Case Report

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Embryonal Rhabdomyosarcoma of the Prostate in Adults: A Case Report

Mouhamadou Bachir Ba*, El Hadji Aliou Baldé, Franck Aurélien Chouamou Cheumaga, Fatimatou Néné Sarr, Maimouna Mané, Papa Macoumba Gaye

Radiotherapy Department, Dalal Jamm Hospital, Dakar-Senegal

SUMMARY

Rhabdomyosarcomas (RMS) are malignant tumors that develop from normal skeletal muscle tissue. It is a rare soft tissue cancer in adults. We report the case of a 26-year-old patient with an embryonal RMS from the prostate extended to the bladder with pelvic lymph node involvement. He is received for erectile dysfunction, lower back pain on his right side, pollakiuria, urgent urination and decreased urine output. The clinical examination revealed a prostate tumor. A CT scan shows a pelvic mass developing at the expense of the early-enhanced prostate, with irregular contours, invading the posterior wall of the bladder. Pelvic MRI gave similar results. Standard histology with immunostaining of the pieces of the transrectal biopsy concluded in an RMS of the embryonal type. No distant metastasis is detected on the extension workup. The tumor was classified as T2bN1M0 stage 3, group III. Neo-adjuvant multidrug therapy is administered in six courses and a complete response results. The patient then refuses surgery. Radiotherapy using a three-dimensional conformational technique (RC3D) is delivered to the entire pelvis with overprinting on the initial prostate lesion up to a total dose of 70Gy. The patient is in remission 16 months after his diagnosis. Embryonal prostate RMS is a rare disease in adults. The treatment is multidisciplinary by neo-adjuvant chemotherapy then surgery and / or radiotherapy.

*Corresponding author

Mouhamadou Bachir Ba, Radiotherapy Department, Dalal Jamm Hospital, Dakar-Senegal; Tel: +221 781780679; E-mail: mbbachir21@gmail.com

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Introduction

Rhabdomyosarcomas (RMS) are malignant tumors that develop from normal skeletal muscle tissue. This is a rare type of soft tissue tumor. It is predominant in children under 10 years old and rarely seen in adults. Three histological entities are described. The two subtypes that affect both children and adults are embryonal and alveolar rhabdomyosarcomas, with pleomorphic RMS occurring almost exclusively in adults. Although these tumors can appear almost anywhere, the most frequent sites of the development of these tumors are at the level of the structures of the head and neck (approximately 40% of all cases), at the level of the male or female genitourinary system (approximately 25% of all cases) and limbs (approximately 20% of cases) [1-4]. We report the case of a 26-year-old patient with an RMS of the embryonal type of the prostate extended to the bladder with pelvic lymph node involvement treated by chemotherapy and radiotherapy.

Patient and Observation

A 26-year-old adult presents with erectile dysfunction, lower back pain on the right side, pollakiuria, urgent urination and decreased urine output. The administration of oral antibiotics, antispasmodics and alpha-blockers does not generate any improvement. A CT scan shows a 10.2 x 8.2 x 6.4 cm pelvic mass developing at the expense of the early-enhanced prostate, with irregular contours, invading the posterior wall of the bladder, in contact with the anterior wall of the rectum, causing bilateral ureterohydronephrosis, with

bilateral external iliac lymphadenopathy and left obturator. The urethro-cystoscopy finds a prostate tumor of irregular appearance, invading the bladder and bleeding on contact with non-visualized ureteral meatus. Pelvic MRI gives similar results with bilateral capsular protrusion (Figure 1).

Figure 1: Axial MRI slice showing a prostate tumor with irregular contours

A transrectal needle biopsy reveals tissue comprising a neoplastic proliferation of round cells, medium in size, monomorphic with cytoplasm, site of a rhabdoid and eccentric nucleus eosinophilic inclusion, focus positive for desmin, vimentin, actin, and myogenin during immunostaining. CT scan and bone scintigraphy detected

no distant metastasis. The diagnosis concludes with an embryonal T2bN1M0 RMS of the prostate stage 3, group III. Multi-agent chemotherapy including doxorubicin, bleomycin, vinblastine and dacarbazine (ABVD), is administered in six courses and a complete response results. Erectile function returns to normal. The patient then rejects the surgery. Complementary radiotherapy using a three-dimensional conformational technique (RC3D) is delivered to the entire pelvis with overprinting on the initial prostate lesion at a dose of 46Gy and 70Gy respectively (Figure 2). Radiotherapy treatment is complicated by the occurrence of grade II diarrhea relieved by anti-diarrhea drugs. The patient returns to activities within three months of completing their 8-month treatment and is still in complete remission 16 months after diagnosis.

