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Case Report- Giant Non Functional Pheochromocytoma

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Abstract:
Adrenal pheochromocytomas(PCCs) are rare neuroendocrine tumours that arise from neural crest cells of adrenal medulla, however their prevalence is probably underestimated - in some series 50% were diagnosed at autopsy. Their clinical presentations are varies among patients, that is why diagnosis might be difficult to establish.a small proportion of tumors hardly synthesize or release any catecholamines and may have no symptoms and are termed as non functional or subclinical PCCs.the nonfunctional tumors are commonly picked up as incidentalomas and biochemical tests are usually negative. Undiagnosed subclinical normotensive PCCs could lead to catastrophic consequences during surgery and subsequently.

Keywords:*nonfunctional pheochromocytoma ,incidentaloma.*

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I. Introduction :

Pheochromocytoma(PCC)(greek phaios"dusky",chroma"color" , cytoma"tumor") is a neuroendocrine tumor arising from the chromaffin cells of the adrenal medulla.it is a sympathetic tumor with diverse manifestations due to variations in the catecholamine secretion and their metabolites, unlike the head and neck paraganglioma which are parasympathetic in origin and do not usually produce catecholamines.it may present with the classical triad of symptoms are palpitations , headache , and diaphoresis.about 14-55% of incidental lesions are asymptomatic which can be challenging .here we review a case of pheochromocytoma which presented as incidentaloma,completely asymptomatic and histology revealed the presence of large PCC.

II. Case Report:

55-year old man was admitted for bilateral inguinal hernia. During his admission his blood pressure was completely normal. And he had no symptoms apart from those related to hernia. Ultrasound was done for hernia and incidental renal mass was found.further evaluation on Ct scan shows approx. 10.5*6.8*9cm(AP*TR*CC) size well defined heterogenous density (predominantly hypodense)lesion in right suprarenal region showing peripheral intensely enhacing with internal non enhacing areas onarterial phase with its extension.medially it abuts inferior vena cava , anteriorly it abuts portal vein ,posteriorly it reaches upto posterior adbdominal wall ,superio-laterally it abuts right lobe of liver ,inferiorly it abuts upper pole of right kidney with preserved fat plane.

biochemical test in the form of urinary metanephrines were elevated.Ihc markers like synaptophysin and chromogranin were positive.subsequently he underwent open right adrenalectomy with intra operative bp fluctuations that were controlled with anti-hypertensive agents and post-operative recovery was uneventful.histology shows nested and solid pattern without capsular invasion and vascular invasion.

CLINICAL MANIFESTATIONS :

hypertension is the most common symptom it can be paroxysmal or persistent. Patients often also present with the classical triad of symptoms which are palpitations , headache , and diaphoresis (bouts of sweating). anxiety and tremors may also found. Metabolic derangements such as lactic acidosis, weight loss, and

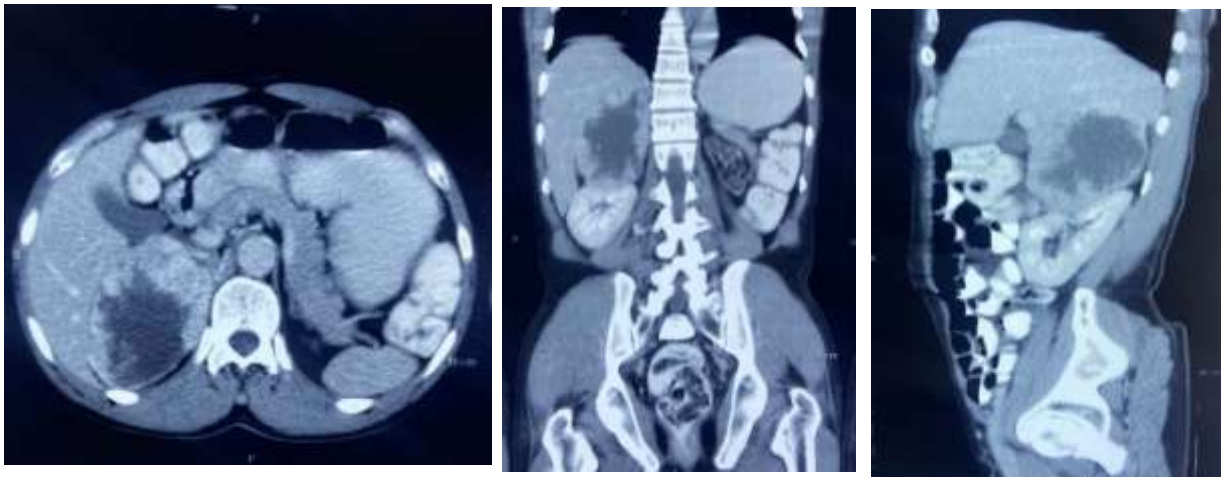
hyperglycemia are found in one in three patients secondary to catecholamine excess. Without treatment, patients face an increased rate of cardiovascular morbidity due to catecholamine-induced malignant hypertension, stroke, heart failure, and fatal arrhythmias

