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CASE REPORT

Prostatic sarcoma of the Ewing family in a 33-year-old male — A case report and review of the literature

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Abstract Ewing sarcoma is the second most common primary bone tumor seen in children and adolescents, typically presenting between 10 and 20 years of age. Extraosseous sarcomas of the Ewing family in adults are rare. We report a manifestation of this tumor entity in the periprostatic tissue of a 33-year-old male and discuss our treatment approach. Transrectal biopsy is a feasible and simple diagnostic tool for unclear pelvic masses. Multi-modal therapy and central registries are needed to gain knowledge of rare pelvic tumors like Ewing sarcoma.

1. Introduction

Ewing sarcoma is the second most common primary bone tumor seen in children and adolescents. However, extraosseous sarcomas of the Ewing family in adults are very rare. We report a manifestation of this uncommon tumor entity in the periprostatic tissue of a 33-year-old male.

2. Case presentation

A 33-year-old patient who had recently migrated from Afghanistan presented to the emergency department with symptoms of pelvic pain and urinary tract infection (dysuria, urgency, mild pyuria on dipstick) for several days. Patient's past medical history was unremarkable and without any recollection of exposure to toxic substances or genetic predisposition. Upon suspicion of a pelvic mass on transrectal ultrasound during initial assessment, magnetic resonance imaging (MRI) of the pelvis was performed. It revealed a 6.0 cm × 4.5 cm × 4.6 cm mostly solid pelvic mass showing signs of central necrosis located between bladder and rectum with suspected infiltration of the prostate and left internal obturator muscle (Fig. 1).

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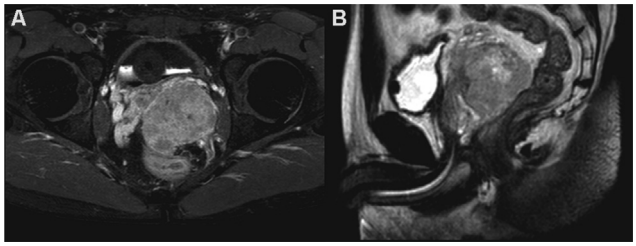

Figure 1 The 6.0 cm × 4.5 cm × 4.6 cm mostly solid pelvic mass on T1-weighted, contrast-enhanced (A) as well as T2-weighted MRI-study (B). It shows signs of central necrosis and displacement of bladder and rectum with suspected infiltration of the prostate and left internal obturator muscle; partly contrast-filled bladder with placed foley catheter.

In order to complete tumor-staging, additional cranial MRI, chest/abdominal computed tomography (CT), bone-scan as well as colonoscopy were performed. Besides unspecific enlargement of a cervical lymph node on the left hand side, no signs of metastatic disease were detected.

For further histological evaluation of the mass, ultrasound-guided transrectal biopsies were obtained. Upon pathological examination the tumor was preliminarily described as a "small, round and blue-cell-like tumor" of the periprostatic soft-tissue infiltrating the capsule of the prostate without further classification (Fig. 2).

Besides the Ewing's sarcoma family of tumors, the initial differential diagnoses included desmoplastic round cell tumor and neuroendocrine carcinoma. Other possible differential diagnoses, such as neuroblastoma, rhabdomyosarcoma as well as non-Hodgkin's lymphoma [1—3], were excluded by immunohistochemistry. With a symptomatic aggressive tumor and in order to get the complete histology, an initial surgical resection was chosen. Radical tumor resection with prostatovesiculectomy, extended regional lymph node dissection as well as resection of the pelvic floor were performed using an open retropubic approach. The bladder neck was then closed and a cystostomy-catheter was inserted for urine-diversion (Fig. 3).

Course of postoperative recovery was unremarkable. The intraoperative setting suggested at least an R1-situation, even though surgical margins were later reported as tumor-free (R0) by pathology. After further molecular profiling by a reference pathologist (Prof. Leuschner, Kiel, Germany)

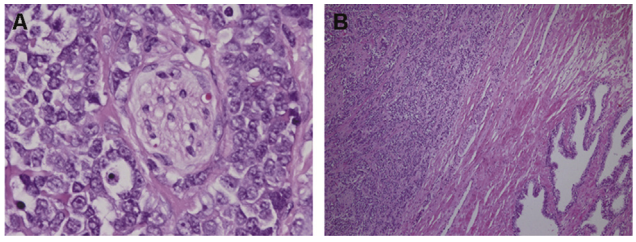

Figure 2 Upon pathological examination the transrectal biopsy was preliminarily described as a small-blue-round-cell tumor and later classified as a Ewing-like tumor of the periprostatic tissue. (A) Nerve fibre infiltration of the tumor (400×); (B) Tumor formation with close proximity to the prostatic tissue (40×).

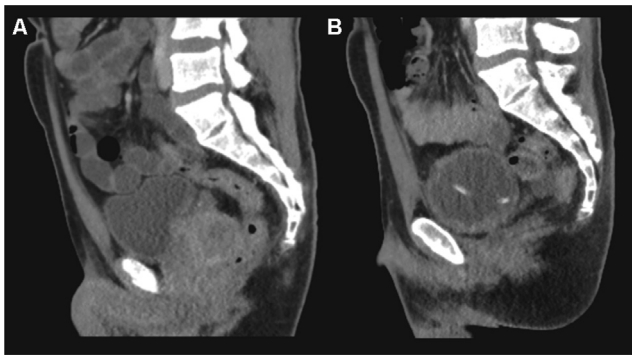

Figure 3 Contrast enhanced staging CT-scan of the pelvis (A) and re-staging 2 months after surgery and first cycle of adjuvant chemotherapy (B): no signs of residual disease or recurrence of the Ewing-like tumor. Urine-diversion via cystostomy-catheter after closure of the bladderneck.

the tumor was shown to hold genetic alterations of the *EWSR1*-gene consistent with the tumor of the Ewing sarcoma family, even though cytokeratin positivity as expressed by this tumor is a rather uncommon histopathological feature only found in a minority of cases in this entity [4,5]. Due to the aggressive nature of this disease and given the likely presence of residual disease an intensive regime of adjuvant chemotherapy was initiated shortly after surgery. It consisted of six courses of VIDE (vincristine, ifosfamide, doxorubicin, etoposide) applied 3-weekly. Most recent restaging 12 months after surgery and after completion of the VIDE-regime showed no signs of residual disease or recurrence. Currently further chemotherapy is being administered with an alternating VAI- (vincristine, actinomycin, ifosfamide) and VAC-regime (vincristine, actinomycin, cyclophosphamide). Continuous remission provided, a second surgery for definitive orthotopic or cutaneous continent urinary diversion is intended upon completion of adjuvant therapy.

3. Discussion

Ewing sarcoma is a rare malignant disease in which cancer cells are found in the bone or in soft tissue. Approximately 25% of patients with Ewing sarcoma have metastatic disease at the time of diagnosis. Ewing sarcoma occurs most frequently in teenagers and young adults, with a male/female ratio of 1.6:1 [6]. In patients aged 10—19 years, the incidence is between nine and 10 cases per million people, the incidence for all ages is even more rare with one case per million people in the United States. The polymorphism (*EGR2*-gene on 10q21.3) associated with the increased risk is found at a much higher frequency in whites than in blacks or Asians, possibly explaining the epidemiology of the relative infrequency of Ewing sarcoma in the latter populations [7].

For extraosseous primary tumors, the most common primary sites of disease include the following: trunk (32%), extremity (26%), head and neck (18%), retroperitoneum (16%), other sites (9%). The Surveillance, Epidemiology, and End Results (SEER) database was used to compare patients younger than 40 years with Ewing sarcoma who presented