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Radical Cystectomy with Ileal Neobladder in a patient with Hemophilia! A case report

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ABSTRACT

A 38 year old hepatitis C and Hemophilia Type A patient presented with recurrent hematuria since 6 months. Investigations revealed a 5 cm lesion in the lateral wall of the bladder with perivesical fat stranding. PET CT showed a FDG avid lesion measuring 4 cms in the bladder with non FDG avid Iliac nodes. Bladder preserving protocols were not considered in this patient in view of the need for further adjuvant treatment. Patient underwent a Radical cystectomy with bilateral pelvic lymph node Dissection and Ileal Neo-bladder after optimisation with Factor VIII transfusion and correction of bleeding parameters.

Introduction

Hemophilia is an X linked recessive genetic disorder of the coagulation pathway. Hemophilia A (Factor VIII deficiency) affects 1 in 5000–10000 and Hemophilia B (Factor IX deficiency) affects 1 in 40,000. Complications are related to bleeding especially internal bleeding in the joints and intestines. The increased life expectancy of hemophiliac patients, with the availability of recombinant Factor replacements, has also lead to recognition of previously undetected malignancies. Although recurrent hematuria is seen but bladder malignancy as a cause is rare. Various guidelines for carcinoma bladder suggest adjuvant treatments (intravesical BCG, chemotherapy or radiation) in all bladder preserving protocols that can lead to severe hematuria in hemophiliacs. Radical cystectomy with bilateral pelvic lymph node dissection provides the only alternative for a cure. Ileal conduit is a good option especially in elderly patients. In young patients with associated social and sexual needs, neobladder recreation using the intestine facilitates continence and voiding per urethra. Such a complex procedure in a Hemophiliac provides a unique challenge to the surgeon.

Case description

38 year old gentleman presented with complaints of recurrent hematuria since 6 months. Conservative treatment failed and hence he was further investigated.

Ultrasound revealed multiple growths in the bladder largest measuring 5 cms.

A contrast CT abdomen and pelvis confirmed the findings along with perivesical fat stranding. FDG PET CT showed a metabolically active lesion in the bladder with a few non FDG avid Iliac Nodes (Fig. 1).

Coagulation parameters revealed a normal Pro-Thrombin time and an elevated Activated Partial Thromboplastin time. Further Haematological work up revealed a severe Factor VIII deficiency. (Fig. 2).

All options were discussed with the patient and his relatives.

Hemato-oncologists were involved to optimise the patient for surgery. 100% correction with Factor VIII (4500 IU) was done on the day of surgery. Factor VIII increment assay done half an hour after Factor VIII infusion showed good increment of 104%.

Patient then underwent Radical Cystectomy with Bilateral Pelvic Lymph Nodal Dissection with Ileal Neo-Bladder reconstruction on 11th December 2019. Intra-operatively Factor VIII assay done revealed decrease in Factor VIII to 76% 4 hours after the first injection. Patient was given 15 units/kg of Factor VIII (1350 IU). Intra operative blood loss was around 1.5 L. Patient was transfused 4 PRBCs and Factor VIII transfusions.

Day 0: 4500 IU stat, 1350 IU intra operative, 2000 IU at night.
Day 1: 2500 IU in the morning and 2000 IU at night.
Day 2: 2000 IU BD.
Day 3–7: 2000 IU BD.

Patient was discharged from the hospital on the eight post operative day.

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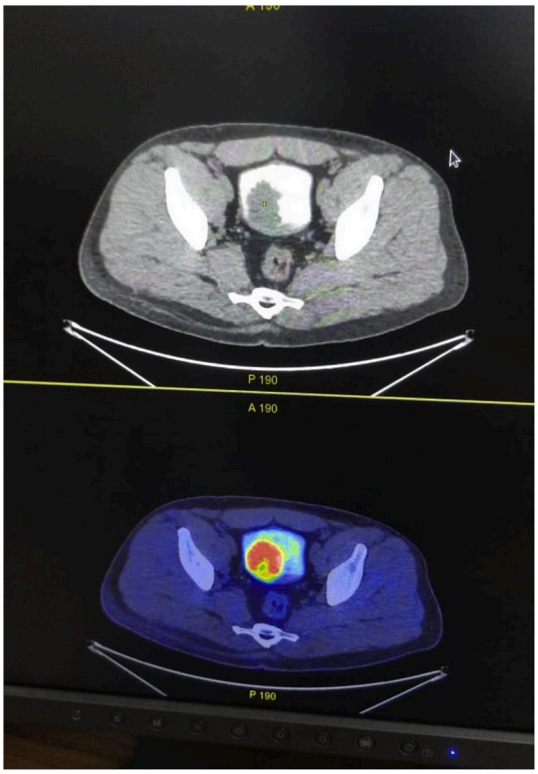

Fig. 1. PET CT revealing the FDG avid growth in the bladder.

