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Case Report

Surgical Treatment of Sacral Chordoma: The Role of Laparoscopy

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Abstract
Sacral chordoma is a rare tumour that represents the most common malignancy of the sacral region. Its diagnosis can be delayed because of unclear clinical manifestation. This tumour can involve surrounding anatomical structure such as the rectum, and its surgical treatment is still challenging. We report on 3 patients with sacral chordoma. Two of them were successfully treated using a laparoscopic approach and one by open surgery. We present all details of the surgical technique and patients' outcome. Minimally invasive methods in the surgical treatment of chordoma allow to perform a radical dissection of the tumour, minimizing the operative trauma. A laparoscopic approach can be considered safe and radical for sacral chordoma treatment.

Introduction

Chordoma is a rare tumour that develops from embryonic notochordal cell remnants. It has a slow growth rate that hinders its early diagnosis [1]. Usually, clinical presentation is unclear with uncertain symptoms that often lead to a working diagnosis of another more frequent disease. In 29–50% of all cases, this tumour involves the sacral region, and it affects men twice as often as women [2].

Even though sacral chordoma is considered to be a benign tumour, it can evolve with time, representing the most common malignant tumour of the sacral region. Sometimes chordomas are diagnosed when they are already at an advanced stage with distant metastasis to the lymph nodes, liver, bones, lungs, brain, soft tissues and/or the peritoneal cavity [3].

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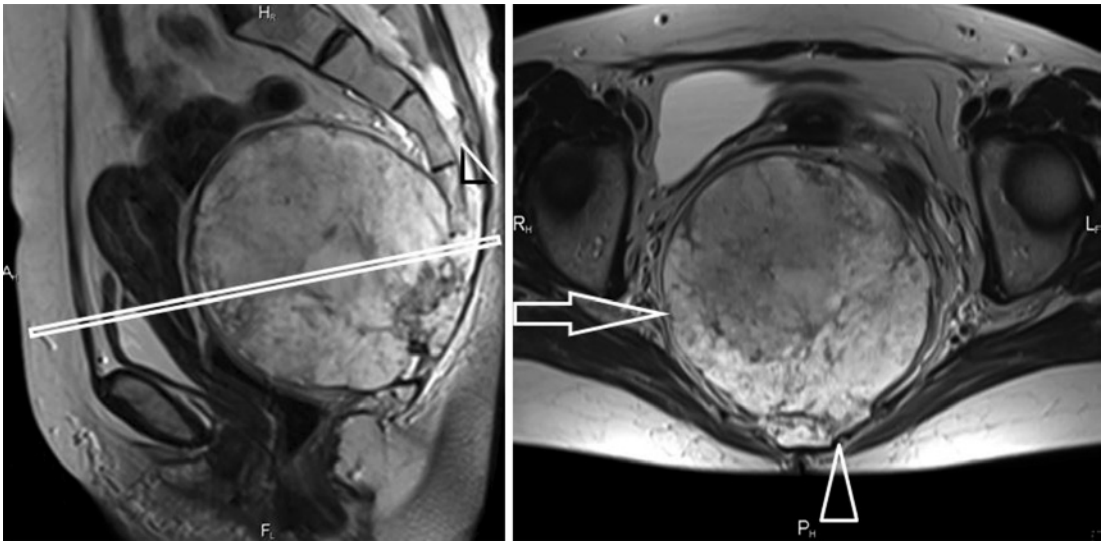

Fig. 1. MRI T2WI. Left panel, sagittal slice demonstrating large heterogeneous hyperintense signal of tumour located in a female pelvis, which pushes on the pelvic organs and involves the sacrum. The thin oblique rectangle indicates the position of the axial slice. Right panel, the arrow shows the tumour, the arrowhead shows the sacral invasion. Bone marrow biopsy was carried out. The histological results revealed chordoma. Two courses of radiotherapy (12.25–15.4 Gy) were given. The third course was interrupted because of a peptic ulcer. This was treated by common antiulcer medications.

Surgical resection plays a key role in the treatment of sacral chordoma, and there is a significant difference in survival between operated patients and those who do not undergo surgery [4, 5]. Surgical treatment of primary sacral tumours can be challenging, considering the complex surrounding anatomical structures and the (generally) large size of the tumour [6].

Here, we reported our experience with the surgical treatment of patients with sacral chordoma by open and laparoscopic approaches.

