cervical tumor | no | surgery RT(45Gy) | disease free after 21 mo. |
| 21 | 46 | f | 46 mo. | sacral tumor | sacral | small sacral tumor | no | surgery | lost, disease free after 11 mo. |
| 22 | 21 | f | 12 mo. | neck pain,cough cranial nerves dysfunction (VI, IX,X,XI,XII) | clivus | large cervical tumor with respiratory obstruction | surgery | surgery | disease free after 3 mo. |
| 23 | 18 | f | 2 mo. | abdominal pain | out of spine | retroperitoneal tumor | biopsy | CT(vbl+cis+ ble) | dead after 11 mo. |
| 24 | 23 | m | 18 mo. | leg pain, rectal dysfunction | sacral | advanced sacral tumor | palliative surgery | RT(50Gy) | dead after 12 mo. |

m-male, f-female, RT-radiation therapy, CT-chemotherapy, act-actinomycin, cyc-cyclophosphamide, vbl-vinblastine 5FU-5 fluorouracil, vin-vincristine, MTX-methotrexate, adr-adriamycin, cis-cisplatin, dti-dacarbazine, ble-bleomycin

sometimes with obstruction of air passage and dysfunction of cranial nerves.[8]

The metastases rate found in our series (33.4%) is within the range described in the literature.[4,8,9,18]

Conventional x-ray, computerized tomography and magnetic resonance are very important in the diagnosis and to plan the better treatment. In the histopathology, the presence of large cells with lobular arrangement and intracytoplasmatic mucus, called physaliphora cells, is characteristic of chordomas (Fig. 3).

Surgery with adequate margins is the best available treatment for chordomas, but this is not always possible due to the advanced stage of the disease and difficult access[5] (Fig. 4).

The extent of curative surgery is a dilemma, because the total removal of the tumor many times implies rectal and urinary bladder impairment.

The results of radiation therapy alone are not satisfactory, however is indicated in the absence of better therapeutic options[8,18] and the dosage varies from 70 to 80 Gy.[11,16]

The chemotherapy (PVB scheme - cisplatin, vinblastine, bleomycin) was used in our retroperitoneal case of chordoma and was unsuccessful. Azzarelli reported one case with complete remission with chemotherapy (PVB scheme) that allowed surgical treatment. However, this patient had local recurrence 3 months later.[2]

A 50% five-year survival rate is reported by Smith in patients with sacral lesions.[14] In the Queen Elizabeth Hospital series no difference was found between patients submitted to palliative radiation therapy and those without treatment.[4] The same author reported a 5-year survival rate between 50% and 60% for cases submitted to surgery with adequate margins followed by radiation therapy, 25% for cases submitted to palliative radiation therapy and 15% for patients submitted to palliative surgery.[4] Keisch reported a 5-year survival rate of 100% for cases submitted to surgery with adequate margins and radiation therapy, and 68% for cases submitted only to surgery with adequate margins.[10] This excellent result indicates the necessity of combined treatment and early diagnosis. The five year overall survival reported by some authors are shown in Table 2.

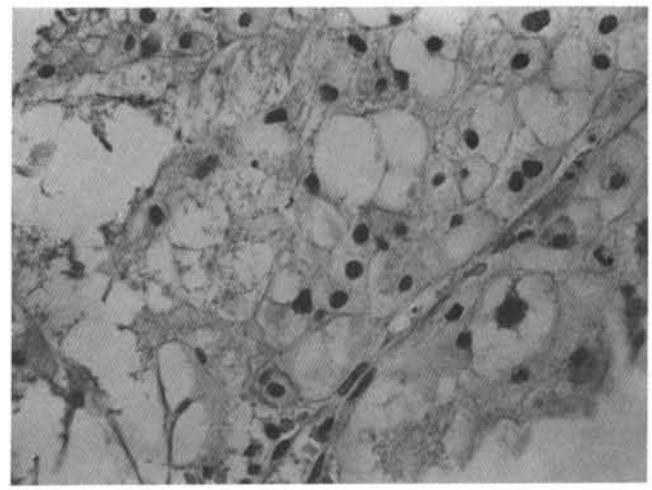

**Figure 3** - Chordoma cells with cytoplasmatic vacuolization. Classical physaliferous large cell on the right side (hematoxylin-eosin stain magnification x400).


**Figure 4** - Necropsy specimen of a large sacrococcygeal chordoma invading adjacent structures of the pelvis, destroying equine tail.

**Table 2**
**Five-year overall survival reported by some authors for chordomas**

| Author | # cases | 5 yr(%) |
|---|---|---|
| Keisch et al(1991) | 21 | 74 |
| Rich et al(1985) | 48 | 50 |
| Smith et al(1987) | 60 | 50 |
| Chetiyawardana(1984) | 69 | 40 |
| Azzarelli et al(1988) | 33 | 30 |
| This series | 24 | 4.2 |

The 4.2% five year survival obtained in our series is an indication of the advanced stage of the disease and recurrence after previous inadequate treatment.

Since chordomas are a slow-growing and rare tumors, multidisciplinary approach and early diagnosis are required for adequate treatment.


## Resumo

**Introdução:** O cordoma é um tumor raro e de crescimento lento, com agressividade local e localização preferencial na coluna vertebral. **Objetivo:** O objetivo principal deste estudo é analisar a história natural e os resultados de tratamento dos cordomas. **Metodologia:** Este é um estudo retrospectivo realizado entre 1953 e 1993. **Material e Métodos:** A idade variou de 2 a 86 anos (média=34,5). Doze pacientes eram do sexo masculino e 12 do sexo feminino. A localização do tumor era: região sacral em 20 casos, cabeça e pescoço em 3 casos e fora da coluna em um caso. **Resultados:** O tratamento, sozinho ou combinado, foi cirurgia, radioterapia e quimioterapia. A sobrevida para pacientes com lesões no sacro variou de 4 a 119 meses, desde a data dos sintomas. A sobrevida global em 5 anos foi de 4,2%. **Conclusão:** Cordoma é um tumor raro e de crescimento lento, com uma abordagem terapêutica cirúrgica difícil devido a sua localização preferencial no sacro e resultados pobres com radioterapia ou quimioterapia, principalmente em pacientes com doença avançada.


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