Granular cell tumor of the Urinary bladder: Case Report

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ABSTRACT
Granular cell tumors (GCT) are extremely rare lesions of the urinary bladder with only 12 cases have been reported in the literature. Although usually benign, these lesions are commonly confused for malignant tumors since they may present as solid tumors with ill defined margins and ulcerating surface and hence could be confused with transitional, squamous cell carcinoma or sarcoma in the urinary bladder. Only two cases of malignant granular cell tumor of the urinary bladder were reported. A careful pathologic assessment for establishing the appropriate diagnosis is required and the treatment is either a conservative or aggressive surgical treatment for benign or localized malignant GCT of the urinary bladder, respectively. We report the case of a forty six year old patient who had benign GCT of the urinary bladder.

Key words: Granular cell tumor, urinary bladder, S100 protein, immunohistochemistry

ÖZET
İdrar Kesesinin Granüler hücreli tümörü: Olgu Sunumu
Granüler hücreli tümörler (GHT) literatürde sadece 12 adet olgu sunumu ile belirtilmiş olan idrar kesesinin oldukça nadir görülen lezyonlardır. Genellikle iyi huylu olgamalarına rağmen bu tümörler kötü huylu tümörler ile karşılaştırılır. Çünkü bu lezyonlar idrar kesesinde sarkom veya transizyonel skamoz hücreli kanseri ile karşılaşıldıklarında dolaylı yuzyıl olarak sınıflandırılır ve sınırlar düzgün solid tümörler olarak görülebilir. İdrar yollarının kötü huylu granüler hücreli tümörü sadece iki olguda bildirilmiştir. Uygun tanı konulabilmesi için dikkatli bir patolojik değerlendirme gereklidir. İyi huylu tümörlerin tedavisinde semptomatik koruyucu tedavi iken, kötü huylu tümörlerin tedavisini cerrahidir. İdrar kesesinde iyi huylu granüler hücreli tümörü olan bir hastayla olgu sunumu olarak bildiriyoruz.

Anahtar Sözcükler: Granüler hücreli tümör, idrar kesesi, S100 protein, immünohistokimya

CASE REPORT
A forty six year-old lady presented with abdominal pain associated with nausea and vomiting of 1 week duration. She reported intermittent painless hematuria for the last year associated with some urinary symptoms such as dysuria, urgency and frequency. The patient has a significant smoking history. Her temperature was 97.5, breathing at 18 breaths per minute, blood pressure 170/79 and pulse of 80 beats per minute. Physical exam was negative except for suprapubic tenderness. Laboratory studies showed white count of 9.9 k, Hemoglobin 11.7 mg/dl, hematocrit 35.8 %, creatinine 0.8mg/dl. Urinalysis showed 10 RBC and 5 WBC/HPF. Urine culture and Cytology were negative. CT pelvis showed a soft tissue mass along the anterior bladder wall on the left measuring 3.5 cm. MRI of the pelvis with intravenous gadolinium revealed solid well defined intramural mass 2.5x2.9x2cm at the anterior bladder wall (Figure 1). Cystoscopy revealed the presence of submucosal mass near the dome of the bladder to the left of the midline with no intravesical extension. The remainder of the urinary bladder was unremarkable except for mild erythema along the posterior bladder wall. Under general anesthesia, patient underwent bladder biopsy with fulguration. Microscopic examination of the tissue revealed monotonous cells arranged in sheets with abundant eosinophil cytoplasm and small blunt nuclei with no mitotic activity and no necrosis (Figure 2-3). Immunohistochemical studies showed the tumor cell to be S-100 positive consistent with granular cell tumor of the urinary bladder (Figure 4). Neuroendocrine markers such as neuron specific enolase and chromogranin were negative. Patient symptoms improved.

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Figure 1. MRI, T2. An oval solid mass 2.5 x 2.9 x 2 cm, homogeneous with smooth margins within the anterior bladder wall with no evidence of bladder outlet obstruction.

Figure 2. Histology of benign granular cell tumor of the bladder (Haematoxylin and eosin stain, Lower Power).

Figure 3. Histology of benign granular cell tumor of the bladder (Haematoxylin and eosin stain, High Power).

Figure 4. Immunohistochemical staining of granular cell tumor of the bladder, granular cytoplasm is reactive for S-100 protein.

DISCUSSION
Granular cell tumors are unusual, rare neoplasm; they most commonly involve the head and neck region in 30-50% of cases (1), especially the tongue. Involvement of the genitourinary system is rare with only 12 cases of granular cell tumor of the urinary bladder were reported.

Granular cell tumor was initially described by Abrikosoff in 1926 (2). It is believed to be a benign tumor that arises from neural cells (3, 4) probably the Schwann cell. Malignant GCT comprise fewer than 2% of all GCT (5). Only two cases of malignant granular cell tumors were reported in the literature (6, 7). In few cases of granular cell tumors, Schwann cells in the pathology were not present (8).

Granular cell tumors (GCT) are typically solitary and smaller than 3 cm. Up to 10% of GCT are multiple (9, 10). GCT are particularly multiple in black patients (11, 12).

Granular cell tumors are usually benign but are commonly confused for malignant tumors since these lesions may present as solid tumors with ill-defined margins and ulcerating surface and their size vary up to 12 cm, and hence could be confused with transitional, squamous cell carcinoma or sarcoma in the urinary bladder. Although most patients affected are middle-aged, it can affect persons of varying ages with a peak incidence in the fourth through the sixth decades of life. A slight female predominance exists with an estimated female to male ratio of approximately 3:2.

The malignant lesions are usually larger (4-15 cm) and may be locally destructive, causing symptoms depending on the site (Pressure, obstruction, hemorrhage, ulceration, secondary infection). The malignant GCT has two distinct variants. The first variant has a benign histopathology indistinguishable from those found in benign tumors (13). The second consists of lesions that are more readily identifiable as malignant. Necrosis, nuclear pleomorphism, spindling and increased mitotic activity are the features of malignancy (13, 14).

The symptoms of GCT depend on the location and the size of these tumors. Gross hematuria is the most common symptom in patients with GCT of the urinary bladder but the
patient may be asymptomatic and GCT could be found incidentally.

The tumor cells are large polygonal, oval or bipolar cells with abundant, fine or coarsely granular eosinophilic cytoplasm with well defined cell borders and small, pale-staining or vesicular nucleus eccentrically located in the cell. They can be arranged in nests or sheets. Granular cells demonstrating nuclear enlargement, hyperchromatism and pleomorphism, or with mitotic activity, are suggestive of malignant variant of this tumor. The confirmatory immunohistochemical stain pattern consists of positive staining for S100 protein, neuron specific enolase, laminin, CD 68 and various myelin protein (15) and negative staining with epithelial (cytokeratins), sarcoma (vimentin, desmin, alpha smooth muscle actin) markers. The non reactivity to epithelial and muscle markers differentiates GCT from carcinomas and sarcomas (16).

Granular cell tumors (GCT) mostly follow a clinically benign course. Conservative surgical treatment with transurethral resection of the tumor or partial cystectomy appears to be the treatment of choice. Malignant GCT is extremely rare and the treatment include radical cystectomy.

REFERENCES