CASE REPORT

Embryonal rhabdomyosarcoma of urinary bladder in an adult patient: an unusual manifestation

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SUMMARY

An adult man presented with off and on painless haematuria, which was diagnosed as bladder mass on contrast-enhanced CT. He underwent transurethral resection of bladder tumour with complete resection. A diagnosis of embryonal rhabdomyosarcoma was made following histopathology and immunohistochemistry report. The patient was planned for radical cystectomy but was found to have large recurrent infiltrating bladder mass with liver and bone metastasis. Hence, the plan for radical cystectomy was deferred and the patient was counselled regarding chemotherapy (vincristine, cisplatin and doxorubicin regime). The patient tolerated the first cycle of chemotherapy, but his condition deteriorated prior to the second cycle and the patient expired.

BACKGROUND

Rhabdomyosarcoma (RMS) is a malignant soft-tissue tumour that exhibits skeletal muscle differentiation. The most common variant is embryonal whereas alveolar variant is rare. It accounts for 6% of childhood malignancies.¹ Twenty per cent of cases occur in the urogenital tract (eg, vagina, urinary bladder, prostate, paratestis and uterus).² In adults, embryonal rhabdomyosarcoma of the urinary bladder is a rare presentation with anecdotal reports in scientific literature. Because of the rarity, clinicopathological features in this age group such as histological subtype, biological behaviour and treatment response (ie, chemosensitivity) are not well defined as compared with their paediatric counterpart. Here, we report a case of a 25-year-old man who presented with

haematuria and has been diagnosed as a case of embryonal rhabdomyosarcoma.

CASE PRESENTATION

A 25-year-old man presented with complaints of off and on painless haematuria for the past 3 months. There was no history of fever, weight loss, suprapubic pain, previous surgery or smoking. General physical and local examination was unremarkable.

INVESTIGATIONS

Routine laboratory investigations were within normal limits. Ultrasonography abdomen reported normal bilateral upper tracts along with a mass measuring 41×36mm on the anterior wall of the urinary bladder. Contrast-enhanced CT (CECT) revealed a heterogeneously enhancing soft-tissue attenuation lesion measuring approximately 52×15mm arising from the anterior wall of the urinary bladder with subcentimetric and perivesical lymph nodes (figure 1A,B). On cystourethroscopy, an irregular solid mass was seen arising from the anterior wall of the urinary bladder.

DIFFERENTIAL DIAGNOSIS

Transitional cell carcinoma of urinary bladder may be considered in differential diagnosis.

TREATMENT

Intraoperatively, hard growth (4×4 cm) was present on the right anterolateral wall of the urinary bladder with bilateral normal ureteric orifices. The patient was managed by transurethral resection

Figure 1 (A, B) Contrast-enhanced CT showing a heterogeneous lesion arising from the anterior wall of the urinary bladder.

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Figure 2 Histopathology and immunohistochemistry photographs showing rhabdomyoblasts (A) with positivity for myogenin (B), desmin (C) and vimentin (D).

of bladder tumour with complete resection. Postoperative course was uneventful.

OUTCOME AND FOLLOW-UP

Histopathology and immunohistochemistry findings confirmed embryonal rhabdomyosarcoma arising from the urinary bladder, positive for myogenin, vimentin and desmin (figure 2A–D). The patient was planned for radical cystectomy, but the patient developed a hard lump in the suprapubic area. Repeat CECT scan showed extramural enhancing supravescical mass (79×123 mm) with infiltration into the omentum, mesentery and sigmoid colon with multiple liver space occupying lesion (SOL) suggestive of metastatic deposits (figure 3A,B). The patient was given first cycle of intensive chemotherapy regimen (vincristine, cisplatin and doxorubicin) and was sent home. Unfortunately, the patient deteriorated prior to the second cycle of chemotherapy and passed away.

DISCUSSION

Embryonal rhabdomyosarcoma is generally regarded as a neoplasm occurring primarily during childhood; there are few case reports of this neoplasm in adults.^{3–9} Only one case is

reported from the Indian subcontinent.⁵ Embryonal rhabdomyosarcoma has three histopathological forms that can occur either singly or in combination: botryoid, spindled and anaplastic. The botryoid form arises beneath epithelial mucosal surfaces and has a cambium layer. The spindled variant most commonly occurs in scrotal soft tissues and the rest occurring in the head and neck region. Immunohistochemical methods with antimyoglobin antibody (myogenin, desmin) and antisarcomeric actin antibody, characteristic of striated muscle, are helpful in the diagnosis of rhabdomyosarcoma.

In the paediatric age group, the prognosis for urinary bladder embryonal rhabdomyosarcoma is less favourable compared with disease at other sites (vagina, uterus, paratesticular region; 70% to 73% and 84% to 89%, respectively).¹⁰ However, the prognosis and optimal management of embryonal rhabdomyosarcoma in the adult age group is uncertain owing to its rarity.

Due to lack of proper guidelines, various treatment options (surgery, chemotherapy, radiotherapy) have been tried for the management of embryonal rhabdomyosarcomas in adults in the past as per surgeon's choice. Hence, there is an urgent need for uniform and systematic guidelines.

Figure 3 (A, B) Contrast-enhanced CT showing extramural enhancing supravescical mass with multiple liver space occupying lesion (SOL) suggestive of metastatic deposits.

Learning points

- Embryonal rhabdomyosarcoma of urinary bladder is rare in the adult population.
- Immunohistochemistry (antimyoglobin antibody and antisarcomeric actin antibody) may aid in confirming the diagnosis.
- Embryonal rhabdomyosarcoma should be managed aggressively.
- Multimodal treatment strategies (surgery, radiotherapy, chemotherapy) are available, but proper guidelines are lacking.

As seen in this case, tumour metastasises to liver and bones in short course of follow-up even after complete resection done endoscopically. This depicts very aggressive behaviour of RMS. So, embryonal rhabdomyosarcoma of the genitourinary tract should be managed very aggressively.

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