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A CASE OF BIOCHEMICAL TUMOUR

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Abstract : Background - Pheochromocytoma (PCC) and paraganglioma are rare neuroendocrine tumors arising from chromaffin cells. PCCs are typically found in the adrenal medulla, accounting for 80 85 of cases. Estimates of the prevalence of PCC in hypertensive populations vary between 0.1 and 0.6(18). Case Presentation - A 42 year old male presented with complaints of pain abdomen for 1 month with no other co morbidities. Imaging studies found a left adrenal mass and bio-chemically proved as PCC. Laparotomy and surgical excision of the tumor was done. Conclusion - Advances in imaging and screening have led to an increased frequency of diagnosis of pheochromocytoma in normotensive and asymptomatic patients. It is estimated that 1.523 of all incidentally detected adrenal masses (incidentalomas) are PCCs(19).

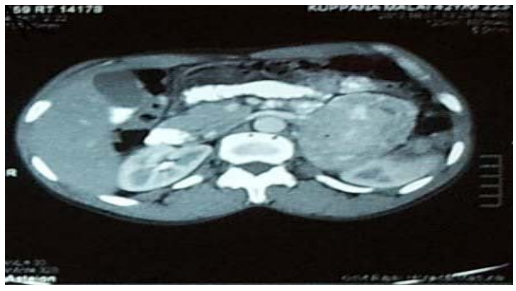
Keyword :pheochromocytoma, metanephrines, adrenalectomy

Introduction

The first account of pheochromocytoma was published in 1886 by Felix Frankel, who described a young woman suffering from intermittent attacks of palpitations, anxiety, vertigo, and headache. Autopsy revealed bilateral adrenal tumors that stained brown when treated with chromium salts. The characteristic positive chromaffin reaction lends these adrenomedullary tumors the name pheochromocytoma ("dusky-colored tumor" from the Greek phaios, or dusky). Successful surgical management of pheochromocytoma was initially described in 1926 by both César Roux and Charles Mayo. [20]

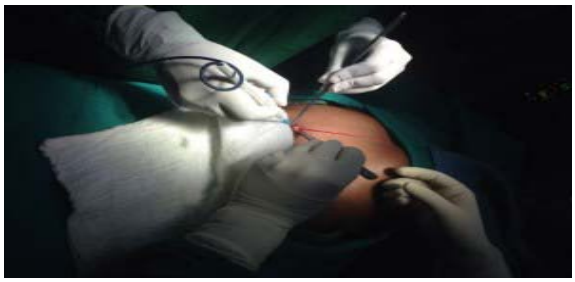
Case report

A 42 year old male presented with complaints of vague abdominal pain for 1 month which was insidious in onset , not progressive ,no radiation or aggravation. The patient reported no headaches, palpitations, or diaphoresis. The results of a clinical examination were unremarkable. Given his abdominal discomfort , the patient was referred for an abdominal ultrasound evaluation, which revealed 6*7 cm mixed echogenic mass in the area of the left kidney. Further imaging with computed tomography scanning suggested that the mass arose from the left adrenal gland of size 8*7*7cm with increased vascularity and displacing the left kidney postero inferiorly.

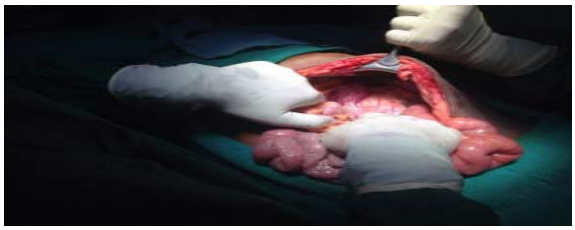


Pheochromocytoma

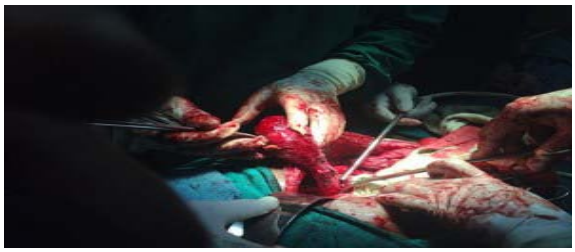
The patient was referred for further investigation. Total fractionated urine metanephrines were measured in a sample of an acidified 24-h urine collection. Urine analysis results were concordant, with increase in total metanephrines with value of 2385units/day (normal value 25-312). This all routine blood investigations and urine tests were normal. Medical endocrinologist opinion was obtained for pre operative preparations. The patient was started on prazosin 1mg tds, monitoring of blood pressure done both in supine and upright procedure, high salt diet of 3-5 g per day, normal saline infusion of 1-2 litres per day. Before posting the patient to surgery, anaesthetists were informed about the case who were prepared for pre op induction and intra operative management. Surgery was proceeded with a upper midline laparotomy incision , retroperitoneum entered and the highly vascular tumor was identified ,meticulously dissected out from kidney and excised in toto. Intra operative and post operative period was uneventful. Histopathology reported as cells with fine basophilic cytoplasm. Zellballen pattern of cells with intracytoplasmic hyaline globules. No capsular and vascular invasion. No mitotic figures. Picture is suggestive of pheochromocytoma.



