

## Case Report

# Total Penectomy for Recurrent Chordoma of the Corpus Cavernosum

Lennert Eismann <sup>1</sup>, Sabine Kess,<sup>2</sup> Christian G. Stief,<sup>1</sup> and Frank Strittmatter<sup>1</sup>

<sup>1</sup>Department of Urology, Ludwig-Maximilians-University, Munich, Germany

<sup>2</sup>Department of Gynecology and Obstetrics, University Hospital of Heidelberg, Heidelberg, Germany

Correspondence should be addressed to Lennert Eismann; [lennert.eismann@med.uni-muenchen.de](mailto:lennert.eismann@med.uni-muenchen.de)

Received 2 December 2019; Accepted 9 January 2020; Published 13 February 2020

Academic Editor: Francesco M. Solivetti

Copyright © 2020 Lennert Eismann et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Chordomas are rare low malignant neoplasm arising from remnants of the notochord with predilection site of the clivus or the os sacrum. Standard therapy is radical excision and adjuvant radiation. Due to invasive growth and adjacent to vital structures resection is often incomplete, and therefore, local recurrence is frequent. First, to the best knowledge of our authors, we present a 70-year-old man with a recurrent chordoma infiltrating the corpus cavernosum. Asymptomatic recurrence was diagnosed by magnet resonance imaging according to the standard follow-up. Our interdisciplinary tumor board recommended surgical resection. We performed a total penectomy and perineal urethrostomy to achieve negative resection margins and preserve best quality of life for the patient.

## 1. Introduction

Chordoma is a rare malignancy with an incidence reported up to 0.08/100,000 person per year with a predominance in male patients according to the Surveillance, Epidemiology, and End Results (SEER) [1]. They arise from the notochord along the axial skeleton and occur most frequently in the os sacrum and the skull base region [1]. According to the histopathology, chordomas are considered low-grade malignant neoplasms; therefore, slow but locally aggressive growth patterns explain late clinical appearance [2]. Independent of primary tumor location, standard therapy remains radical en bloc resection [3]. Total resection is often not feasible due to infiltration of surrounding vital structures [1]. Local recurrence is frequently seen after incomplete resection or after violation of tumor capsule [1, 4]. The 5-year local relapse rate for completely resected sacral chordoma remains 30% [5]. Local recurrence determines patient's survival [1]. In 5% of patients suffering from chordomas, distant metastasis in the lungs, bone, skin, and the central nervous system are reported at initial diagnosis [1].

Here, we report about a rare case of local recurrence of a sacral chordoma after multiple resections. The patient was

referred to our urological department for surgical treatment of a clinically inapparent recurrence located on the basis of the corpora cavernosa. We performed a complete resection of the recurrent chordoma by total penectomy and perineal urethrostomy.

## 2. Case Presentation

A 70-year-old patient was referred to our Department of Urology, Faculty of Medicine, Munich Germany, for a surgical treatment of a recurrent chordoma located at the radix penis. The primary chordoma of the os sacrum was diagnosed in 1996 and primarily resected. In the following eight years, the patient underwent three resections (R2) of local recurrences before starting medical therapy with imatinib 400 mg per day in 2009. In 2014, a dorsal complete compartment resection was performed for a fifth local recurrence (R0). Imatinib was paused until in the same year, a single pulmonary metastasis was diagnosed and treated by surgery (R0). Consequently, the medical therapy with imatinib was restarted. In the following year, a local recurrence of the proximal femur was diagnosed. Furthermore, the complete resection (R0) of the gluteal metastases was performed in

