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A rare case of metastatic prostatic rhabdomyosarcoma in a young adult: Clinical challenges and therapeutic perspectives

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ABSTRACT

Embryonal rhabdomyosarcoma of the prostate in adults is rare and often diagnosed at an advanced stage, with metastases. We report the case of a 23-year-old young adult presenting with low back pain and dysuria, whose imaging revealed a voluminous metastatic prostate mass. Biopsy confirmed embryonal rhabdomyosarcoma. Treatment was initiated with chemotherapy, resulting in significant regression of the tumour mass and metastases after 3 courses. Pediatric advances suggest improved survival with a multimodal approach, but its efficacy in adults requires further investigation.

1. Introduction

Embryonal rhabdomyosarcoma of the prostate in adults is a very rare occurrence, with only a handful of published cases. Diagnosis usually occurs at an advanced stage of the disease, often already metastatic, and is associated with an aggressive clinical course.[1] Several therapeutic approaches are used in its management, including radical surgery, radiotherapy and chemotherapy. We present here a case of metastatic embryonal rhabdomyosarcoma in a young adult.

2. Case presentation

A 23-year-old young adult with no significant medical history presented with bilateral low back pain and dysuria. Clinical examination revealed a very enlarged prostate, hard and irregular on rectal examination. The PSA level was measured at 0.38 ng/ml, within the normal range. Ultrasound revealed bilateral ureterohydronephrosis, with serum creatinine of 40 mg/l, indicating renal failure. Pelvic Magnetic resonance imaging (MRI) revealed a large, multilobed prostate mass measuring 87 × 80*116mm, with T2 isointense and diffusion hyperintense features, strongly enhanced after contrast injection (Fig. 1). The mass had contacts with the iliopubic branches, rectal wall, bladder neck, iliopubic muscle and psoas muscles, with multiple retroperitoneal adenopathies and secondary bone lesions in the right iliac wing, third sacral vertebra and T12 and L1.

Transrectal prostate biopsy under local anesthesia confirmed embryonal rhabdomyosarcoma, with poorly differentiated tumour proliferation. Pathological examination revealed small malignant cells, arranged in patches and sheets, with marked anisokaryosis. The stroma was mainly composed of lymphocytes and plasma cells. An immunohistochemical complement was performed, showing positive anti MYOGENIN and anti DESMINE antibodies (Fig. 2).

The subsequent thoracic CT scan revealed a bilateral pleural effusion with left supra-clavicular adenopathy and tumor involving bilateral axillary lymph nodes. In addition to the prostatic focus, the PET scan showed a pelvic mass in the psoas muscle, suggesting secondary muscle involvement, hypermetabolic adenopathy above and below the diaphragm, suggesting lymph node involvement, and multiple hypermetabolic foci in the bone, suggesting secondary sites.

After bilateral nephrostomies had been inserted and renal function had returned to normal, the decision taken at the multidisciplinary consultation meeting was to institute combined treatment including radiotherapy and chemotherapy.

The chemotherapy protocol, comprising Vincristine, Ifosfamide, Mesna and Doxorubicin, was initiated. A significant reduction in the size of the prostate mass was observed after three cycles of chemotherapy (47 × 22mm vs 87 × 80), accompanied by the disappearance of adenopathy and pleural effusion on the follow-up thoraco-abdomino-plevic scan (Fig. 3). Despite the introduction of a combined course of radiochemotherapy, with clear regression of the prostate mass after three

