Neuroblastoma involving urinary bladder: A rare type of tumor with diagnostic ambiguity

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Abbreviations & Acronyms CT = computed tomography NHL = non-Hodgkin's lymphoma NSE = neuron-specific enolase

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Received 6 January 2019; accepted 19 May 2019. Online publication 10 June 2019 **Introduction:** Neuroblastoma of urinary tract is an extremely rare type of tumor and only a few cases have been reported worldwide.

Case presentation: We presented an adult case of neuroblastoma involving urinary bladder. Patient was a 24-year-old man with a history of repeated attack of hematuria. Patient underwent cystoscopy and transuretheral resection of bladder tumor. Biopsy result was "Blastema cell (suggestive of neuroblastoma) with immature ganglion cells." Immunohistochemistry workup was continued and neuron-specific enolase and S100 staining were positive. Patient lost to further follow-up visit.

Conclusion: Although neuroblastoma of skin and peripheral nerve is not a rare disease entity, involvement of deeper structures (especially urinary bladder) is exceptional.

Key words: cystoscopy, hematuria, neuroblastoma, transuretheral resection of bladder tumor, urinary bladder.

Keynote message

Adult neuroblastoma involving urinary bladder is rarely reported in literature. This patient is also the first reported case of adult neuroblastoma of urinary bladder in Myanmar.

Introduction

Myanmar has a high incidence of bladder tumor mainly transitional cell carcinoma. But other types of solid tumors such as sarcoma and neuroblastoma are rarely encountered and there is not even a single report until now. This is the very first case of adult neuroblastoma of urinary bladder in Myanmar.

Case presentation

We report the case of previously healthy 24-year-old man who was referred to the Department of Urology, Mandalay General Hospital following a 3-week history of visible hematuria. It was described as total painless hematuria. He also complained about lower urinary tract symptoms such as frequency and straining. Moreover, there was also significant loss of weight and appetite.

Although abdominal examination was normal, digital per rectal examination revealed a lobulated extraluminal mass at the anterior wall. Upper limit of the mass could not be reached.

All the laboratory results were within normal limits except neutrophil leukocytosis. Initial pelvic ultrasound scan showed a large echogenic mass in the retrovesical area about 69×69 mm invading into the posterior vesical wall.

Contrast enhanced CT scan of abdomen and pelvis showed a well-defined hypo-dense mass 79×73 mm between the bladder and rectum with slight enhancement in post-contrast scan (Fig. 1). There was no enlarged lymph node. Radiologist's impression was pelvic tumor (lymphoma probably NHL) and differential diagnosis was teratoma.

Because of inconclusive imaging results, joint cancer team decided for cystoscopy and transuretheral resection of tumor for tissue diagnosis. Cystoscopy revealed a large solid tumor



Fig. 1 Axial CT scan of neuroblastoma of urinary bladder of the patient.



Fig. 2 (a) $10 \times$ magnification of neuroblastoma of urinary bladder of the patient. (b) $40 \times$ magnification of neuroblastoma of urinary bladder of the patient (stained with NSE).

occupying the whole bladder neck and trigonal area. Size was about 80×80 mm and the tumor occupied almost the whole bladder. Both ureteral openings could not be identified. Then transure theral resection of tumor was done by monopolar diathermy. It was only for diagnostic purpose.

Histological report of resected tissue was "blastema cells (suggestive of neuroblastoma) with immature ganglion cells and schwannian stroma, Ganglioneuroblastoma" (Fig. 2a). Pathologist also suggested for immunohistochemistry workup. Immunohistochemistry staining resulted in positive for both NSE and S100 and the remark was "Ganglioneuroblastoma" (Fig. 2b).

While waiting for further surgery, the patient was admitted to the oncology unit with features of raised intracranial pressure. CT scan of the head showed a brain metastasis in the right temporal lobe. Emergency radiotherapy to metastasis was given for one. Since then, we have lost contact with the patient.

Discussion

Neuroblastoma is an extra-cranial solid tumor arising from neural crest cells. So they can arise anywhere along the sympathetic nervous system.¹ Previously, neuroblastoma of urinary bladder was mainly reported in pediatric patients. Adult neuroblastoma of urinary bladder was rarely reported.

In 2008, the International Neuroblastoma Risk Group introduced pretreatment classification system and staging system. The classification system includes four simple stages – L1, L2, M and MS.² Depending on risk factors such as age, grade and genetics, patients can be categorized into four different prognostic groups – very low, low, moderate and high.^{3,4}

On literature review, the oldest reported case of neuroblastoma of urinary bladder was from Iran. The patient was a 52year-old man. Unlike our patient, that patient came to the hospital with the chief complaints of recent low back pain and night sweats. The patient underwent surgical excision (bladder preserving surgery) and received six cycles of vincristine, dacarbazine and cyclophosphamide. Until the time of case report (2011), the patient was still free of tumor recurrence.⁵

Adults with neuroblastoma have a significantly worse outcome than children. This may be due to tumor biology, a more virulent clinical course, or the fact that adults are less sensitive or have poor tolerance to pediatric chemotherapy regimens. The observed 3- and 5-year survival rates were the lowest among adults (45.9% and 36.3%, respectively), but 86% and 84.6% of infants survived for 3 and 5 years. A long-term evaluation of adult neuroblastoma survival in a 10-year follow-up study revealed a continuous decrease in survival during the first 7 years after diagnosis.⁵

Treatment for neuroblastoma of urinary bladder mainly depends on the International Neuroblastoma Risk Group pretreatment classification system and staging system. Treatment options include surgery (mostly bladder preserving surgery), chemotherapy, and radiotherapy.¹ In our case, because of the nature of the tumor (large, aggressive and occupying the bladder neck), primary surgical treatment could not be applied.

Conclusion

We report a case of a 24-year-old man presenting with painless total hematuria who was found to have neuroblastoma in a rare location (urinary bladder). According to this patient, it is imperative to arrange for early cystoscopy and high-resolution imaging for a young patient with unresolved hematuria.

Conflict of interest

The authors declare no conflict of interest.

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