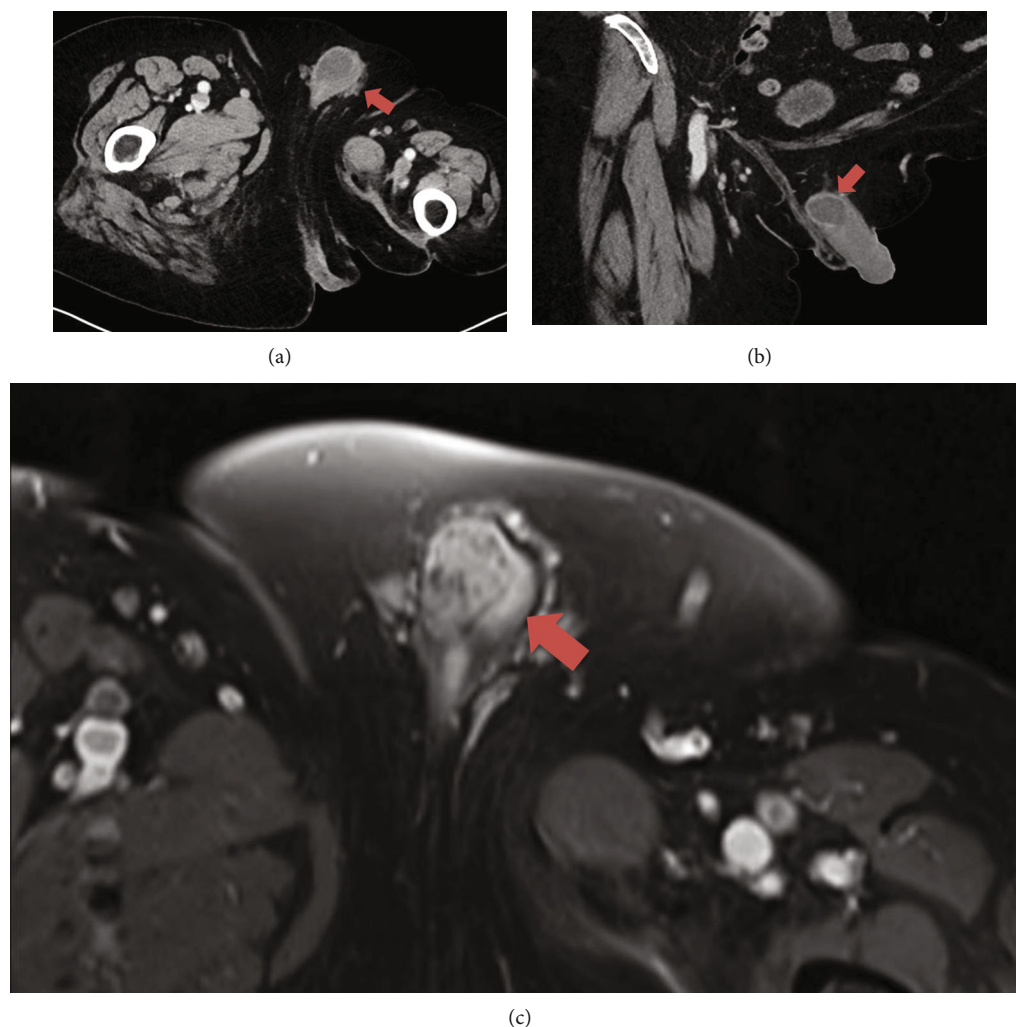


FIGURE 1: Preoperative MR imaging: (a) axial, (b) coronal, and (c) axial in T1 perfusion. Red arrows mark chordoma metastasis at the right corpus cavernosum of the penis as solid mass of about  $2.3 \times 3.0$  cm.

2017. In February 2019, another pulmonary metastasis was treated by atypical pulmonary resection (R0).

The follow-up computer tomography (CT) and magnet resonance imaging (MRI) indicated a suspicious lesion of the radix penis (Figure 1). Therefore, a biopsy was taken to prove the seventh local recurrence. The histopathology showed a chondroid chordoma. Clinically, the recurrence was unapparent. The interdisciplinary tumor board advised a surgical treatment.

**2.1. Surgical Treatment.** For preoperative preparation, a standard blood sample was taken, an abdominal CT scan in order to plan surgical procedure was performed and, antibiotic perioperative prophylaxis was given. We performed a total penectomy and perineal urethrostomy (Figures 2(a) and 2(b)). The intraoperative surgical margin was negative. Postoperatively, a transurethral catheter was used for 5 days to decrease risk of infection and to improve wound healing. A total inpatient stay of 7 days was noted. We reevaluated wound healing and functional outcome 14 days postoperatively.

**2.2. Histopathology Findings.** The macroscopic preparation showed an asymmetric indurated lesion with a maximal expansion of  $4.1 \times 2.5 \times 2.6$  cm. The resection margin to the corpus cavernosum was measured 0.3 cm and to the corpus spongiosum 1.9 cm. The tumor showed a suppressive, but not infiltrative growth of the corpus cavernosum.

The microscopic evaluation revealed a microcystic and partly solid tumor embedded in a myxoid and chondroid matrix (Figures 2(c) and 2(d)). In the center of the tumor, hemorrhagic and necrotic parts were identified. The tumor cells were voluminous and rich of vacuoles with rough chromatin.

