Botryoid rhabdomyosarcoma of the bladder in an adult female

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Summary

We report on a case of rhabdomyosarcoma of the bladder in an adult patient. A 65-year-old woman presented with macroscopic hematuria. Ultrasonography revealed a large bladder tumor and severe hydronephrosis of the right kidney. Computed tomography (CT) confirmed these findings and revealed extravesical tumor extension. Cystoscopic biopsy of the tumor showed botryoid rhabdomyosarcoma. The patient’s local and general conditions did not allow any adjuvant treatment. The patient died of disease 6 months later.

Key words: bladder, rhabdomyosarcoma

Introduction

Rhabdomyosarcoma is the most common soft tissue sarcoma in children and adolescents, accounting for 5% of all childhood tumors. Twenty percent of all rhabdomyosarcomas involve the genitourinary tract [1]. However, malignant urinary bladder tumors of mesenchymal origin in adults are extremely uncommon. Their incidence has been reported to range from 0.23% to 0.67% of all bladder tumors [2]. Usually, bladder rhabdomyosarcoma arises from the region of the trigone and posterior vesical wall. Apart from its ability to invade local tissues rapidly, lymphatic and hematogenous dissemination are possible [3]. Clinical findings are hematuria and more frequently bladder irritative symptoms. Survival is usually poor due to the tumor’s aggressive clinical course. We report on a case of rhabdomyosarcoma of the bladder in an elderly patient.

Case presentation

A 65-year-old female patient presented with a 2-month history of asymptomatic macroscopic hematuria. Her medical history consisted of blindness, diabetes mellitus and severe cardiopulmonary disease. Clinical examination showed a firm mass in the bladder region. Urinalysis was positive for hematuria. The hemoglobin and creatinine levels were 10 g/dl and 1.8 mg/dl, respectively. Ultrasonography of the kidneys and the bladder showed severe hydronephrosis of the right kidney and a large bladder mass with a diameter of 8 cm occupying the right wall of the bladder. A CT scan confirmed these findings revealing also posterior and right-sided extravesical tumor extension and a contact with the sigmoid colon and the vaginal cuff (Figure 1). Cystoscopy revealed a large polypoid bladder tumor arising from the right and posterior bladder wall that almost filled the bladder cavity. A cystoscopic biopsy was subsequently performed.

Histopathologic examination of the specimens showed spindle-shaped, undifferentiated tumor cells with hyperchromatic nuclei and abundant eosinophilic cytoplasm lying in between a loose-textured myxoid stroma. Multiple areas of necrosis and haemorrhages were also seen (Figure 2). Immunohistochemically, cells were reactive for vimentin, neuron-specific enolase (NSE) and desmin (Figure 3). The histological and immunohistochemical findings were consistent with a rhabdomyosarcoma of the botryoid subtype.
Figure 1. CT scan shows a 8×7.7 cm solid mass in the bladder and tumor infiltration of right vesical wall.

The patient’s local and general condition did not allow a definitive cancer treatment (radical surgery, radiation therapy and/or chemotherapy). The patient died of the disease 6 months later.

Figure 2. Rhabdomyosarcoma consisting of undifferentiated (a) and spindle-shaped (b) cells lying in between a loose myxoid stroma. (H&E ×40).

Figure 3. Immunohistochemical staining revealing a strong positive reactivity for vimentin (a), NSE (b), and desmin (c). (H&E ×40).

Discussion

Rhabdomyosarcoma is the most common soft tissue sarcoma in children. Approximately 20% of these patients have genitourinary rhabdomyosarcomas [1]. However, the incidence of this tumor in adults is extremely low [2].

Rhabdomyosarcoma arises from primitive totipotential embryonal mesenchymal cells. Bladder rhabdomyosarcoma is classically an endophytic tumor that grows usually in the submucosa of the trigone and posterior vesical wall and expands intraluminally in a botryoid configuration [4]. The clinical presentation of bladder rhabdomyosarcoma is variable. The predominant symptoms are hematuria, frequency, dysuria and urgency. These symptoms become aggravated as the tumor grows and obstructive symptoms may ensue, potentially leading to infection or urinary retention. Involvement of the trigone (the most common location in the bladder) can result in hydronephrosis and deterioration of the renal function due to obstruction [5,6]. Rhabdomyosarcoma is a highly malignant neoplasm with a propensity for early local in-
filtration and eventual metastatic dissemination, and its prognosis is poor [7]. In most cases, diagnosis can readily be made by histological morphology. However, immunohistochemistry is helpful in distinguishing rhabdomyosarcoma from others malignant mesenchymal tumors of the bladder [2].

The use of combined-modality therapy comprising chemotherapy, radiation therapy and surgery has improved the long-term survival rates of young patients from approximately 25% in 1970 to more than 70% in recent years. During the last 2 decades, the Intergroup Rhabdomyosarcoma Study Group (IRSG) has identified prognostic factors for these patients that help improve subsequent therapeutic manipulations [1,8]. However, a uniform multimodality treatment strategy has not been established for adult genitourinary rhabdomyosarcoma despite the success of this approach in the treatment of pediatric genitourinary rhabdomyosarcoma. Therapy for rhabdomyosarcoma has been empiric and, due to the limited experience with elderly patients, comparisons among series are difficult to make. Adult patients need to be carefully selected for radical surgery, radiation therapy and chemotherapy because the morbidity associated with these approaches is not negligible. It must also be remembered that aging brings an increase in associated illnesses and a deterioration in immune function [5,9]. In our case, because of extensive local invasion of the tumor and the general condition of the patient, radical surgical therapy or palliative treatment was not performed.

References