

Diagnosis and treatment of 29 cases of adrenal ganglioneuroma

J. LI, C.-H. YANG, L.-M. LI

Department of Urology, Tianjin Medical University General Hospital, Tianjin, China

Abstract. – **AIM:** To evaluate diagnosis and treatment experience for adrenal ganglioneuroma and provide data for clinical surgery.

PATIENTS AND METHODS: Analysis clinical feature and iconography and endocrine examination and clinical data of 29-cases adrenal ganglioneuroma in our Hospital.

RESULTS: Back discomfort in 10 cases and convulsivum dizziness in 6-cases (hypertension in 2 cases), central obesity in 1 case. 12-cases were found by physical examination. 9-cases were diagnosed as adrenal ganglioneuroma and others were diagnosed as adrenal tumor. After operation, all of the cases were diagnosed as adrenal ganglioneuroma by pathology. Beside one patient were still dizzy with BP (blood pressure): 150/95 mmHg, all of patients completely recovered.

CONCLUSIONS: For diagnosis on adrenal ganglioneuroma, we should depend on iconography and pathology. The operation is main method and most of patients can be cured.

Key Words:

Adrenal gland neoplasms, Ganglioneuroma.

In this report, we present 29 cases adrenal ganglioneuroma admitted from July 1988 to August 2011 in our Hospital diagnosed by pathology.

Patients and Methods

Clinical Material

There were 29 patients (18 males and 11 females, mean age 35 years) admitted to our Hospital from July 1988 to August 2011. The tumor occurred in the left side in 11 cases (11/29), in the right side in 17 cases (17/29, 1 case of unilateral multifocal tumors), both sides in 1. The tumor was 2.5-12.3 cm in diameter. The waist discomfort in 10 cases, paroxysmal dizziness and lack of power in 6 cases (6/29, 2 patients with paroxysmal hypertension history for 2 months and tumor in both sides). Central obesity in 1 cases (1/29). 12 cases (12/29) were accidentally discovered by B ultrasound discovery for a physical examination. 29 patients were examined by routine 24h VMA (vanillyl mandelic acid) in urine, plasma aldosterone, renin angiotensin II, urine cortisol and electrolyte biochemical examination. Patients with bilateral lesions (1/29) had 2 times (examined 3 times) beyond the normal range by 24h VMA urine examination. The level of the patient (1/29) who had maximum tumors (tumors with a diameter of 12.3 cm) was beyond the normal range both of the 3 times by 24h VMA urine examination, with dizziness and waist discomfort, normal by other examination. The level of plasma aldosterone and angiotensin II were beyond the normal range in 2 cases (2/29) and the others were normal. The centripetal obesity level (1/29) was beyond the normal range for 24h urine 17-hydroxysteroid, without hypertension, and other examinations were normal.

The remaining patients for VMA, aldosterone, angiotensin II, renin, hematuria, cortisol, and electrolyte biochemical tests were normal. 29 patients underwent B ultrasound and CT examination. For undergoing MRI (magnetic resonance imaging) in

Introduction

Ganglioneuroma (GN) is a rare tumor arising from the neural crest tissue and is most commonly located in the posterior mediastinum and retroperitoneum, and rarely in the adrenal gland¹. Because imaging procedures such as ultrasonography (US) and computed tomography (CT) have become prevalent, the number of incidentally identified adrenal GNs has recently increased². Adrenal GN is usually hormonally silent, and can therefore asymptomatic even when the size of tumor is large³. Therefore, it is generally difficult to obtain a precise differential diagnosis of adrenal GN before surgery. In addition, the size and imaging characteristics of adrenal GNs are variable, and thus, some of them are very similar to other adrenal tumors such as adrenocortical carcinoma (ACC) and pheochromocytoma (PC)⁴.

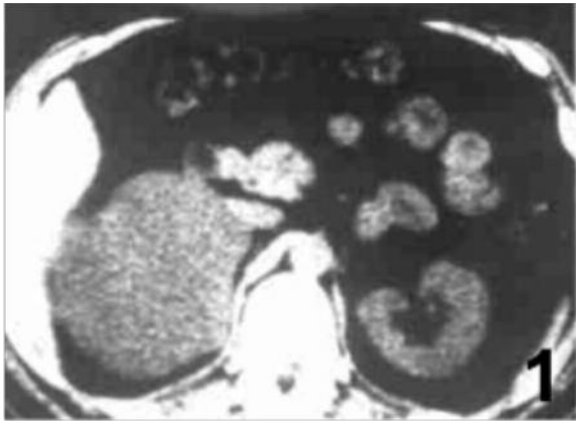


Figure 1. A plain CT scan showed a relatively homogeneous adrenal tumor.

