Prostatic rhabdomyosarcoma in the adult, A case report

Sajad Ahmad Malik, Arif Hamid Bhat, Sajad Sultan Lone

Department of Urology, Sher e Kashmir Institute of Medical Sciences, Soura, Srinagar, Kashmir, India

ABSTRACT

Prostate embryonal rhabdomyosarcoma (ERMS) is a common tumour in infants and children, but it is rare in adults. It is characterized by a high degree of malignancy, both local rapid growth with formation of large pelvic masses, often leading to renal failure due to bladder outlet obstruction, and systemic spread, commonly to the lungs, liver and bone.

We report on a case of a stage III prostate ERMS, approached with combined-modality treatment, with the administration of 5 courses of doxorubicin, ifosfamide and 2-mercaptoethane sulfonate sodium (mesna), with planned subsequent radiotherapy to the prostatic bed (60 Gy/30 fractions).

1. Introduction

Rhabdomyosarcoma (RMS) is the most frequent soft tissue tumour in paediatric patients, accounting for up to 50% of all soft tissue sarcomas. In adults RMS is quite rare. Clinical experience is limited to case reports. Three histological subtypes have been identified: embryonal with botryoid and alveolar histiotypes, generally affecting young children and adolescents. The pleomorphic histiotype is the one encountered in adult patients. In patients still in their childhood, it is most frequently located in the cervical-cephalic region and genitourinary system. In adults, RMS can involve the thorax and extremities accounting for 2-5% of all soft tissue sarcomas. Possible therapeutic management options are radical surgery, radiotherapy and chemotherapy. This combined approach has significantly improved the outcome, in terms of survival and quality of life in children, while in adults the prognosis overall has been poor. We present a case of 28-year-old male affected by stage III ERMS of the prostate treated with a combined chemo-radiotherapy approach followed by radical surgery, resulting in a disease-free survival of 2 years and a good quality of life, with a complete remission of the urinary symptoms.

2. Case presentation

A 28-year-old male presented to us with complaints of severe suprapubic pain accompanied by inability to pass urine for one day. He admitted having difficulty in passing urine for the last one year for which he had not sought any medical advice and had self medicated. On examination he had a palpable urinary bladder and was promptly catheterised without any difficulty. A rectal examination revealed a large mass lesion filling the entire pelvis with smooth overlying rectal mucosa. Serum PSA was 1.6 ng/dl. An ultrasound examination of the pelvis revealed a large heterogenous mass apparently arising from and replacing the prostate gland (Fig. 1A).

CT scan of the abdomen and pelvis (Fig. 1B) revealed a large (8x7x7 cm) heterogeneously enhancing mass lesion in the pelvis posterior to the bladder having ill-defined plane with posterior wall of urinary bladder and rectum. Mass was seen abutting the pelvic side walls. These findings were corroborated on MRI pelvis (Fig. 1C,1D,1E,1F) which showed an enlarged prostate gland (160ml) replaced by a T2 bright signal causing loss of zonal differentiation and lobulated outline with extra capsular breach involving both seminal vesicles with multiple enlarged bilateral internal iliac nodes. Bone marrow examination revealed normal marrow cells. FDG PET scan (Fig. 2A,2B,2C,2D) showed intense activity in the enlarged prostate, seminal vesicles, and right internal iliac nodes. Tc 99 m bone scan did not reveal any bony metastasis.

Biopsy sections showed features of small round blue cell tumour with focal areas of pleomorphism, spindling and high mitotic activity suggestive of embryonal rhabdomyosarcoma (Fig. 2E). Immunohistochemistry revealed positivity for desmin and myogenin consistent with embryonal rhabdomyosarcoma (Fig. 2F).

The patient underwent chemotherapy comprising of doxorubicin, ifosfamide and 2-mercaptoethane sulfonate sodium (mesna) for 5 cycles in our institution. Check imaging with MRI revealed a partial response to chemotherapy according to the RECIST Criteria. Post neoadjuvant chemotherapy patient underwent a successful catheter free trial. Patient underwent radical cystoprostatectomy with ileal conduit after proper

* Corresponding author.
E-mail addresses: drmalekajad33@gmail.com (S.A. Malik), drarifhamid@yahoo.co.in (A.H. Bhat), sslone012@gmail.com (S.S. Lone).

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Fig. 1. A USG Picture. B CT Axial cut. C MRI Axial cut; D, E MRI Sagittal cuts; F MRI Coronal cut.
counselling of the risks and benefits involved in view of residual tumour. In hospital post-op period was uneventful. After discharge patient developed enterocutaneous fistula with fecal discharge from wound site. It was a low output fistula with rectal content and settled with conservative management only. Histopathology showed near complete excision of the mass lesion with embryonal histology (Fig. 2F). Involvement of the seminal vesicles bilaterally and the cut urethral margins was demonstrated as was the presence of lymphatic and vascular invasion. Owing to the development of the enterocutaneous fistula (Fig. 3), adjuvant radiotherapy to the prostatic bed and the pelvis was withheld until recovery. Patient subsequently received adjuvant radiotherapy (60 Gy/30 fx). Patient currently doing well and is on regular follow-up.

3. Discussion

Primary ERMS of the prostate gland is an extremely rare pathological and clinical entity in adults, with fewer than twelve cases reported in literature. The presenting symptoms are often related to urethral obstruction, with frequency, hesitancy, and dysuria as predominant symptoms or, less often, with hematuria and acute urinary retention. The compression of the rectum can cause constipation, rectal bleeding and a sense of rectal fullness. CT scan and MRI study reveal a large soft tissue mass with areas of necrosis replacing the whole prostate, but the radiological differential diagnosis with prostate adenocarcinoma can be very difficult. MRI can clearly show the site of origin as the central prostate area, with compression of a recognizable peripheral portion. The diagnosis is usually performed with a transurethral, transrectal or perineal biopsy. Histopathological evaluation can reveal variable grade of differentiation along the myogenesis pathway. Immunohistochemistry for skeletal muscle specific markers, such as against myoglobin and skeletal muscle actin and myosin can help identify the tumour as rhabdomyosarcoma. A localized prostate disease can be treated with radical surgery.

Fig. 2. A Coronal PET CT Cut. B, C Axial PET CT Cuts. D Saggital PET CT Cut. E Hematoxylin and Eosine showing primitive mesenchymal cells having variable degrees of skeletal muscle differentiation. Both hypo- and hypercellular areas with a loose myxoid stroma are present. F HIC slide showing positivity for desmin and myogenin.
4. Conclusion

No definitive conclusions can be drawn up to now regarding the best available treatment option towards prostate ERMS in adults. However, in an organ-confined disease, radical surgical extirpation should be performed. As alternative to surgery, radiation therapy combined with chemotherapy might be useful to achieve local control with organ functional preservation and good quality of life. The most effective chemotherapeutic drugs have yet to be established. Further studies are needed in the future in order to improve the understanding of biological behaviour of prostate ERMS in adults and to define the most appropriate therapeutic approach, as well as the timing of local therapy in metastatic disease.

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