Figure 2: RC3D treatment plan with Dose-Volume histogram

Discussion

Rhabdomyosarcoma is a very rare cancer. Only 350 new cases of RMS are diagnosed each year in the United States in young people under the age of 21, and about 4 per million healthy children under 15 develop RMS each year. This cancer affects boys slightly more than girls and is more common in young children under the age of 7 [4]. RMS are rare in adults. Five studies with large numbers of patients have been published comprising 400 adult cases of RMS in some of the largest expert centers, in the United States and Europe, in the last 20 to 30 years. Although pleomorphic histology is more common among the adult population, the embryonal type is not as exceptional [5]. The genitourinary localization is found in 25% of cases of RMS. Sarcomas of the prostate represent less than 0.1% of all malignant prostatic tumors in adults, including RMS which are the most frequent, ahead of leiomyosarcomas, phyllodes tumors of the prostate and other rare sarcoma entities [6]. The diagnosis is based on standard histology, immunostaining with search for t(2; 13) PAX3-FKHR translocation by RT-PCR. The CT scan and Magnetic Resonance Imaging (MRI) are the morphological examinations most used in the assessment of the extension of RMS. The CT scan being advantageous for the evaluation of bone lesions and abdominal lymphadenopathies and MRI provides a better definition of the primary tumor and its relations with neighboring organs [7]. Embryonal RMS has a better prognosis than other histological types (alveolar and pleomorphic). Other prognostic factors are determined by age (less than 10 years), tumor site, tumor size (less than 5cm) and TNM stage.

In vesico-prostatic RMS, specific treatment can be preceded by a urinary diversion if necessary. In our case, there was no deterioration in renal function despite bilateral ureterohydronephrosis and the patient had refused any surgery.

Currently all patients with RMS are treated with chemotherapy. Depending on the size and location of the original tumor and the extent to which it can be resected with surgery, most patients also undergo multimodal treatment with radiation therapy and surgery. The chemotherapy drugs used are commonly used in sarcomas of other tissues soft (dactinomycin, ifosfamide, vincristine) in the form of VAC (vincristine, dactinomycin, cyclophosphamide) or VAI (vincristine, dactinomycin, ifosfamide), or even IVADO, with the addition of doxorubicin(8)Under our working conditions, the patient had received adriamycin, Bleomycin, Vinscritine and dacarbazine with a good clinical and radiological response. The implementation of this neo-adjuvant chemotherapy allowed a marked improvement in the prognosis and allows the realization, if possible, of a conservative treatment [6].

Surgery is the local treatment of choice for RMS except in certain locations inaccessible to it (RMS para meningeal, orbit, etc.) or in the case of an organ conservation protocol. Oncologic surgical treatment includes resection of the original site with a circumferential margin of at least 1cm [4]. Our patient had refused surgery after neoadjuvant chemotherapy. Radiation therapy is indicated in all patients with alveolar RMS, even those whose tumors were completely removed before the start

of chemotherapy, and almost all with embryonal RMS with incomplete resection (microscopic or gross) or those without surgery. Our patient received external radiotherapy using a three-dimensional conformational technique at a total dose of 70Gy. Brachytherapy because of these ballistic advantages is often used in RMS of vesico-prostatic origin. This radiotherapy technique allows more precision on the tumor volume and better protection of neighboring healthy tissues [8,9].

In this prostate location, embryonal RMS has a better prognosis in children than adults, even if the results of multimodal treatment are very encouraging compared to alveolar or pleomorphic ones.(2.4).

Conclusion

Prostatic localization of embryonal RMS is rare in adults. We report the case of a 26-year-old patient suffering from an embryonal RMS of the prostate with bladder extension and pelvic lymph node treated exclusively by chemotherapy and external radiotherapy. In order to improve their prognosis, these patients should be referred if possible to a specialized multidisciplinary team, both medically and surgically.

Competing Interests

The authors declare no conflict of interest.

Contributions from the Authors

All authors have read and accepted the final manuscript

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