III. Discussion:

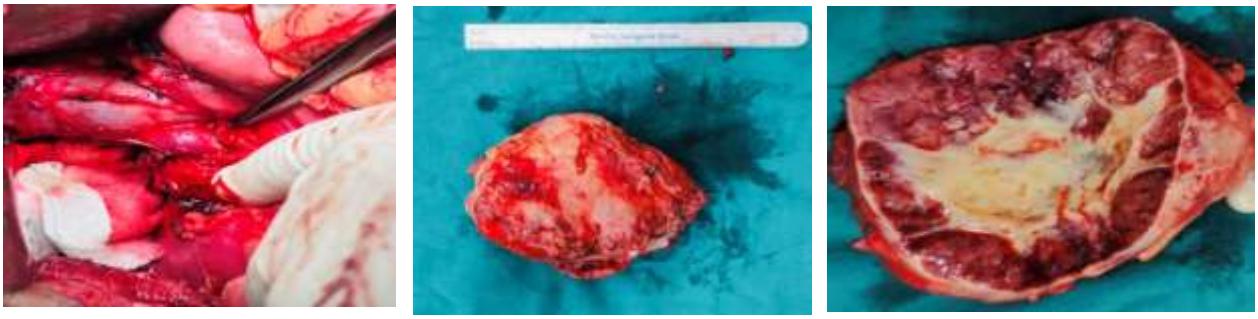
This report describes the case of a 55 year old male with an incidental nonfunctioning pheochromocytoma. pheochromocytoma is a rare neuroendocrine neoplasm of chromaffin cells, occurring in approximately 0.95 per 100,000 persons.more recent data indicate that 1.5% to 18% of adrenal incidentalomas discovered during abdominal imaging for various reasons. Though 80-85% neoplasms have been described within the adrenal medulla, 15-20% are found in the sympathetic nervous system ganglia and are referred to as paragangliomas(PPGs). While PPGs in the abdomen secrete high levels of catecholamines, most paragangliomas found in the head and neck are nonfunctional.

Pheochromocytomas have been described in all ages. Peak incidence occurs in the fourth and fifth decades in sporadic cases and earlier in associated hereditary conditions.

Recent advances in genome sequencing have demonstrated that approximately 35% of these chromaffin cell neoplasms are hereditary.inherited PCCs are associated with conditions such as multiple endocrine neoplasia type ii (MEN-2A OR MEN 2B), neurofibroma type i(NF-1),von hipplelindau syndrome and familial paraganglioma due to genes encoding succinate dehydrogenase(SDH) subunits b,c,d .



[Figure 1,2,3ct scan showing a tumor 10.5 cm in diameter in upper pole of right kidney]



(Figure:1showsing abutment of tumor with inferior vena cava ; figure:2 showing gross specimen adrenal mass; figure:3 showing cut section of adrenal mass)

The differential diagnosis of adrenal incidentaloma in patients without a history of malignant disease is following.