Histopathology

Low grade urothelial carcinoma with no evidence of muscle invasion (Fig. 3).

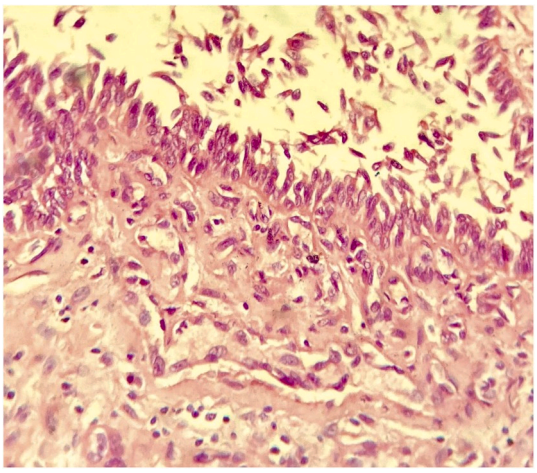

Fig. 3. Microscopic picture of the histopathological slide.

Discussion

Hemophilia is a rare X linked recessive genetic disease affecting the production of Factors VIII and IX ranging from mild (>5%), moderate (1–5%) and severe disease (<1%).[1] Hemophiliacs can present with hematuria frequently. Hematuria is mostly due to benign causes but those not responding to conservative treatment require further investigations to rule out a malignancy.[2,3,4] Various guidelines (AUA/EAU) suggest Radical Cystectomy as one of the treatment modalities for intermediate and high risk bladder tumors.

Radical cystectomy particularly in this patient was mandatory in view of the Hemophilia wherein adjuvant therapies would have resulted in severe hematuria.

Radical Cystectomy in a hemophiliac involves a high risk of bleeding complications. A team effort requiring Hemato-oncologists, Anaesthetists and Surgeons with adequate Factor VIII replacement can ensure

Test Name	Result	Unit	Reference Range
VON WILLEBRAND WORK UP			
PT- Prothrombin Time Test	14.0	Sec	11-14
PT Control	12.0	Sec	
APTT clot based	58.0	Sec	26-36
APTT CONTROL	30.0	Sec	
FIBRINOGEN LEVEL clauss	220.0	mg/dl	200-400
THROMBIN TIME clot based	21.0	Sec	18-22
FACTOR VIII ASSAY clot based	3.9	%	50-150
FACTOR IX ASSAY clot based	70.0	%	50-150
FACTOR VIII Chromo chromogenic	5.0	%	50-150
VWF ANTIGEN ELFA	115.0	%	60-150
VWF – RICOF ASSAY LTA	66.21	%	50-200

Fig. 2. Hematological investigation revealing severe Factor VIII deficiency.

good outcomes in such patients. A quick in and quick out approach was followed, hence the decision to conduct an open procedure.

Regular monitoring of the Factor VIII levels ensured a smooth post operative recovery in our patient. Patient is continent, potent and voiding per urethra using the creeds manoeuvre and clean intermittent catheterisation at 3 months follow up. No recurrent episodes of hematuria or Malena has been reported by the patient till date. This is the first reported case of Radical cystectomy with Ileal Neo bladder in a Hemophiliac patient. Although radical cystectomy with ileal conduit has been reported earlier for severe hematuria.[5]

Conclusion

Hemophilia with hematuria is a nightmare to any practising urologists. The need for frequent clot evacuations and failed conservative therapies mandates a cystectomy even for a benign cause. In carcinoma bladder Radical Cystectomy probably is the only option to achieve favourable cancer free survival rates and improving the quality of life. Neo bladder diversion using the ileum though a complex procedure provides a continent diversion especially in young patients. Team approach can ensure good outcomes even in such patients with Hemophilia who can have significant bleeding complications in the perioperative period if not managed properly.

Author contribution

Karthik Rao: Writing - original draft, Writing - Reviewing and Editing. **Mohan Keshavamurthy:** Conceptualisation, Validation, Resources. **Shakir Tabrez:** Supervision. **Sreeharsha Harinatha:** Supervision. **Niti Raizada:** Supervision, Resources.

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