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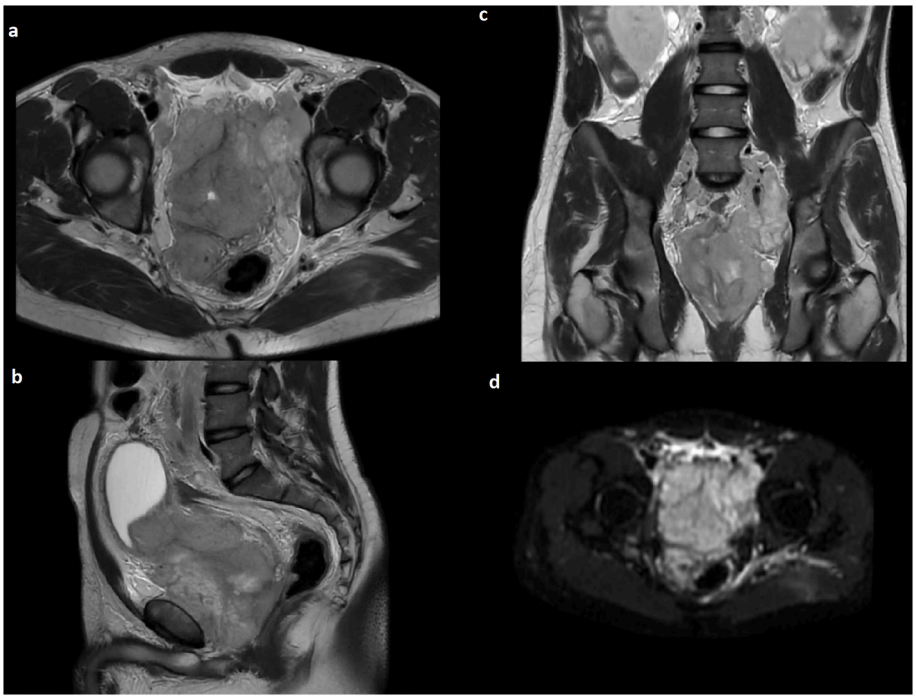

Fig. 1. Pelvic Magnetic resonance imaging (MRI) (a-transversal T2. b-sagittal T2. c-Frontal T2. d-transversal diffusion) showing a large, multilobulated neoplastic mass within the prostate, exhibiting isointensity on T2-weighted imaging and hyperintensity on diffusion-weighted imaging, markedly enhanced following contrast administration, with delineation of some central necrotic areas measuring 87 × 80*116mm. This mass extends anteriorly in contact with the iliopubic branches, posteriorly abutting the anterior rectal wall with loss of separation line in places, superiorly and anteriorly infiltrating the bladder neck, laterally contacting the iliopubic muscle and psoas muscles with infiltration of the right psoas muscle. Additionally, multiple retroperitoneal, para-aortic, primitive iliac, internal iliac, and external iliac lymphadenopathies are note.

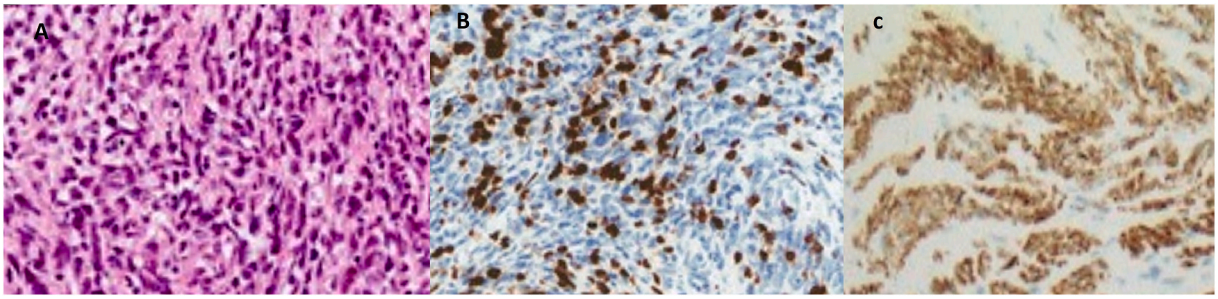

Fig. 2. A) Histology imaging showing tumor cell type of embryonal rhabdomyosarcoma showing malignant cells organised in patches and sheets. The tumour cells are small, with round or oval nuclei with marked anisokaryosis (modificationx400). B)tumor cells with strong nuclear positive for myogenin (myogenin immunostaining x n *x*100). C) cytoplasmatic reactivity for Desmin ((Desmin immunostaining x 100).

cycles of chemotherapy, the patient died before he could benefit from a fourth course of treatment, following respiratory failure caused by a severe lung infection, as a result of a weakened immune system most likely due to chemotherapy.

3. Discussion

Sarcomas of the genitourinary system in adults remain rare, accounting for approximately 2.1 % of all soft tissue sarcomas, with a higher prevalence in the paratesticular and renal regions. Sarcomas of the prostate are exceptionally rare in adults, accounting for around 0.1 %–0.2 % of all prostate cancers.[1] Adult prostatic rhabdomyosarcoma (RMS) has an aggressive clinical course and a poor prognosis, often characterised by large lesions, extensive involvement of the urogenital tract and distant metastases.[2]