Cases Presentation

Case 1

A 55-year-old female with a sacrococcygeal tumour came to our attention presenting with chronic pain in the back. The tumour was identified accidently during a gynaecological examination. In 2010, the patient had hit her back. There were no pathological changes visible on her X-ray results after that injury. Since 2011, the patient had been suffering from back pain while standing, walking, or remaining in the same position for a long time. An MRI of the pelvis showed a tumour in the vertebral part of the sacrum in correspondence of S4 vertebra, distracting the lower and the upper sacrum vertebrae, and expanding ventrally to the pelvic. The size of the tumour was 10 × 10.8 × 10.5 cm. The tumour had well-defined margins, and it compressed the rectum to the left and forward. Also, the uterus was displaced (Fig. 1).

Firstly, the abdominal step was performed. After creating a pneumoperitoneum with a Veress needle, all trocars were positioned (Fig. 2).

The pelvic peritoneum was dissected from the rectum in the space between the common iliac artery and the base of the rectosigmoid junction mesentery. A further dissection of the tissues was performed along the mesorectal fascia to the upper part of the chordoma. The tumour was identified in the sacral region. Therefore, an incision was done between the

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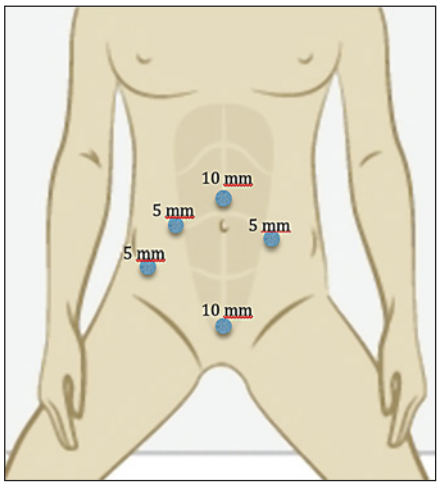

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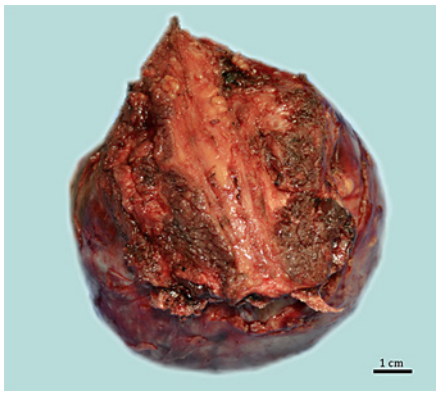

Fig. 2. Scheme of the trocar ports' positions.
Fig. 3. The chordoma with the distal part of the sacrum after the resection.

rectum and the tumour until complete separation of the chordoma from the mesorectal fascia. There was no connection between the tumour and the walls of the pelvis and the main branches of the iliac vessels.

The perineal step started after rotating the patient into the jack-knife position. The skin above the sacrum was dissected performing a linear longitudinal cut from the distal edge of the coccyx up and 5 cm above the planned sacrum resection line. Then, the subcutaneous tissue was also dissected in the direction of the lateral margins of the sacrum. Therefore, the superficial fascia, the medial part of the gluteus maximus muscle, the sacrococcygeal, sacrospinal, and sacrotuberous ligament were dissected. The sacroiliac ligament and a segment of the pyriform muscle were dissected on each side. When the mobilization of a lateral surfaces was extended to the sacroiliac joints, the perineal access was connected to the abdominal one.

Then, the sacrum was dissected using a liston bone cutter at the level of S3. The tumour was dissected from the adjacent structures. The chordoma and the distal part of the sacrum were resected (Fig. 3). The perineum was sutured.

Operative time was 180 min. Blood loss was 300 mL. Length of hospital stay was 26 days. Postoperatively, the patient had urinary dysfunction, which resolved 20 days after the operation. After 40 months of follow-up, the patient is still alive with no signs of recurrence.

Case 2

A 71-year-old female reported a trauma in the sacral region 3 years before coming to our attention. She was followed up by a neurologist, and she underwent physiotherapy protocol. Six months after the accident, the patient reported severe back pain and underwent a CT scan. On the CT scan, a tumour of 10 cm diameter, spreading to the sacrococcygeal junction, was found. The lesion was solid, immobile, and tender upon palpation. An extra-rectal tumour behind the back wall of the rectum, 4 cm from the anal verge, was identified during the digital rectal examination. Biopsy revealed a sacral chordoma.

Surgery was performed with an open approach starting with a median incision till the pubis on the abdomen. The rectum and the rectosigmoid junction were mobilized sparing the hypogastric plexus until the upper margin of the tumour. The dissection was continued up to the level of the pelvic floor muscles. The patient was turned into the jack-knife position for the perineal stage – performing the same steps as described in Case 1.

