

PHEOCHROMOCYTOMA OF THE URINARY BLADDER

G. POURMAND AND M.R. NIKOBAKHT

From the Department of Urology, Sina Hospital, Tehran University of Medical Sciences, Tehran, Islamic Republic of Iran

ABSTRACT

One of the rarest tumors involving the urinary bladder is the pheochromocytoma. A case of pheochromocytoma of the urinary bladder in a young female suffering from headache and cystitis for many years is reported. Twenty-four hour urine VMA was normal. The patient was never hypertensive except during surgical manipulation of the tumor. Biopsy of the bladder resulted in massive hemorrhage.

MJIRI, Vol.2, No.1, 75-77, 1988

INTRODUCTION

Pheochromocytoma is a disease less often encountered by urologists. Pheochromocytoma of the urinary bladder was first reported in 1953 by Zimmerman.¹ Pheochromocytomas (paraganglionomas) of the bladder are mostly benign and constitute 0.4% of all urinary bladder tumors.²

In this article one case of paraganglionoma of the bladder is presented and the current literature reviewed.

CASE REPORT

The patient is a 37 year old female from Andimeshk with a 15 year history of cigarette smoking. She was admitted on August 23, 1987 with complaints of right flank pain and dysuria of 3 months duration with a previous history of hematuria. The patient had had severe headaches for the past 10 years but these had no association with micturition. Upon her admission the patient's blood pressure was always within normal limits while micturating. Routine laboratory workups were also normal. An IVP revealed a small stone in the inferior calyx of the right kidney as well as a filling defect in the left side of the urinary bladder (Fig. 1). Cystoscopy revealed a nodule measuring 1x1 cm above the left ureteric orifice that was covered by normal appearing mucosa. A biopsy was performed and the pathological report was paraganglionoma.

Twenty-four hour urine vanillyl-mandellic acid (VMA) and serum catecholamines were within normal limits. A partial cystectomy was performed for the

patient. The blood pressure was continuously monitored during the operation. During surgical manipulation the blood pressure rose from 110/70 mmHg to 170/110 and returned to a normal level 15 minutes after surgical excision of the tumor. Ten days post-op the patient was discharged from the hospital without com-

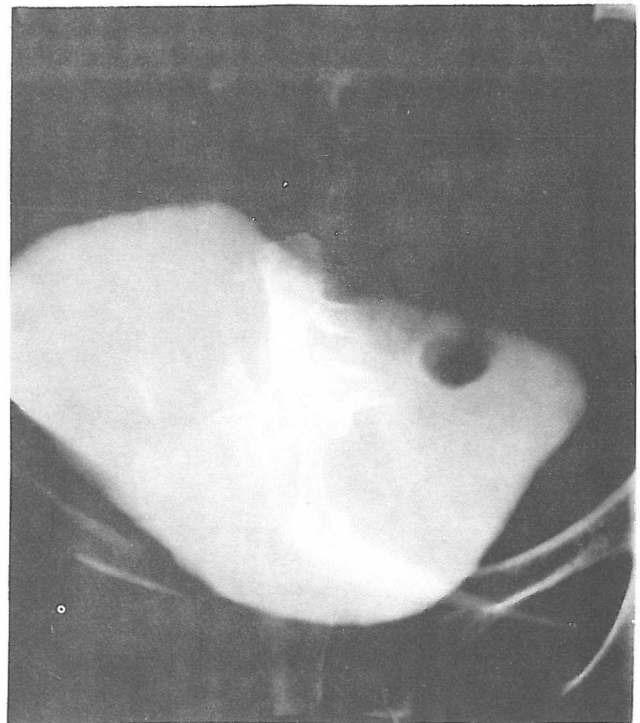


Fig 1: Intravenous urography of the patient with bladder pheochromocytoma demonstrating a filling defect in the left side of the bladder.

plications. Subsequent blood pressure values were within normal limits and the patient's headaches did not recur. The final histopathological report was paraganglionoma of the urinary bladder with mural involvement (Fig. 2).

DISCUSSION

Up until 1967, a total of 24 cases of urinary bladder pheochromocytomas were reported of which only 30% had visible masses upon cystoscopy.³ The majority of patients (56%) had presented with hematuria and all but two patients were hypertensive.⁴ Four of the patients had malignant tumors with metastases, three patients had lymph node involvement, and one patient had tumor invasion from the bladder to the pubic symphysis while subsequent examinations revealed mesenteric and liver metastases.⁴

The tumor was seen with an equal frequency in all age groups and neither benign nor malignant tumors

showed any specific age group preponderance. Most of the patients had presented with symptoms such as headache, fainting upon micturition, and gross hematuria.² The appearance of symptoms during micturition may be the only clue to diagnosis.

