

## Primary clear cell carcinoma of urinary bladder: A case report



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### ARTICLE INFO

#### Article history:

Received 1 September 2017

Received in revised form

22 September 2017

Accepted 5 October 2017

Available online 19 October 2017

#### Keywords:

Bladder

Clear cell

Cancer

Urinary bladder

### Introduction

The most common malignancy of urinary tract is bladder cancer with more than 330000 new cases each year and more than 130000 deaths per year. Incidence of this tumor is more in males than females. Majority of histological type of bladder cancer are urothelial carcinoma, 90%. Other types of bladder tumor such as squamous cell carcinoma and adenocarcinoma are rare.<sup>1,2</sup> In fact the genitourinary carcinomas has been categorized in four groups: Variant forms of urothelial carcinomas (UC), squamous cell carcinoma (SCC), adenocarcinoma and undifferentiated carcinoma.<sup>1</sup> According to the world health organization (WHO) definition bladder adenocarcinoma is a malignant neoplasm which originates from urothelium and has pure glandular phenotype in histological studies.<sup>2</sup> Clear cell type is one of the histological growth patterns of bladder carcinoma.<sup>2</sup> Mullerian clear cell type affects female genitalia and due to some clinical and pathologic characteristics, it has been thought maybe it arises from mullerian ducts.<sup>3,4</sup> PCCUB is rare

and many clinicians are unfamiliar with its clinical and diagnostic presentations.<sup>3</sup> Few cases of PCCUB has been reported in the age of 19–80 years which despite UC females were affected more than males.<sup>1</sup> The tumor presents as a large solitary papillary or sessile lesion which has polypoidal growth.<sup>2,3</sup> The clinical presentation of PCCUB are hematuria, lower urinary tract symptoms and discharge.<sup>3,4</sup> The patients may complain of flank or pelvic pain due to local invasion of tumor that resulted in ureterovesical junction involvement and subsequently hydronephrosis.<sup>4</sup> Immunohistochemistry assay (IHC) has high sensitivity and specificity for this tumor diagnosis.<sup>3</sup> Pan cytokeratin, CK7 and CA125 are positive for this tumor.<sup>1,3,4</sup> The best treatment and outcome for PCCUB yet is unclear, however surgery is considered as the choice treatment.<sup>3,4</sup> The differential diagnosis of PCCUB are nephrogenic metaplasia, UC with clear cell cytoplasm, diffuse large B-cell lymphoma (DLBCL) and metastatic clear cell carcinoma.<sup>3,4</sup>

### Case presentation

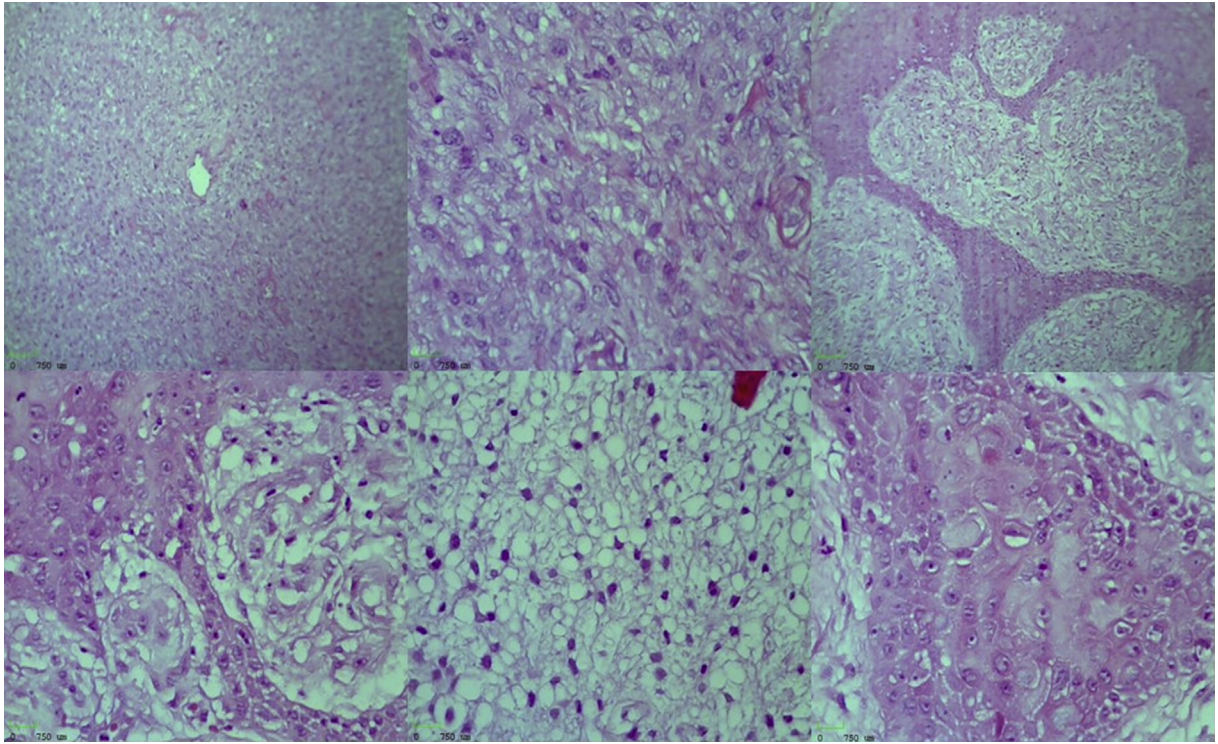
A 78 years old female complained of periodic gross hematuria and dysuria. She had no history of past medical disorders, tobacco or cigarette smoking. Her hemoglobin was 12 mg/dl. The urinalysis showed hematuria. The other lab results were normal. GU tract sonography revealed a 34 × 42 mm solid mass with lobulated margin and discrete calcification on its surface on the anterior wall of bladder. The other radiologic investigations showed no other important pathologies. During the cystoscopy the same polypoid