Operative time was 180 min. Blood loss was 600 mL. Length of hospital stay was 35 days. The patient had no postoperative complications. After 84 months of follow-up, the patient is dead for recurrence.

Case 3

A 35-year-old male came to our department complaining of rectal pain and reporting constipation from 6 months. MRI detected a large tumour (90 × 99 × 110 mm) with multiple septa inside which originated from the sacrum. This tumour pushed forward the levator ani muscle and the rectum. Total body CT scans did not detect distant metastases. Digital rectal examination revealed an immobile tumour with the smooth surface 5 cm from the anal verge. Histopathological examination confirmed sacral chordoma. Longitudinal marks were attached on the skin surface in the sacrum area as in Case 1. During the MRI procedure, the distance from the sacrococcygeal joint to the place of skin incision was measured. The surgical treatment followed the same steps as reported in Case 1.

Operative time was 200 min. Blood loss was 200 mL. Length of hospital stay was 24 days. The patient had no postoperative complications. After 16 months of follow-up, the patient is still alive with no signs of recurrence.

Discussion

Surgical treatment of a sacral chordoma is challenging, since it can involve some critical structures such as nerves and arteries. Usually, a sacral chordoma develops silently and insidiously, and there is a considerable time-lapse between the onset of symptoms and diagnosis. In most of cases, back pain is the main symptom reported by patients.

Conventional X-ray in the sacrococcygeal region can identify the tumour, but MRI is the best imaging method for evaluating the extension and the involvement of the surrounding soft tissue. Prognosis of the patients is heavily depends on the structures involved by the tumour (there is a higher recurrence rate for gluteal invasion) and on the extent of the surgical resection (en bloc resection determines a lower rate of recurrence) [7, 8]. The majority of primary malignant sacral tumours do not respond to common radiotherapy and chemotherapy, and only extensive surgical resection represents a potential curative treatment [9]. However, radiotherapy can be useful, and it can give temporary benefit to patients who underwent inadequate surgery or in case of unresectable tumours [10]. Currently, complete en bloc resection of the tumour with clean margins seems to provide the longest survival [11, 12]. Furthermore, the operation can be associated with many complications and morbidities such as massive bleeding, wound dehiscence, surgical site infection, neurogenic bladder or bowel, damage of the sacral plexus and sciatic nerve [13].

Even if a sacral chordoma rarely invades the rectal wall due to the periosteum and presacral fascia, surgical dissection can be difficult leading to injuries of the neurovascular structures, rectum, and bladder. The optimal surgical approach for excising the tumour is quite debated in the literature, and some authors propose an exclusively posterior approach [14], while others demonstrate the efficacy of a combined anterior and posterior approach [15]. The posterior approach can be indicated for very caudal tumours affecting the third sacral section, while the combined approach (abdominal and posterior) is useful for more cranial lesions. According to Woodfield et al. [16], sacrococcygeal tumours larger than 10 cm should be dissected using a combined approach with a safe dissection of visceral organs away from the tumour.

In this case series, we reported our experience with the combined approach. We performed laparoscopic surgery for the abdominal steps in 2 patients. The possibility to have

a magnification of the view during laparoscopy allows to perform a precise dissection preserving all structures very close to the tumour and reducing the risk of bleeding. In our cases there were no severe complications during the postoperative period except for bladder dysfunction in Case 1 which resolved within 2 months after surgery.

Conclusion

Minimally invasive methods in the surgical treatment of chordoma allow to perform a radical dissection of the tumour minimizing the operative trauma. Two of our cases reflect the successful use of a combined approach using a laparoscopic technique for the treatment of sacral tumours.

Statement of Ethics

Written informed consent was obtained from the patients for the publication of this case report series and any accompanying images.

Disclosure Statement

The authors certify that they have no conflicts of interest to disclose on the subject matter discussed in this paper.

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Author Contributions

S.K. Efetov: second surgeon, conception of the study design, data collection, analysis, manuscript writing, revision, and manuscript submission. A. Picciariello: conception of the study design, data collection, analysis, manuscript writing, and revision. V.S. Kochetkov: data collection, analysis, and participation in the care of the patient. K.B. Puzakov: conception of the study design, manuscript writing, and revision drafting. A.V. Alekberzade: data collection, analysis, and critical revision of the manuscript. I.A. Tulina: conception of the study design, data collection, analysis, manuscript writing and revision, and critical revision of the manuscript. P.V. Tsarkov: first surgeon, conception of the study design, data collection, analysis, manuscript writing and revision drafting, and approval of the final version of the manuscript for submission.

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