

Robot-assisted Radical Prostatectomy Due to a Primary Carcinoid Prostatic Tumor in a Three-Year-Old Child: A Case Report



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We present an extremely rare case of a 3-year-old child with a primary carcinoid tumor of the prostate. A 3-year-old boy presented with failure to thrive, constipation, recurrent respiratory tract infections, and pain in the genital area. His karyotype was normal and cystic fibrosis and coeliac disease were excluded prior to further investigation. An abdominopelvic computed tomography scan revealed a prostatic mass. Transrectal ultrasound-guided prostate biopsy was therefore performed and pathological examination revealed a carcinoid tumor. A robotic radical prostatectomy was performed. As this is an innovative surgical approach, we describe the surgical technique used. *UROLOGY* 133: 216–218, 2019. © 2019 Elsevier Inc.

Neuroendocrine tumors are a heterogeneous group of neoplasms derived from neuroendocrine cells, also known as enterochromaffin cells or amine precursor uptake and decarboxylation cells. They are quite rare, both in adults and in children. They have been described in various locations: 85% are found in the gastrointestinal tract, particularly the appendix and the ileum, but cases have been reported in lung, liver, kidney, ovary and, very rarely, the genitourinary tract. We present an extremely rare case of a primary carcinoid tumor of the prostate in a 3-year-old child.

CASE REPORT

The patient was a 3-year-old male, born after a 38-week twin pregnancy. At age 2 years, the patient started to show weight stagnation, constipation, and repeated respiratory infections. A thorough study of the boy was undertaken at the Paediatric Gastroenterology Department, and food allergies, cystic fibrosis, and coeliac disease were ruled out. The karyotype was normal and the bone age showed a 12-month delay.

Eight months later, the patient presented with poor general condition and significant progressive pain in the genital area. There was no evidence of abdominal masses, visceromegaly or peripheral adenopathies. The laboratory

tests disclosed significant protein-calorie malnutrition and also hypercalciuria.

Abdominopelvic computed tomography (CT) scan revealed a prostatic mass in the retrovesical area, with heterogeneous contrast uptake. The mass measured $2.7 \times 3.5 \times 3.2$ cm and caused an imprint on the base of the bladder (Fig. 1).

Abdominopelvic magnetic resonance imaging demonstrated a 3-cm-diameter prostate tumor with extension towards the extraprostatic fat, and in contact with the obturator muscle. A head CT scan showed a normal brain. Technetium-99m scintigraphy ruled out bone metastasis.

A transrectal biopsy of the tumor was performed, and the pathology suggested the presence of a carcinoid tumor with low proliferative activity.

Robotic radical prostatectomy was performed using a 5-port approach, with the patient in the supine position. Three robotic 8-mm ports were placed: 2 of them on either side of the body, obliquely and 10 cm from the navel, and the third left sided, parallel to the navel. The optical port (12-mm) was located in a supra-umbilical position, and a 5-mm port was placed on the right side for the assistant, parallel to the navel.

A transperitoneal approach was selected. The prostatic fat was dissected and the endopelvic fascia was incised bilaterally to allow separation of the prostate from the urogenital diaphragm. The urethra was exposed and the dorsal venous complex was ligated. The bladder neck was dissected until the prostatic urethra had been exposed; using the fourth arm of the robot, we were able to fix it and hold the urethral catheter, thus exposing the prostate. The posterior aspect of the bladder neck was then divided, to reduce damage and facilitate the dissection of the

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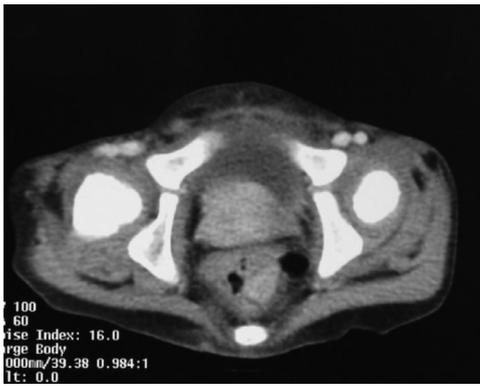


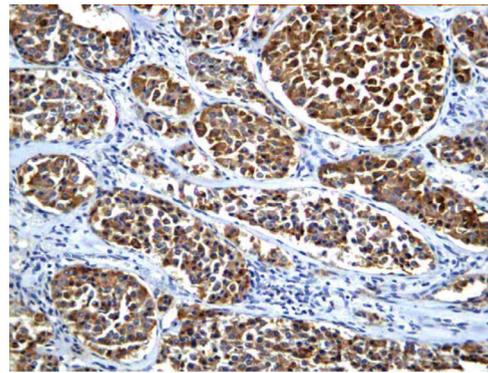
Figure 1. CT scan showed a prostatic mass in the retrovesical area, with heterogeneous contrast uptake. The mass caused an imprint on the base of the bladder.

vesicoprostatic space. Haemostasis of the vesicoprostatic lateral pedicles was ensured by means of Hem-o-lok clips. The seminal vesicles were then dissected with difficulty as they were particularly hard to identify, and the vasa deferentia were divided after opening the Denonvilliers' fascia. Dissection was performed between the rectal and the prostatic fascia, and the urethra was divided after transection of the dorsal venous complex. The prostate was placed in a sterile and airtight bag to prevent tumor seeding. Vesico-urethral anastomosis was performed with a 6/0 PDS double-needle continuous suture, and it was verified that the suture was watertight (Fig. 2). The surgery lasted 4 hours, with 100 mL of blood loss. We placed drainage at the Retzius space, which was removed 3 days after surgery. Also, an 8 Fr silastic urethral catheter was placed, which was removed 3 weeks after surgery and normal cystography.

