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Uncommon Adrenal Tumors: A Case Series

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Adrenal tumors are very rare. These lesions are called adrenal incidentalomas because they are incidentally detected through imaging procedure. Its prevalence ranges from 1.4% to 2.9% worldwide. Clinical suspicion, laboratory testing, Imaging and histopathological examination needed for definite diagnosis. We report three cases of adrenal tumors received at our institute between the year 2021 and 2022. Pheochromocytoma of Adrenal gland scaled score (PASS) tells about the prognosis based on histopathological findings. Earliest management is essential as these tumors are related with secretion of hormones and may have a poor prognosis.

Keywords: Incidentalomas; management; hormones; prognosis.

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1. INTRODUCTION

Adrenal gland is an endocrine gland and divided into cortex and medulla. Cortex is derived from mesoderm and produces steroid hormones. Medulla is derived from neural crest and produces catecholamines. Adrenal tumors are very rare lesions and are called incidentalomas because they are incidentally detected through imaging procedure. Its prevalence ranges from 1.4% to 2.9% worldwide. Thomas A et al concluded that the incidence of adrenal masses found on abdominal CT scans was between 0.6% and 1.3% whereas the incidence of these masses on all CT scans including thoracic, abdominal and pelvic was between 0.4% and 4% [1]. We reported 3 cases of adrenal tumors received at our institute from 2021-2022 for a period of 1 year.

2. CASE PRESENTATION

Case 1-A 38 year old male presented with recurrent bouts of high blood pressure, headache, palpitations, excessive sweating, dizziness, abdominal discomfort and weight loss. He was diabetic and hypertensive for about 5 years. On clinical examination he was conscious and oriented. Pulse was 104 beats/min. Blood pressure was 190/100 mm Hg on supine position and 164/110mm Hg on standing position. Laboratory investigation revealed 24 hr urine fractionated metanephrine and normetanephrine levels greater than 10000mcg/dl. Cardiac examination showed moderate concentric left ventricular hypertrophy and mild mitral regurgitation. Contrast Enhanced Computed tomography scan revealed a round soft tissue density, heterogeneously enhancing lesion of size 9.0x9.7x9.6cm in right suprarenal region (Fig. 1). Right adrenal gland is not seen separately from the lesion. Left adrenal gland is normal. Grossly we received a circumscribed, globular, bosselated soft tissue mass measuring 10x 9x7 cm. Cut surface was bulging and vague lobulations was present. The tumor is tan to red brown in colour. Microscopically marked pleomorphic tumor cells seen with prominent nucleoli, stippled chromatin and abundant amount of eosinophilic cytoplasm in nested and trabecular pattern with diffuse sheets. Tumor cells were strongly positive for IHC markers Chromogranin A and Gata-3. We reported the case as Pheochromocytoma.