Clinical manifestations of prostatic RMS usually include voiding problems such as urinary retention, dysuria and, rarely, haematuria. Typical clinical findings include prostate enlargement and a normal serum PSA value.[1] The disease generally progresses rapidly, so the diagnosis is often made at an advanced stage when there are distant metastases. Macroscopic examinations often show a nodular tumour with greyish-white indurated cut surfaces.[3] Treatment options, ranging from surgery to radiotherapy and chemotherapy, depend on the stage of the disease. However, for metastatic stages, the optimal role of local therapies remains undefined.[4]

Survival data for prostatic rhabdomyosarcoma in adults is poor, partly due to late diagnosis and the difficulty of achieving complete resection. Studies show that the majority of patients have large tumours with metastases at the time of diagnosis.[5] Compared with the paediatric population, where therapeutic advances have considerably improved survival rates, the treatment of rhabdomyosarcoma in adults remains a challenge, due to the lack of data on the efficacy of chemotherapy and radiotherapy.[6] Initiatives such as the Rare Cancer Network multicentre study aim to fill this gap by collecting data to better characterise

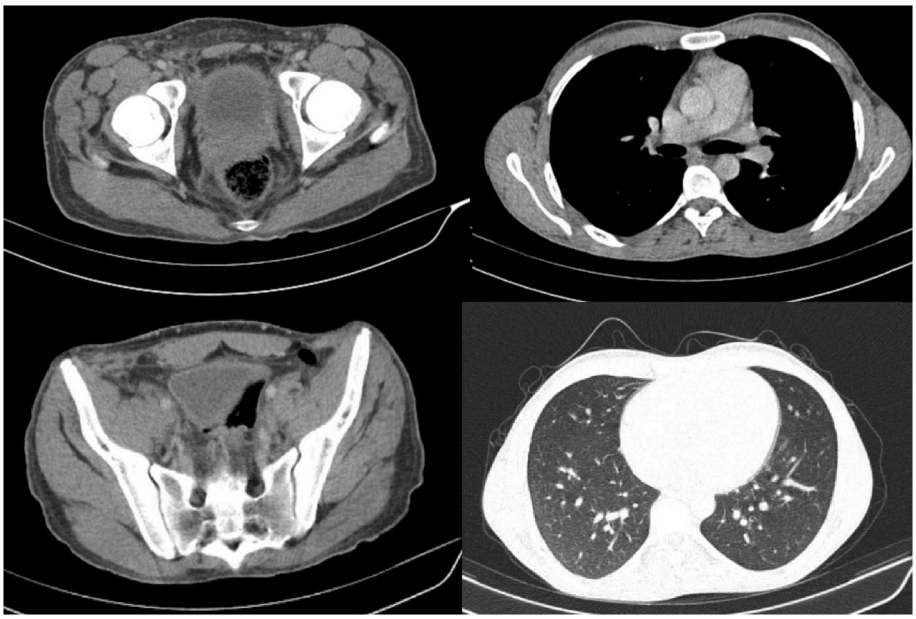

Fig. 3. The follow-up thoraco-abdomino-plevic CT scan images, showing a significant reduction in the size of the prostate mass, decrease in the number and size of lymph nodes and a total regression of the bilateral pleural effusion.

treatment strategies in adults with prostatic sarcoma.[7] The retrospective study included 6 US and 10 European centres with a total of 61 patients diagnosed from 1987 to 2016, 4 % of whom were classified as rhabdomyosarcoma. Local control and overall survival were only marginally improved in patients with advanced disease treated with a multimodal approach.[8]

In the case we describe here, primary radical surgery was not an option as our patient already had very advanced and metastatic disease at the time of diagnosis, highlights the importance of multidisciplinary management and the urgent need to develop more effective therapeutic strategies to improve the prognosis of patients with metastatic prostatic rhabdomyosarcoma in adults.

4. Conclusion

Rhabdomyosarcoma of the prostate in adults is rare and often diagnosed late. Our case highlights the importance of early recognition and multidisciplinary management. Chemotherapy appears to play a role in controlling metastatic disease. However, the role of surgery and radiotherapy in advanced cases requires further evaluation.

CRediT authorship contribution statement

Reda Tariqi: Writing – original draft, Project administration, Data curation, Conceptualization. **Mehdi Farina:** Methodology, Investigation, Data curation. **Abdelmounim Boughaleb:** Resources, Methodology, Investigation. **Hamza El Abidi:** Resources, Funding acquisition. **Imad Boualaoui:** Resources, Methodology. **Ahmed Ibrahimi:** Visualization, Project administration. **Hachem El Sayegh:** Writing – review & editing, Supervision. **Yassine Nouini:** Writing – review & editing, Supervision.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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