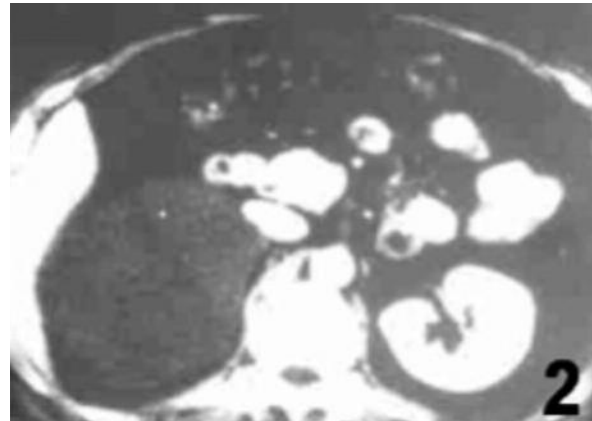


Figure 2. A dynamic enhanced CT scan showed no significant change.

10 patients (10/29). B ultrasound and CT scan showed adrenal solid mass lesions and the boundary of tumors were clear and smooth. B ultrasound examination showed a space-occupying lesions hypoechoic. CT showed low density or mixed density. A plain CT scan showed a relatively homogeneous round or oval-shaped adrenal tumor and the level were 19-40 Hu (Figure 1). A dynamic enhanced CT scan showed no significant change, and the level were 25-60 Hu (Figure 2). CT scan in nine cases containing fine punctate calcification, which were lobulated, and the patients were definitively diagnosed before operating. T1-weighted MRI showed a homogeneous adrenal tumor with a low-signal intensity (Figure 3). T2-weighted MRI showed a heterogeneous adrenal tumor with a high-signal intensity (Figure 4).

Surgical Methods

29 patients were underwent surgical treatment. 11 cases underwent open surgical removal with-

out carrying out Laparoscopy, and 2 patients in which were diagnosed adrenal ganglioneuroma before surgical treatment. 2 cases also underwent open surgical removal because the tumor was large. The remaining 16 patients were removed tumor by after laparoscopic excision and 3 cases in which were diagnosed adrenal ganglioneuroma before operation. We carried out open surgery through incision in 11th intercostal or 11 ribs. 2 cases whose VMA levels in above ranges were given alpha blockers a week in advance. 26 cases were removed adrenal tumors and the ipsilateral adrenal completely. 2 cases underwent partial adrenalectomy. One case with bilateral adrenal masses was removed the left side.

Results

9-cases were diagnosed by CT as adrenal ganglioneuroma and others were diagnosed as adren-

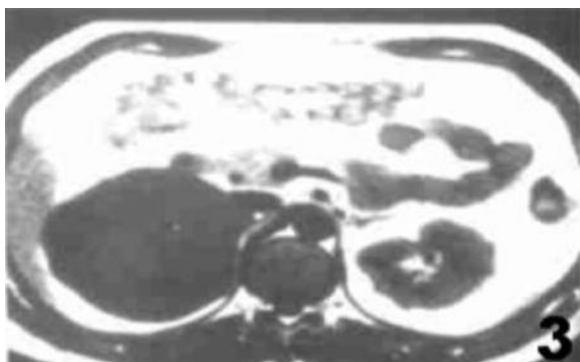


Figure 3. T1-weighted MRI showed a homogeneous adrenal tumor with a low-signal intensity.