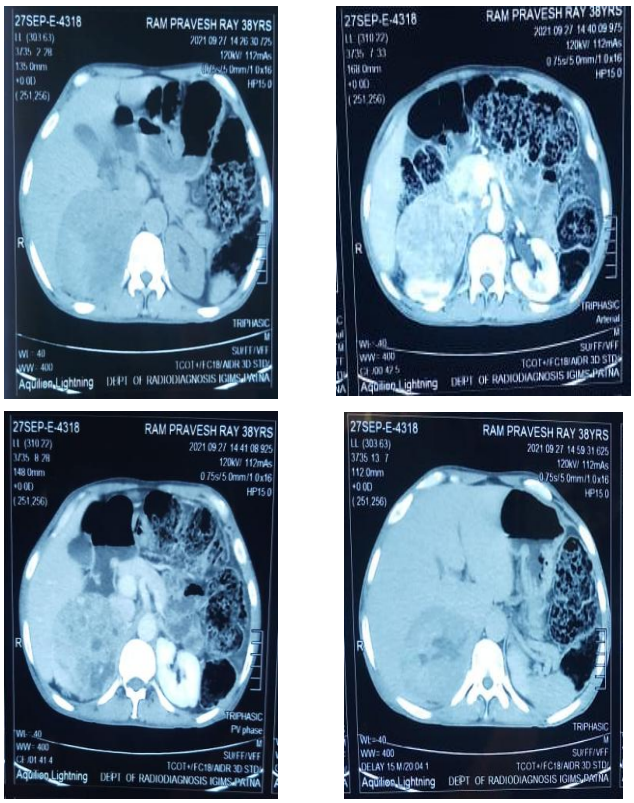

Fig. 1. CECT showing right adrenal mass

Case 2-A 18 year old male presented with gradually increasing abdominal mass and weight loss from last 1 year. He was not diabetic and hypertensive. Free plasma normetanephrine level was 2170 ng/L.24 hr urine VMA was 4.88mg. Serum cortisol was 15.69mcg/dl. CT scan showed large heterogenous lesion in right suprarenal region of size-6.5x7.5cm (Fig. 2). PET–CT scan revealed intense FDG uptake in a well circumscribed hypervascular centrally necrotic suprarenal mass lesion(maximum dimension-85mm) and another smaller well circumscribed heterogenous left adrenal lesion (maximum dimension-20mm). Grossly right adrenal mass was of size-8x6x3cm. Cut section showed a grey brown lesion of size 4x4x3cm involving the adrenal medulla and focally reaching upto the cortex. Left adrenal mass was of size-5x3x1cm. Cut section showed greyish tan haemorrhagic lesion of size 3x3x1cm. Microscopically sections from bilateral adrenal tissues displayed tumour cells in nested pattern. Cells were large, polygonal, uniform with fine granular vacuolated cytoplasm and round to oval nucleus, nucleoli prominent. Brisk atypical mitosis also noted. Tumor cells were positive for IHC-NSE and S-100.We reported the case as Bilateral Pheochromocytoma.

Fig. 2. Cut surface: Tumour is tan to red brown in colour

Case 3-A 19 months male child presented with severe ill health,abdominal mass and pain. The child was severely malnourished and failure to thrive. CECT abdomen revealed a large well defined soft tissue mass lesion of size-7.6x7.4x11.6cm with specks of calcification within is noted in the right suprarenal location (Fig. 3). We received 3 linear core biopsy of right suprarenal mass of size 1.2cm each. Histology demonstrated sheets of densely packed primitive embryonal cells. Intermixed with above sheets are single scattered binucleated, multinucleated cells with ample cytoplasm. No Homer-wright rosettes seen. Tumor cells were positive for IHC -S100 and negative for CD68. We reported the case as Anaplastic Neuroblastoma.

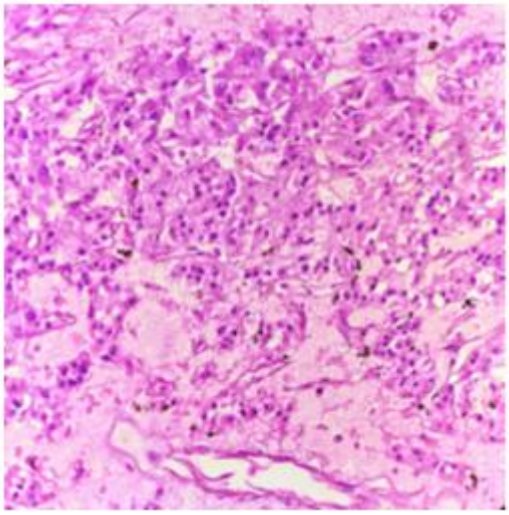

Fig. 3. Nested arrangement of tumour cells in H and E section (H and Es x 40)