**Table 1**  
IHC results of the patient mass.

SMA: negative	EMA: (scattered cell are positive)
NSE: negative	CD10: positive (in 10% of tumor cells)
Chromogranin: negative	Ki 67: positive (in 10% of tumor cells)
CK: negative	
Vimentin: ±	
CD 117: negative	
S100: negative	
CD 45: negative	
CD 34: negative	
CK 20: negative	
CK 7: negative	

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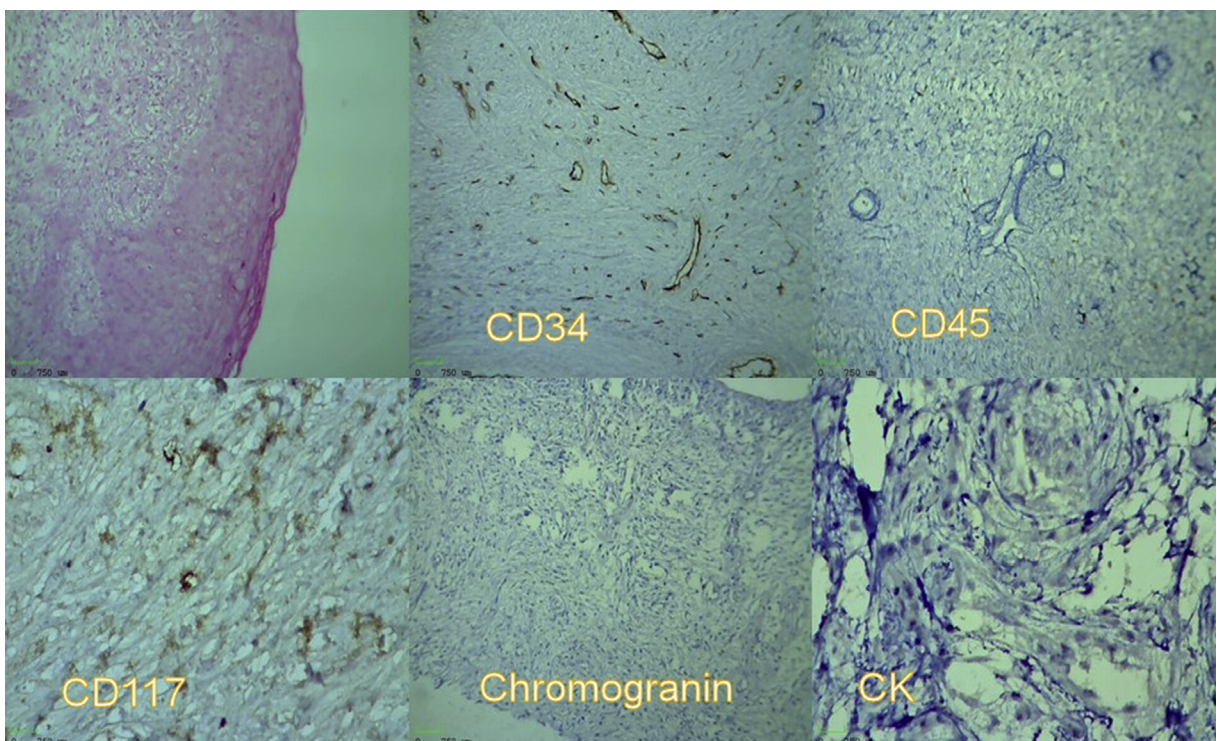
E-mail addresses: [drmmoradi@gmail.com](mailto:drmmoradi@gmail.com), [drmoradi@kums.ac.ir](mailto:drmoradi@kums.ac.ir) (M. Moradi), [shojaeishahram@yahoo.com](mailto:shojaeishahram@yahoo.com) (S. Shojaei), [kaveh.kaseb@yahoo.com](mailto:kaveh.kaseb@yahoo.com) (K. Kaseb), [haressrezaee@yahoo.com](mailto:haressrezaee@yahoo.com) (H. Rezaee).