Pheochromocytomas of the urinary bladder are usually located in the submucosa, but even in benign cases some invasion into the mucosa exists. This results in the formation of elevated nodules in the cavity of the bladder with eventual injury to the epithelial lining and hemorrhage.

Histologically these tumors are highly vascular and have nests of cells (zellballen) lining their vascular sinuses. These nests of cells are composed of typical acidophilic, polygonal cells having elliptical nuclei.⁵ Other names attributed to this tumor are chromaffinoma and paraganglionoma. Some authors have used the term paraganglionoma for those tumors not having catecholamine secretions.

Measuring serum and urine catecholamine levels is

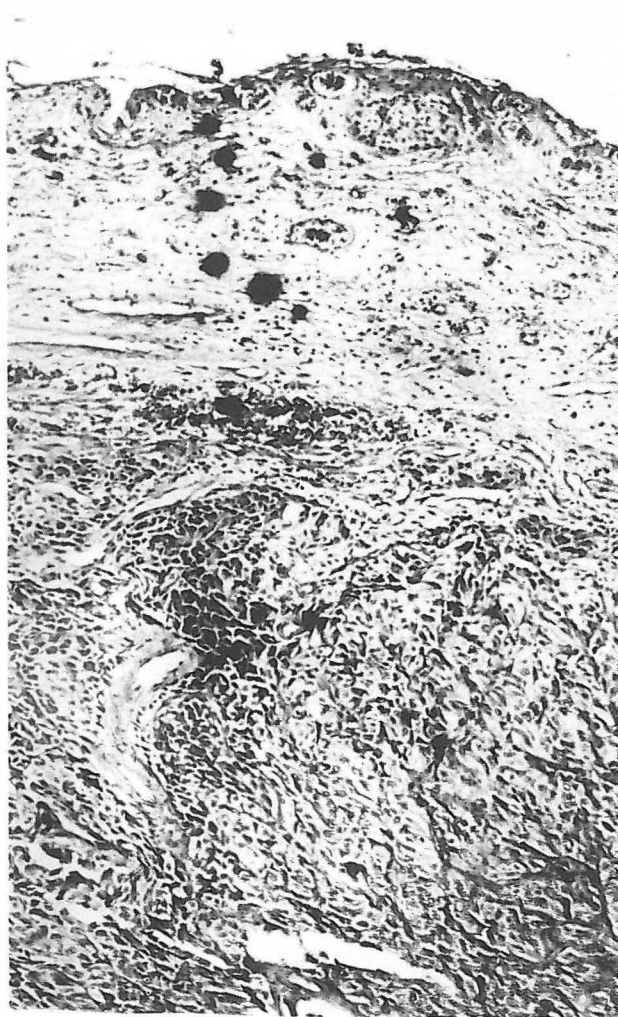


Fig 2: Microscopic view of the bladder pheochromocytoma (paraganglionoma), lower magnification.