### 3. Discussion

Local recurrence of chordoma is common even after aggressive surgical excision of primary tumor depending on surgical margins [4, 6]. Incomplete resection in initial therapy is a prognostic predictor for poor overall survival [7]. The literature describes 49% of disease-free survival after 15 years for negative surgical margins in comparison to 7% after R1 status

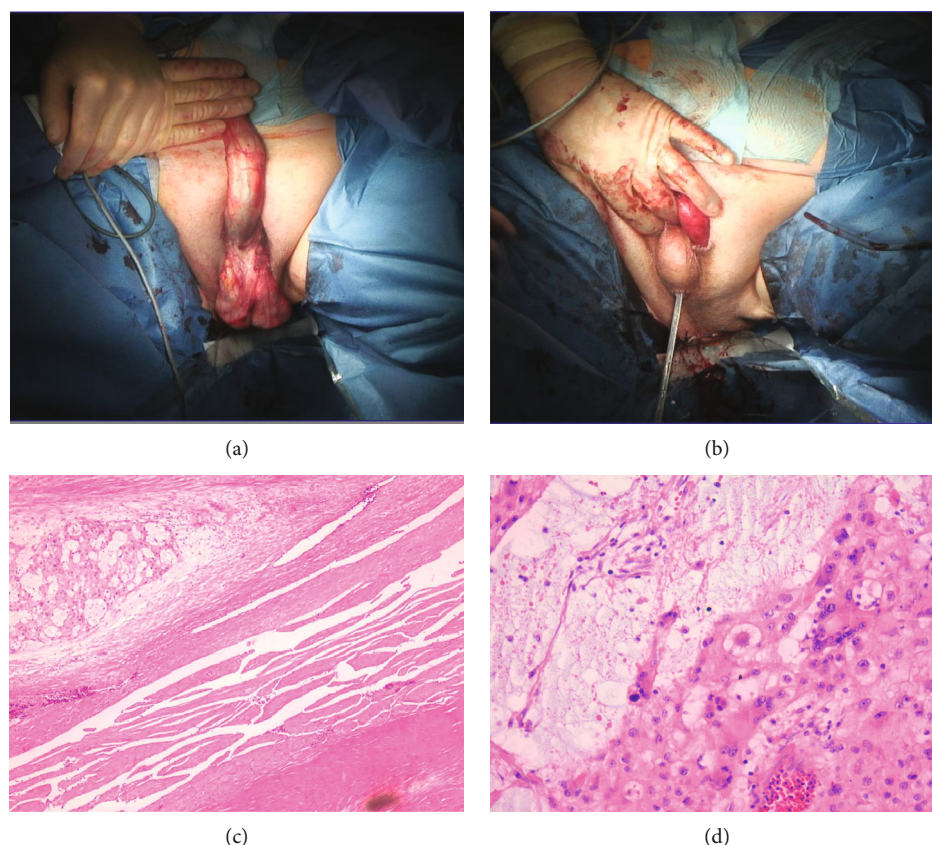


FIGURE 2: (a) First, the penis was denuded and the corpora cavernosa was separated before complete penectomy was performed. (b) In the last step, the urethra was diverted through the perineum. In the histology findings, the hematoxylin and eosin section shows corpora cavernosa infiltrated by typical physaliferous vacuolated large chordoma cells embedded in a myxoid stroma in images (c) and (d) (magnification 20x).

in sacral chordomas [5]. Postoperative follow-up to detect recurrence is important before reaching a size to become clinically apparent [8]. The treatment of recurrence remains the surgical resection and/or radiotherapy [4]. The literature describes that local recurrence of the os sacrum and mobile spine is highly related to tumor-related death [9]. Treatment options have to be chosen carefully by an interdisciplinary team to preserve best quality of life [4].

Most penile tumors are squamous cell carcinoma [10]. Metastases of the penis are uncommon, most frequently are primary malignancies from the urogenital tract [11]. Depending on tumor stage and location of the tumor, there are various treatment options [10]. The gold standard in advanced tumor stage is total penectomy and perineal urethrostomy [12]. Also, perineal urethrostomy is the last treatment option for benign urethral disease such as urethral strictures [13]. Despite invasiveness of total penectomy particularly affecting sexuality [14], Barbagli et al. reports a satisfaction after perineal urethrostomy in cases of urethral stricture of 78% [15].

There is one case that reported of a primary vaginal chordoma treated successfully by surgery; other than that, no chordomas of the genital region have been described so far [16].

We describe the first case of a recurrent chordoma located at the corpus cavernosum treated with total penect-

omy and perineal urethrostomy. For best oncological outcome, wide resection margins were achieved and perineal urethrostomy was performed to preserve continence.