Incision



Entering retroperitoneum



Tumour resection

Uneventful postop period

Discussion

The World Health Organization defines pheochromocytoma as a tumour from catecholamineproducing chromaffin cells in adrenal medulla—an intra-adrenal paraganglioma [1,2]. Usually, found in adrenal medulla but they may arise anywhere in the body [1]. Patient's symptoms reflect excessive secretion of catecholamines into circulation [3]. Clinical presentation varies from triad of episodic headache, diaphoresis and palpitations to a variety of non-specific symptomatology. Most frequent symptom of Pheochromocytoma is hypertension. Prolonged exposure of the circulation to high norepinephrine concentration results in constriction, of both arteriolar and venous circulation, with a decrease in circulating volume [4,5]. The best confirmatory test is to measure free catecholamines and their metabolites in a 24 hrs urine collection [6,4]. MRI and CT both provide accurate and consistent identification of Pheochromocytomas [7]. Tumours in unusual sites and metastases may have to be identified with metaiodobenzguinidine (MIBG) scintigraphy [5]. A clinical evaluation of the cardiac status of the patient is to be done. A baseline and serial monitoring of full blood count and haematocrit provides an assessment of the adequacy of volume expansion when -blockade has been started [8]. Blood glucose level measurement is needed as hyperglycaemia is common. ECG: ST and T changes secondary to myocardial ischemia, ventricular hypertrophy, arrhythmias. Chest X-ray may reveal cardiomegaly or pulmonary oedema [4]. 2D ECHO: to estimate myocardial function if cardiomyopathy is suspected. Renal function can be assessed. Prior to surgery, it is necessary to control arterial pressure, heart rate and arrhythmias and to restore the blood volume to normal [9, 8]. This is conventionally done with -blockade over a period of 10-14 days and subsequently, additional -blockade is required to treat any tachyarrhythmias [4]. Prazosin (2-5 mg twice or thrice a day), selective 1-blocker, is favoured because of its shorter half life and ease of titration to the desired end point. -blockade is not instituted until -blockade is established. A non selective blocker should not be instituted before blockade is achieved as blockade of 2 vasodilatory receptors leads to unopposed stimulation and worsening of hypertension.

-blockers with additional -blocking properties are synergistic with -blockers in reducing B.P. eg. labetalol and carvedilol Calcium channel antagonists can be used for preoperative control of B.P. and reduce preoperative and intraoperative coronary spasm. Preoperative sedation and anxiolysis and assurance will decrease anxiety and prevent marked haemodynamic fluctuation in immediate preoperative period [4]. Communication between operating and anaesthesia teams is crucial for success of intraoperative management of patients undergoing resection. Routine monitoring along with CVP monitoring. Sodium Nitroprusside (SNP), Phentolamine, Prazosin, Nitroglycerine (NTG) and various other agents like Magnesium Sulphate ,Nicardipine, Diltiazem, Esmolol are used to control rises in B.P. [4,12,5,8,13,14].

Lidocaine is useful for ventricular arrhythmias while Amiodarone is an alternative. Volume loading before tumour ligation and fluid boluses should be tried before choosing adrenaline, noradrenaline and phenylephrine for Catecholamine withdrawal following venous ligation. Patient is usually kept in ICU unit for first 24 hrs and monitored for haemodynamic instability [15,5,16,17]. Epidural analgesia provides good post-operative pain relief and may be supplemented by oral medications. Approximately 50% patients remain hypertensive for few days, likely related to elevated catecholamine levels which persist for one week after resection [11].

Conclusion

Successful operative treatment of pheochromocytoma is dependent on close communication between the surgeon and anesthesiologist. Manipulation of the tumor needs to be minimized, and the anesthetic team must be prepared to manage the intra operative emergency. Surgery is curative in greater than 90% of pheochromocytoma cases. Although these tumors are highly vascular and tend to adhere to adjacent structures the great majority of them can be removed successfully via a laparoscopic approach. Advances in surgical technique have resulted in reduced operative complication rates.

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