sequence (NCBI Reference Sequence: NG_009272.1). The oligonucleotide sequences of the forward and reverse primers are: exon 8 forward, 5’cctttaatgagatcctcccttc3´, exon 8 reverse, 5’ggggaaatgtggggtgtttcc3´, exon 9 forward, 5’ccetactgtcaccattgt3´ and exon 9 reverse, 5’gcactattcctctcaactgag3´. The PCR programme consisted of an initial denaturation step at 95°C for 15 min followed by 40 cycles, for exon 8 and 35 cycles, for exon 9, of the following steps: 95°C-1 min, 59°C-1 min and 60°C-1 min for exons 8 and 9, respectively, 72°C-1 min and a final elongation step 72°C-8 min. The amplicons were sequenced directly using the 7-Deaza-dGTP Cytm5/Cy5.5 Dye Primer Cycle Sequencing Kit (Bayer HealthCare, Leverkusen, Germany) sequencing kit in an OpenGene Visgen automated sequencer (Visible genetics, Ontario, Canada) and analyzed using the Unix Based Gene Objects 3.1 software. Following data analysis, a point mutation in intron 9 was identified (IVS9+4C>T).

**Clinical Progression**

Antihypertensive therapy was initiated and haemodialysis was started immediately via an arteriovenous fistula. Remarkably, concomitant with the completion of laboratory and genetic investigations the patient experienced a painful swelling of the left testis. At that time, a noticeable enlargement of the left testis, leading to a palpable mass of approximately 25 ml volume with hard consistency, was observed. Ultrasound scan showed an enlargement of the left testis with diffuse echogenicity and microcalcifications as well as three hypoechogenic lesions of 21mm, 10mm and 14mm diameter, respectively. Additionally, hydrocele was detected, whereas no ectopic right testis was found. Magnetic resonance imaging further confirmed the enlargement of the left testis with heterogeneity of the signal and disordered architecture, features implying a neoplasm. Additionally, in the right groin an oval shaped lesion measuring 17mm in diameter with hypointensity in T1 sequence and intermediate intensity in T2 sequence was revealed. The presence of a right hypoplastic testis could not be excluded. Computed tomography confirmed that there were no pathological lymph nodes. The right kidney had a maximum diameter of 8 cm and the left kidney a maximum diameter of 7.1 cm. Serum tumor markers are summarized in Table 2. The patient was submitted to bilateral testicular resection. Prosthetic testes were implanted at the time of gonadectomy. Sperm preservation could not even be considered in the constellation of highly elevated FSH levels.

Histological analysis performed in the left testis revealed a germ cell tumor with morphological and immunohistochemical features consistent with a Sertoli cell tumor. The cells were arranged in tubules with solid areas, a characteristic that made the differentiation from seminoma challenging. Furthermore, the cells demonstrated clear or slightly eosinophilic cytoplasm, nuclei with atypia, mitosis and a great range of appearance as well as areas of necrosis, calcifications and elements of inflammation. Immunohistochemistry supported the discrimination between Sertoli cell tumor and seminoma. Indeed, the staining disclosed a strong positivity for vimentin in the cytoplasm as well as around the nuclei, positivity for cytokeratin and negativity for placental alkaline phosphatase and c-kit-1, which are all features characteristic of Sertoli cell tumor. Markers for neuroendocrine tumors were negative. Nuclei were positive for Ki 67 at a percentage higher than 40%. The size of the neoplasm (6.5 cm), the increased number of mitotic figures as well as the atypia were all indicative of a malignant progress. The tumor was surrounded by an atrophic testis with hyalinized structures and hyperplastic Leydig cells. In the hypoplastic right testis, a gonadoblastoma measuring 0.5 cm in diameter was identified. Subsequently, the patient received adjuvant chemotherapy with Bleomycin, Etoposide and Cisplatin (Platinol) (BEP), with the prospect of continuing with androgen replacement therapy after the completion of chemotherapy.

**DISCUSSION**

Gonad and kidney development depends on the appropriate expression of the *WT1* gene, located on chromosome 11p13. Mutations at the intron 9 donor splice site of *WT1* lead to a modification of the relative