#### 4. Conclusions

The treatment of chordoma and its recurrence disease demands an aggressive surgical excision for best long-term survival in consideration of conserving vital structures and preserving best quality of life.

#### Conflicts of Interest

All authors have stated that they have no conflict of interest.

#### References

- [1] B. P. Walcott, B. V. Nahed, A. Mohyeldin, J.-V. Coumans, K. T. Kahle, and M. J. Ferreira, "Chordoma: current concepts, management, and future directions," *The Lancet Oncology*, vol. 13, no. 2, pp. e69–e76, 2012.
- [2] V. Colia and S. Stacchiotti, "Medical treatment of advanced chordomas," *European Journal of Cancer*, vol. 83, pp. 220–228, 2017.
- [3] S. Stacchiotti and J. Sommer, "Building a global consensus approach to chordoma: a position paper from the medical

- and patient community," *The Lancet Oncology*, vol. 16, no. 2, pp. e71–e83, 2015.
- [4] S. Stacchiotti, A. Gronchi, P. Fossati et al., "Best practices for the management of local-regional recurrent chordoma: a position paper by the Chordoma Global Consensus Group," *Annals of Oncology*, vol. 28, no. 6, pp. 1230–1242, 2017.
- [5] S. Radaelli, S. Stacchiotti, P. Ruggieri et al., "Sacral chordoma: long-term outcome of a large series of patients surgically treated at two reference centers," *Spine*, vol. 41, no. 12, pp. 1049–1057, 2016.
- [6] P. Ruggieri, A. Angelini, G. Ussia, M. Montalti, and M. Mercuri, "Surgical margins and local control in resection of sacral chordomas," *Clinical Orthopaedics and Related Research*<sup>®</sup>, vol. 468, no. 11, pp. 2939–2947, 2010.
- [7] M. X. Zou, J. Li, X. B. Wang, and G. H. Lv, "Prognostic significance of resection degree in skull base chordoma: a systematic review and meta-analysis," *World Neurosurgery*, vol. 100, pp. 692–694, 2017.
- [8] S. A. Hanna, W. J. S. Aston, T. W. R. Briggs, S. R. Cannon, and A. Saifuddin, "Sacral chordoma: can local recurrence after sacrectomy be predicted?," *Clinical Orthopaedics and Related Research*, vol. 466, no. 9, pp. 2217–2223, 2008.
- [9] P. Bergh, L.-G. Kindblom, B. Gunterberg, F. Remotti, W. Ryd, and J. M. Meis-Kindblom, "Prognostic factors in chordoma of the sacrum and mobile spine: a study of 39 patients," *Cancer*, vol. 88, no. 9, pp. 2122–2134, 2000.
- [10] E. Marchionne, C. Perez, A. Hui, and A. Khachemoune, "Penile squamous cell carcinoma: a review of the literature and case report treated with Mohs micrographic surgery," *Anais Brasileiros de Dermatologia*, vol. 92, no. 1, pp. 95–99, 2017.
- [11] K. Zhang, J. Da, H.-j. Yao et al., "Metastatic tumors of the penis: a report of 8 cases and review of the literature," *Medicine*, vol. 94, no. 1, p. e132, 2015.
- [12] A. Simonato, A. Lissiani, S. Galli, A. Bozzola, A. Gregori, and F. Gaboardi, "Total penectomy with perineal urethrostomy for carcinoma of the penis," *European Urology Supplements*, vol. 1, no. 1, p. 193, 2002.
- [13] N. Lumen, M. Beysens, C. Van Praet et al., "Perineal urethrostomy: surgical and functional evaluation of two techniques," *BioMed Research International*, vol. 2015, Article ID 365715, 6 pages, 2015.
- [14] R. Sosnowski, M. Kulpa, M. Kosowicz et al., "Quality of life in penile carcinoma patients - post-total penectomy," *Central European Journal of Urology*, vol. 69, no. 2, pp. 204–211, 2016.
- [15] G. Barbagli, M. De Angelis, G. Romano, and M. Lazzeri, "Clinical outcome and quality of life assessment in patients treated with perineal urethrostomy for anterior urethral stricture disease," *The Journal of Urology*, vol. 182, no. 2, pp. 548–557, 2009.
- [16] S. L. Wei He, X. Gan, and L. Hu, "Chordoma of vagina a case report and review of the literature," *International Journal of Clinical and Experimental Medicine*, vol. 11, no. 5, pp. 5164–5170, 2018.