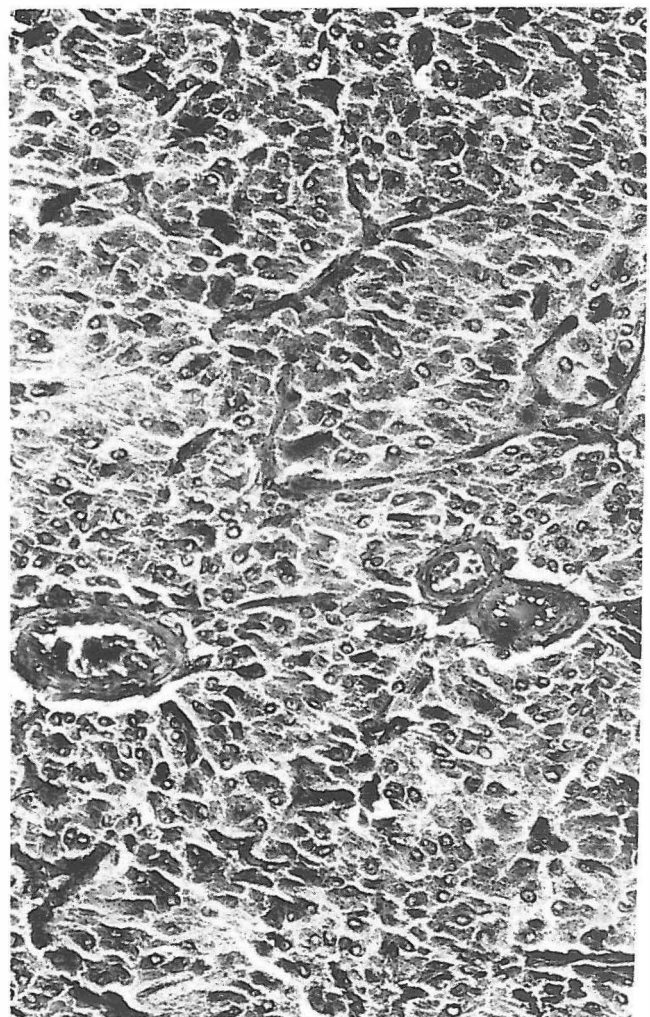


Fig 3: Pheochromocytoma (paraganglionoma) of the bladder. (Higher magnification).

of diagnostic importance; approximately 60% of the patients have elevated levels of catecholamine in their serum and urine.

Malignant pheochromocytoma constitutes 5-10% of all bladder tumors.² Differentiation between benign and malignant paraganglionomas is difficult. i.e. malignancy can not be determined by histological studies or the extent of mural invasion. It has been suggested that in malignant pheochromocytoma serum levels of dopa and dopamine are elevated. However malignancy can only be determined by the finding of secretory chromaffin cells in sites where this tissue does not normally exist. Most authors consider local recurrence, multiple metastases and vascular invasion as signs of malignancy.

Lymph node metastases have been reported in some cases of paraganglionoma. The most common sites for these metastases are the hypogastric and iliac lymph nodes, locations which do not normally contain chromaffin tissue. Also, there have been cases in which iliac node involvement existed but tumor recurrence was not seen. The treatment of choice is partial cystectomy.

Anesthesia

The best premedication for alleviating anxiety and controlling blood pressure is phenoxybenzamine. In addition sodium pentobarbital has been administered selectively to decrease adrenal activity. It is for this reason that sodium thiopental is the drug of choice for induction of anesthesia.

Succinylcholine has been used for intubation with a minimal amount of curare to prevent muscle fasciculation resulting from catecholamine release during the surgical manipulation of the tumor. Diethyl ether, halothane, and methoxyfluorane can be utilized for the anesthesia.

Blood pressure is monitored through a CVP catheter. If an adrenal crisis occurs, intravenous phentolamine is helpful.

Surgery

In the treatment of pheochromocytoma, the surgeon should always bear in mind that secondary tumors may exist in other sites. In the past, transabdominal palpation of the sympathetic chains along the inferior vena cava, aorta, celiac ganglion, hypogastric plexus, and periaortic tissues known as Zuckerkandl bodies was performed. However due to the possibility of catecholamine release and impending adrenal crisis this method is not recommended, rather decision is based on angiographic findings.

It is essential that local lymph node resection be performed in addition to partial cystectomy since local lymph node metastases have been reported in different series and their presence is associated with a poorer prognosis.^{3,4}

Post-op follow up

Long term follow up with close observation is suggested for these patients, since secondary benign pheochromocytomas have been found in some cases. Annual determination of catecholamine levels and frequent blood pressure monitoring in patients with documented pheochromocytoma is mandatory.

REFERENCES

1. Zimmerman JJ, Biron RE, MacMahon HE: Pheochromocytoma of the urinary bladder. *N Engl J Med* 249: 25, 1953.
2. Flanigan RC, Wittmann RP, Huhn RG, Davis CJ: Malignant pheochromocytoma of urinary bladder. *Urology* 16 (4): 368-8, 1980.
3. Bourne RC, Beltaos E: Pheochromocytoma of the bladder: Case report and summary of literature. *J Urol* 98: 361, 1967.
4. Gittes RF, Mahoney EM: Pheochromocytoma. *Urol Clin North Am* 4 (2): 239-52, 1977.
5. Glenner GC, Girinley PM: Tumors of the extra-adrenal paraganglion system (including chemoreceptors). *Urinary bladder paraganglionoma*. Washington D.C. Armed Forces Institute of Pathology (AFIP), 80, 1974.