Fig. 4. Tumor cells are strongly positive for IHC marker chromogranin

Fig. 5. Tumor cells are strongly positive for IHC marker GATA 3

3. DISCUSSION

Adrenal Incidentalomas are very difficult to diagnose whereas radiologic studies are the foundation for diagnosis [2]. Pheochromocytoma

occurs in people of all races although diagnosed less frequently in blacks and equal frequency in male and female. The peak incidence is between 3rd and 5th decade of life. Approximately 10% occur in children. Most of cases are sporadic and 10-25% are associated with genetic syndromes such as Von-Hippel Landau (VHL) disease , type 1 neurofibromatosis (NF1) and multiple endocrine neoplasia type 2 (MEN2) [3,4]. The clinical features are diverse ranging from asymptomatic to cardiac arrest. Hypertension is one of the most common clinical feature of pheochromocytoma. The prevalence of pheochromocytoma in patients with hypertension is 0.1-0.6%. Persistent or paroxysmal hypertension is the most frequent sign of pheochromocytoma in 90% of cases [5,6]. It is also associated with episodic headache, suderosis and tachycardia. Paraneoplastic syndromes are also seen for eg cushing syndrome (most common), diarrhea, hypokalemia, achlorhydria and polycythemia [7]. The increased level of serum catecholamines especially norepinephrine, urinary metanephrine, chromogranin A and adrenal gland mass confirm the diagnosis. The assessment of metanephrines in urine and serum are the most sensitive and reliable tests to confirm the diagnosis. Ancillary tests such as abdominal CT or MRI and I123-MIBG or I131-MIBG scintigraphy are also essential for tumor localization. CECT in case 1 showed heterogeneously enhancing lesion of size 9.0x9.7x9.6cm in right suprarenal region. PET CT scan in case 2 showed intense FDG uptake in a well circumscribed hypervascular centrally necrotic suprarenal mass lesion (maximum dimension-85mm) and another smaller well circumscribed heterogenous left adrenal lesion (maximum dimension -20mm). Pheochromocytoma produces excessive amounts of epinephrine and norepinephrine either continuosly or intermittently. Norepinephrine increases peripheral vascular resistance and leads to increased systolic and diastolic blood pressure. In case 1, both urinary epinephrine and nonepinephrine levels were elevated. The patient presented with classic triad of pheochromocytoma along with recurrent bouts of hypertension with dizziness, abdominal discomfort and weight loss. He was known diabetic and hypertensive for about 5 years. The cardiovascular complications include cardiac hypertrophy, heart failure, arrhythmias, ischeamic heart disease and acute myocardial infarction due to catecholamine secretion. Free plasma nor metanephrine level was 2170ng/L in case 2. Pheochromocytoma of Adrenal gland scaled score (PASS) tells about the prognosis based on histopathological findings. Malignant Pheochromocytoma can be recognized by PASS score greater than and equal to four with a sensitivity of 50% and a specificity of 45% [8]. PASS score in Case 1 and Case 2 are three. Malignant pheochromocytoma constitute 10% of cases. Malignancy is determine by presence of metastasis and direct invasion of surrounding structures. Giant pheochromocytomas (>7 cm in size) are rare entities and around 20 cases reported in literature [9,10,11]. Surgical excision is the treatment of choice and results in cure of hypertension. Neuroblastoma is the 3rd most common extracranial solid tumor in childhood [12]. It arises anywhere along the peripheral sympathetic nervous system. Its prevalence is approx 1/7000 live births. It is sporadic or nonfamilial in origin. It commonly arises from adrenal gland,nerve tissues of neck, chest, abdomen or pelvis. The marked pleomorphism and anaplastic features of the cells in these tumors demonstrated more aggressive clinical behavior. The exact incidence of anaplastic neuroblastoma is difficult to assume because of widely scattered case reports. Morphologic indicators of poor outcome are high mitosis-karyorrhexis index and lack of differentiation. MKI is calculated by using 5000 tumour cells as denominator.Microscopy in case 3 demonstrated singly scattered binucleated and multinucleated cells densely intermixed with primitive embryonal cells.

4. CONCLUSION

Primary adrenal tumors are very rare tumors including unilateral and bilateral Pheochromocytoma and Anaplastic neuroblastoma. Early diagnosis and treatment is essential because these tumors may secrete hormones and have a poor prognosis.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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