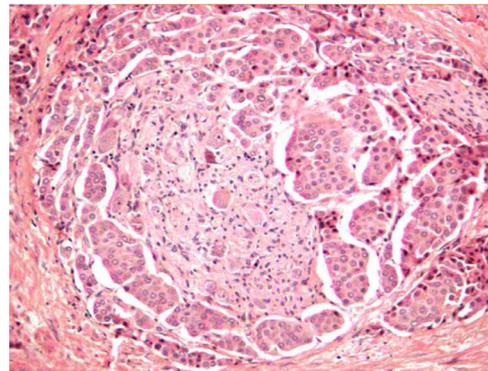
A macroscopically off-white, irregular tumor measuring $2 \times 2 \times 1.5$ cm and weighing 5 g was extracted. Microscopic examination revealed 90% infiltration of the organ by a carcinoid tumor positive for chromogranin A and negative for PSA, with capillary, perineural, and intraneural invasion. There was also extension to the periprostatic fat tissue without compromising of the seminal vesicles,



Figure 2. Vesico-urethral anastomosis. (Color version available online.)



(A)



(B)

Figure 3. (A) Chromogranin A staining showing neuroendocrine neoplasia. (B) Perineural and intraneural invasion by the carcinoid tumour. (Color version available online.)

and a positive surgical margin of 1×1 cm (Fig. 3A, B). The tumor was classified as pT3aN0M0R1.

The child's hospitalization lasted 6 weeks after surgery, due to 2 febrile episodes, cachexia, and requirement of nasogastric tube for nutrition. The use of urethral catheter relieved genital pain.

The patient was therefore discharged to be followed up by the Paediatric Department and to start adjuvant radiotherapy due to positive margins. Chemotherapy wasn't indicated. No postoperative complications occurred. The patient died 1 year after the surgery due to local recurrence and bone metastasis.

DISCUSSION

Carcinoid tumors in children are very rare. Some case series have reported a prevalence between 0.08% and 0.19%, but none of these series have described tumors located in the prostate.¹ The extreme youth of our patient at diagnosis is thus a striking feature, and the case can be considered extraordinary. Most of the limited number of childhood prostate tumor cases reported in the literature have been rhabdomyosarcomas, and Arena et al described 1 such tumor in a 2-year-old child.² In this context it should be noted that rhabdomyosarcomas account for 10%–15% of malignant solid tumors in children.²

Recently, a robot-assisted prostatectomy was performed in a child with an embryonal prostatic rhabdomyosarcoma.³ There is only 1 published report of a primary carcinoid tumor of the prostate, and this occurred in a 7-year-old child within the context of multiple endocrine neoplasia syndrome type IIB.⁴

The prognosis of prostate tumors in childhood is uncertain owing to the shortage of case reports; in our case, however, there were several poor prognostic factors, including capillary, perineural and intraneural infiltration, and positive surgical margins.

In adulthood, the prognosis depends on whether the disease is confined to the prostate and on the type of neuroendocrine differentiation. Six types of differentiation exist in the prostate: usual prostate adenocarcinoma with neuroendocrine differentiation, adenocarcinoma with Paneth cell-like neuroendocrine differentiation, carcinoid tumor, small cell carcinoma, mixed neuroendocrine carcinoma–acinar adenocarcinoma and large cell neuroendocrine carcinoma.⁵ True carcinoid tumors of the prostate are extremely rare and predominantly occur in the elderly. Their prognosis is uncertain because of the scarcity of cases, but they are said to follow a comparatively indolent course.⁶ Focal neuroendocrine differentiation in prostate cancer is seen in some 5%–10% of prostate carcinomas and represents a negative prognostic parameter.^{6,7}

A recent study⁸ showed better diagnostic performance of fluorodeoxyglucose-positron emission tomography/computed tomography compared to technetium-99m scintigraphy for detection of bone metastases in staging of prostate cancer. The approach adopted for the treatment of prostate tumors in children is multidisciplinary, combining surgery, chemotherapy and/or radiotherapy, and sometimes brachytherapy. In contrast, treatment of neuroendocrine tumors in adults rests largely on radical prostatectomy, even though there are adjuvant treatments such as chemotherapy and radiotherapy.⁹

As to surgical treatment of prostate tumors in children, the literature cites radical prostatectomy by means of infraumbilical laparotomy, as well as one case of radical

prostatectomy via the transpubic approach¹⁰ and one robotic-assisted radical prostatectomy.³ Use of the da Vinci robot improves surgical manoeuvrability, increases the depth of field thanks to the 3D image, and magnifies the structures; these advantages allow better dissection and greater precision in the movements and sutures.

CONCLUSION

Further studies are required to define the best treatment for prostatic neuroendocrine tumors in children. Robot-assisted radical prostatectomy is an optimal surgical option for paediatric prostatic tumors.

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