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CLINICAL PRESENTATION, MANAGEMENT AND OUTCOME OF RARE ENDOCRINE TUMOR: PHEOCROMOCYTOMA

General Surgery

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ABSTRACT

Pheochromocytoma is a rare endocrine tumor of the adrenal gland that can release high level of adrenalin and noradrenalin. These hormone producing cells are increases in pheochromocytoma which causes fight or flight type of responce. Extra adrenal pheochromocytoma also present in abdomen. Pheochromocytoma incidence is between third and fifth decade of life. It is very rare tumor and most of them are benign.

KEYWORDS

DISEASE:

These are tumours of the adrenal medulla and sympathetic ganglia that are derived from chromaffin cells and most commonly produce supraphysiological levels of circulating catecholamines. The prevalence of phaeochromocytoma in patients with hypertension is 0.1–0.6% with an overall prevalence of 0.05% in autopsy series.

Etiology:

Multiple endocrine neoplasia type 2 (MEN 2)
Familial paraganglioma (PG) syndrome
von Hippel–Lindau (VHL) syndrome
Neurofibromatosis (NF) type 1

Pathology

Phaeochromocytomas are greyish-pink on the cut surface and are usually highly vascularised. Areas of haemorrhage or necrosis are often observed . Microscopically, tumour cells are polygonal but the configuration varies considerably. The differentiation between malignant and benign tumours is difficult, except when metastases are present. An increased PASS (**p**haeochromocytoma of the **a**drenal gland **s**cale **s**core), a high number of Ki-67-positive cells, vascular invasion or a breached capsule all lean more towards malignant rather than benign. Phaeochromocytomas may also produce calcitonin, ACTH, vasoactive intestinal polypeptide (VIP) and parathyroid hormone-related protein (PTHrP). In patients with MEN 2, the onset of phaeochromocytoma is preceded by adrenomedullary hyperplasia, sometimes bilateral. Phaeochromocytoma is rarely malignant in MEN 2.

Clinical features

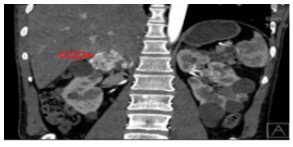
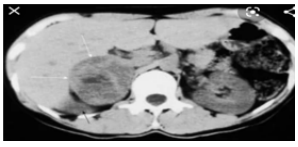
Symptoms and signs are caused by catecholamine excess and are typically intermittent . In total, 90% of patients with the combination of headache, palpitations and sweating in the presence of an adrenal tumour have a phaeochromocytoma. Hypertension may occur continuously, be intermittent or absent. A subset of patients are asymptomatic. More than 25% of apparently sporadic phaeochromocytomas are caused by germline mutations in the *RET, SDHB*, *SDHC, SDHD* and *NF1* genes; genetic testin for these and other genes is therefore recommended, particularly in those patients aged under 50 years.

Diagnosis

The first step in the diagnosis of a phaeochromocytoma is the measurement of adrenalin and noradrenalin level in the blood and then CECT abdomen or MRI imaging is important modality.

Treatment

Operative management is done by open or laparoscopic approach, in present modality most of pheochromocytoma is managed by laparoscopic adrenalectomy. Once a phaeochromocytoma has been diagnosed, an α-adrenoreceptor blocker (phenoxybenzamine) is used to block the effects of catecholamine excess and its consequences during surgery. A dose of 20 mg of phenoxybenzamine initially should be increased daily by 10 mg until a daily dose of 100–160 mg is achieved and the patient reports symptomatic postural hypotension. Additional β-blockade is required if tachycardia or arrhythmias develop; this should not be introduced until the patient is α-blocked. With adequate α-blockade preoperatively, anaesthesia should not be more hazardous than in patients with a non-functioning adrenal tumour; however, in some patients, dramatic changes in heart rate and blood pressure may occur and require sudden administration of pressor or vasodilator agents. A central venous catheter and invasive arterial monitoring are used. Special attention is required when the adrenal vein is ligated as a sudden drop in blood pressure may occur. The infusion of large volumes of fluid or administration of noradrenaline can be necessary to correct postoperative hypotension in the presence of unopposed α-blockade.

POSTOPERATIVE

Patients should be observed for 24 hours in the intensive care(ICU) or high dependancy unit as hypovolaemia and hypoglycaemia may occur. Lifelong yearly biochemical tests should be performed to identify recurrent, metastatic or metachronous phaeochromocytoma.

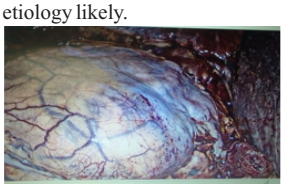
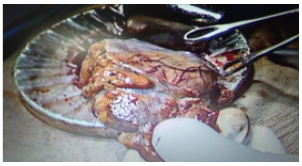
Malignant phaeochromocytoma

10% of phaeochromocytomas are malignant. The diagnosis of malignancy implies metastases of chromaffin tissue, most commonly to lymph nodes, bone and liver. Surgical excision is the only chance for cure. Even in patients with metastatic disease, tumour debulking can be considered to reduce the tumour burden and to control the catecholamine excess.

CASE REPORT:

A 52 year old female presented in opd with chief complain of generalized abdominal discomfort and significant weight loss since last 3 months. Patient having DMII since 3 years and she stopped treatment since 4 months other then this no any comorbidity present. On clinical examination of abdomen was soft without tenderness, lump, guarding or rigidity.

CECT Abdomen s/o well circumscribed,markedly enhancing heterogenous right adrenal lesion of size 44*41*38mm, p/o neoplastic etiology likely.

Patient was managed by right side laparoscopic adrenalectomy. Post operative patient was shifted to ICU for close vital monitoring. Blood

pressure was decreased below 90/70 which was managed by inotropics and vasopressin. Patient recovered on 2nd POD.

Histopathology finding s/o malignant pheochromocytoma of adrenal gland scaled score: Mitotic figure >3 per high power field-present, High grade nuclear pleomorphism, tumor necrosis and capsular invasion-present.

CONCLUSION:

Pheochromocytoma of adrenal gland is rare endocrine tumor with unique presentation of weight loss without any significant blood pressure changes operated with laparoscopic adrenalectomy and patient referred for chemotherapy.