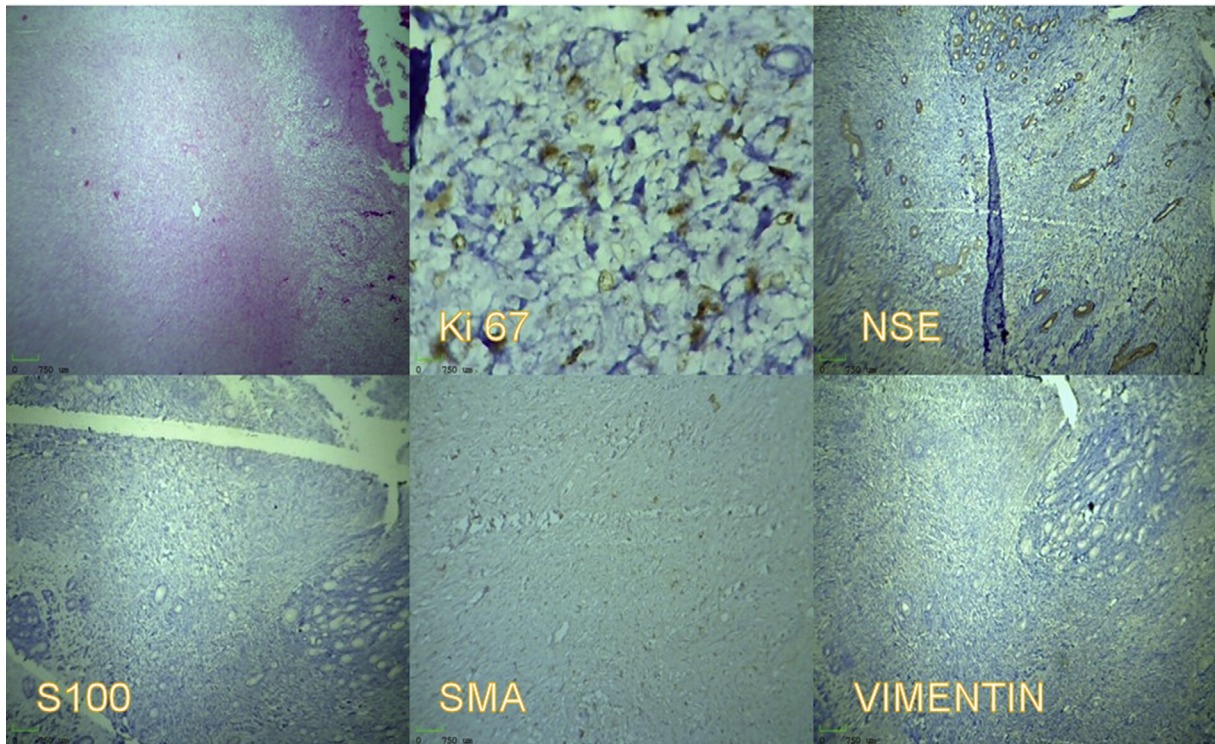
**Fig. 1.** Immunohistochemistry staining of the mass.

and pedunculated mass with calcification on anterior wall obviously seen. The mass resected completely (TUR-BT) but hardly due to calcified lobulated margins. Macroscopically tumor was creamy-gray and  $5 \times 4 \times 3$  cm. Microscopically the tumor had highly clear

cytoplasm with severe precipitation of calcium in the mass with focal glandular structures specific for PCCUB. Muscularis propria was free of tumor. The IHC results of tumor are represented in [Table 1](#) and [Figs. 1–3](#). Because of patient dissatisfaction radical



**Fig. 2.** Immunohistochemistry staining of the mass (continue).



**Fig. 3.** Immunohistochemistry staining of the mass (continue).

cystectomy and chemoradiotherapy ignored.

### Discussion

PCCUB is an uncommon tumor which is diagnosed through its histopathologic characteristics in IHC.<sup>3</sup> The first time PCCUB reported by Dow & Young Jr. in 1968 and up to now less than 50 cases has been reported.<sup>3</sup> Mostly of cases tumor size has been 1–7 cm and often reported on trigone, posterior and left wall of bladder.<sup>1</sup> In pathologic study “cells with abundant clear cytoplasm arranged in solid, glandular or tubulocystic patterns” reports.<sup>2,4,5</sup> In the majority of reported cases in IHC results CA 125, CK 7, Ki 67, P53, CEA and AMACR were positive.<sup>3–5</sup> Because of low reported cases still definite treatment is unclear, however TUR and cystectomy are as the most common procedure of treatment. The treatments which have been done for this tumor are TURBT, radiotherapy, chemotherapy, partial or radical cystectomy.<sup>3</sup> The effect of this therapeutic modalities on long term survival and prognosis of patients due to short follow up of reported cases is unclear too, although can conclude poorer prognosis of this tumor in comparison with common UC of bladder.<sup>1,3</sup> Intravesical chemotherapy following TURBT cannot prevent recurrence of tumor.<sup>3</sup> In our case no significant difference in IHC results viewed but severe macro and microscopic calcification of tumor to the knowledge we have seems as the first time is reported despite all previous reports. Despite the

gross tumor size muscular invasion was not occurred and during 6 months follow up no recurrence observed in cystoscopy.

### Conclusion

PCCUBs are rare. As definite diagnosis is made by IHC and precise treatment and outcome of interventions are vague, we suggest maybe it is better that all urologist report their cases in order to solve this problems.

### Conflicts of interest

None.

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