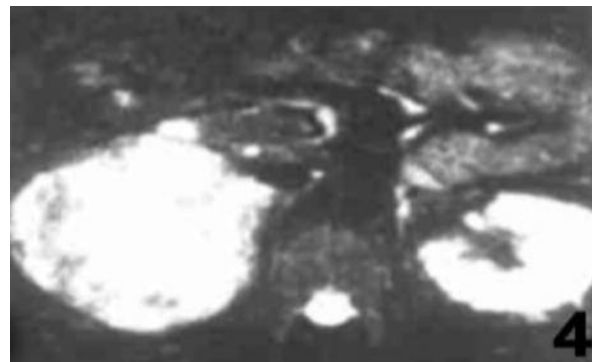


Figure 4. T2-weighted MRI showed a heterogeneous adrenal tumor with a high-signal intensity.

al tumor in the 29 patients admitted to our Hospital. There were not significant intraoperative blood pressure fluctuations in operation. The removed tumors had complete capsule measuring about 10 cm × 9.5 cm × 9 cm. 1 case included four complete capsule tumor tightly together with a diameter of 2.5 cm to 4.0 cm. The sections were light brown, with a fleshy appearance, showing nodules with gelatinous areas. There were hemorrhage and necrosis in 2 cases of tumor and cystic degeneration in 1 case. The slice containing ganglion cells were found by pathological evaluation (Figure 5), which was signal of adrenal ganglioneuroma tumor. Largest tumor lesions contained the pheochromocytoma ingredients. The symptoms of patients who had bilateral adrenal masses were alleviated after operation, and the remaining patients preoperative symptoms disappeared. Patients with maximum tumor patients were carried out B ultrasonic examination annually for 5 years and there were no tumor recurrence or metastasis. We followed-up patients with telephone 2 times a year. Patients with bilateral tumors still had paroxysmal dizziness, with blood pressure of 150/95 mmHg, and 24h urinary VMA was above the normal level, but lower than the preoperative level. Blood pressure was normal in other patients who without discomfort and no tumor recurrence or metastasis. Blood pressure and heart rate of the central obesity patients were normal and they had not given glucocorticoid treatment.

Discussion

Ganglioneuroma is a rare tumor arising from the neural crest tissue and is most commonly located in the posterior mediastinum and retroperitoneum, and rarely in the adrenal gland^{1,5}. Adrenal ganglioneuroma is uncommon below the age of 30 and its frequency increases progressively in adults and the elderly⁶. The prevalence approaches 0.2% in young patients, 3% in people over 50 years of age, and reaches 7% in those over 70 years of age⁷. In a Brazilian study⁸, adrenal ganglioneuroma was found in 2.5% of patients undergoing chest and abdomen CT. A more recent study from China reported that the misdiagnosis rate of adrenal GNs on CT and MRI before surgery was 64.7%⁹. Tumor could increase dopamine level which caused hypertension². We did not use dopamine detection.

In this report, we presented 29 cases of adrenal GN that were differentially diagnosed. The treat-

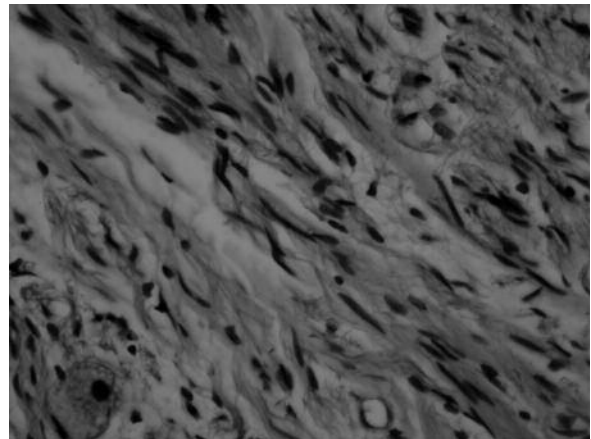


Figure 5. A microscopic view of the adrenal GN. The tumor was composed of mature ganglion cells and Schwann cells with abundant fibrous stroma (H&E, x 400).

ment of any adrenal mass > 6 cm or smaller but progressive growth objectified in various imaging tests, as in our case report was unilateral adrenalectomy, having completed the full functionality study. We used iconography and pathology to diagnose adrenal ganglioneuroma. 9-cases were diagnosed by CT as adrenal ganglioneuroma and others were diagnosed as adrenal tumor in the 29 patients admitted to our Hospital. Two cases of aldosterone increased and one case of abnormal urinary 17 ketosteroids with central obesity may be associated with tumor stimulates the adrenal cortex.

At imaging studies, it appears as a relatively homogeneous, encapsulated mass, with well defined edges and without invading nearby structures. Calcification occurs in 40%60% of cases.

Conclusions

We present here a report of 29 cases adrenal ganglioneuroma, which indicates careful evaluation by endocrine examinations and multiple imaging procedures is necessary to provide a differential diagnosis, and surgery leads to definitive cure, most of the time.

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