Sacrococcygeal Chordoma: Combined Surgical Approach In An 86-Year-Old Male

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Citation

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Abstract

A case is presented of an extensive sacrococcygeal chordoma in an 86-year-old male with an untreatable rectal pain syndrome. The diagnosis was suspected by imaging tests and confirmed by transperineal biopsy. The patient underwent surgery in a single operation using the combined approach, first anterior, to perform a colostomy, posterior rectal dissection and ligation of hypogastric arteries, then in the prone position for resection of the sacrum at S1-S2 together with the tumour. One year after the operation the patient remains disease-free. The aspects of diagnosis and treatment are discussed.

INTRODUCTION

Chordomas are very uncommon bone tumours derived from the primitive notochord and preferentially located in the sacrococcygeal region, with slow but progressive infiltration of the nerve roots and neighbouring soft parts. Because of their location they are difficult to manage.

CASE REPORT

We present a case of sacral chordoma in an 86-year-old patient who consulted for clinical features, over the previous 6 months, of lumbosacral pain irradiating to both legs and rectal tenesmus, which had worsened and not found relief with strong oral analgesics. Rectal palpation revealed an extramucous tumour on the posterior wall of the rectum, which was painful to the touch.

Endorectal ultrasonography demonstrated a wellencapsulated heterogeneous presacral tumour in close contact with, but without infiltrating, the rectal wall (Figure 1).

Figure 1

Figure 1: Endorectal ultrasonography. Well-encapsulated heterogeneous presacral tumour.

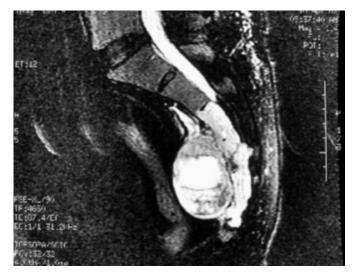


CT and NMR were also done (Figures 2 & 3), which revealed the existence of a well-encapsulated tumour, 5 cm in diameter, de-structuring the distal half of the sacrum, but without affecting the rectum, which suggested chordoma.

Figure 2: CT. 5 cms well-encapsulated tumour destructurating the distal half of the sacrum.

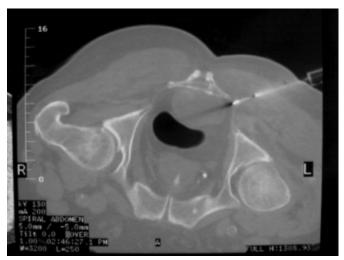


Figure 3: NMR. Sagital view appreciating presacral tumor.



The diagnosis was confirmed by CT-guided FNAB (Figure 4).

Figure 4 Figure 4: CT-guided FNAB.



Since the patient presented with pain and tenesmus requiring continuous i.v. infusion of morphine, surgical treatment was decided on despite his age. This was done in two parts in a single operation: first via the anterior approach, in which the internal iliac vessels were ligated and dissected extraperitoneally, the posterior rectum dissected, with a compress placed between the rectum and anterior part of the sacrum, and a rifle-barrel left colostomy performed; the second part was via the posterior approach, in which the sacrum was resected at the S1-S2 articulation (preserving both S1 sacral roots) and a polypropylene mesh fitted to close the intergluteal defect (Figures 5 & 6).

Figure 5 Figure 5: Surgical specimen.



Figure 6

Figure 6: Polypropylene mesh fitted to close the intergluteal defect



Pathological anatomy of the specimen informed of chordoma with areas of chondroid differentiation massively infiltrating the sacrum. One year after the operation the patient is asymptomatic, with no evidence of recurrence.

DISCUSSION

Chordomas are uncommon tumours, accounting for 3% of all bone tumours₁. They can occur in both sexes at any age, although they are more frequent in males in their 5th to 7th decades of life². Although they may appear on any point of the neuro axis, more than half of the cases have a sacrococcygeal location¹,₂. The clinical features of our patient are typical (lumbar pain associated with anorectal or vesical symptoms), and it is very common for diagnosis to be late, at times after more than a year of insidious discomfort, due to their slow growth rate₃. As for diagnosis, suspicion is established by rectal palpation and confirmed by CT and/or NMR, in which nearly always bone erosion or infiltration of neighbouring soft tissues is observed₄.

Controversy exists as to whether or not to indicate preoperative biopsy in the case of resectable tumours, as it may cause spread and complications (especially if done transrectally)₅. In our case it was done perineally and was complication-free, indicated by the need for a definite diagnosis before surgery due to our patient's old age and surgical risk. Metastases are rare, between 10% and 30%, the most common being in the skin, lung and vertebral bodies₆. As for treatment, there is unanimity on the indication of surgery, since management of the pain is very difficult, even with intrathecal morphine₇. Although some authors prefer the exclusively posterior approach₈, we advocate the

combined approach (anterior and posterior) as it allows a better control of the internal iliac vessels and enables us to perform a temporary colostomy for protection (both in the hypothetical need for rectal excision and with the risk of incontinence due to excision of the 1st sacral roots), and separate the rectum from the presacral space.

Removal should aim to be extensive and complete because, if not, recurrence is certain⁴,⁸. The ideal location for sacral dissection, whenever possible, is the S2-S3 junction (preserving the S2 roots) in order to avoid incontinence. If both S2 roots are dissected, as was necessary in our patient, incontinence is unavoidable²,⁸. When complete excision is not possible, high doses of adjuvant radiation therapy must be administered³, 4. Some authors recommend it systematically after surgery, even in cases in which resection has been complete²,₉. Chemotherapy gives poor results². Although prognosis is generally poor, due to the fact that being multifocal they have a major tendency to recur, it depends on resectability and histology, those with chondroid differentiation (as in our case) having a better prognosis than those with a dedifferentiated pattern₁₀. The percentage of disease-free patients at 5 years ranges from 30% to 40%9, and 5-year survival rates of between 4% and 75% have been reported5.

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