Nonfunctioning adenoma	60%
pheochromocytoma	10%
myelolipoma	9%
Adrenocortical carcinoma	5%
Adrenal cyst	5%
ganglioneuroma	5%

Cortisol producing adenoma	5%
aldosteroloma	1%

The clinical andbiohumoral presentation of PCCs depends on the size and capacity of tumor to synthetize,metabolize and release of catecholamines and their metabolites.

Recent study has shown that PCCs that are normotensive show a downregulation of five genes phenylethanolamine-N-_methayltransferase(PNMT) ,secretogranin II , vesicular monoamine transporter type I, norepinephrine transporter and Np-Y which are involved in key processes of catecholamine metabolism in comparasion to those with hypertensive PCCs.this leads to small amounts of catecholamines that are secreted and thereby present with minimal clinical symptoms.

Other factors that are associated with non functional tumors besides the amount of catecholamines are nature of circulating catecholamines and size of the tumors.there are two types of phenotypes in PCCs-noradrenergic and adrenergic which depends on the activity of PNMT ,the enzyme that converts noradrenaline to adrenaline tumor that lack PNMT activity do not produce adrenaline and exhibit a noradrenergic phenotype.both the phenotypes are found in both sporadic and tumors associated with genetic mutations of identified 10 susceptibility genes namely:VHL,RET,NFI,SDHA.SDHB,SDHC,SDHD,SDHAF2,TMEM127 AND MAX.The adrenergic phenotype is seen in tumor associated with mutations of RET,NF1 AND TMEM127 , whereas noradrenergic phenotype is seen in VHL mutations.

Though 20-30% of pheochromocytoma cases are detected incidentally in asymptomatic patients, approximately 1 in 5000 patients evaluated for hypertension are found to have a pheochromocytoma. However, autopsy studies reveal a prevalence of 0.05%, indicating many tumors are undiagnosed and may contribute to premature mortality.

TREATMENT:

Definitive management of pheochromocytomas includes complete adrenalectomy or partial excision in patients with bilateral or multifocal disease.prior to surgery,patients must be treated with an alpha-blocker to prevent life threatening complications during the surgery.however some study showed no benefit in maintaining intraoperative hemodynamic instability.patient can sustain severe hypertensive crisis during surgery due to tumor manipulation and significant post operative hypotension following surgery.

Laparoscopic or robotic partial adrenalectomy are preferred compared to open surgery due to decreased morbidity.prognosis after surgical removal is excellent ,although hypertension persists in 50% of patients.patients should be followed every year for 10 years after surgery given a recurrence rate of 17%.

OUTCOME OF THE PATIENT :

The patient underwent a open adrenalectomy.post operatively the patient remained hypotensive and required pressure support for 24 hours.he subsequently recoverd and was discharged home with annual endocrine follow up.

Disclosures:
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Conflict of interest:none

References

[1]. rajeevparmeswaran,han boon oh,suannlui,kong b tan.clinical challenges in nonfunctional pheochromocytoma.2019 december11(3):86-90.world journal of endocrine surgery.doi10.5005/jp-journals-10002-1264.
[2]. luigipetramala,matteogalassi,GiuseppeD'ermo,Federicaolmati.nonfunctioning adrenal pheochromocytoma incidentally discovered associated with renal oncocytoma.jonco-nephrol 2017;1(1):62-65.doi:10.5301/jo-n.5000003.
[3]. neha Agarwal,MD.,ganxon ng,MD.,pheochromocytoma :a case report and review of clinical presentation,diagnosis,andmanagement.proceedings of ucla healthcare-vol 20(2016)
[4]. Sundahl N, Van Slycke S, Brusselaers N. A rare case of clinically and biochemically silent giant right pheochromocytoma: case report and review of literature. ActaChir Belg. 2016; 116(4):239-242.
[5]. Moch H, Cubilla AL, Humphrey PA, Reuter VE, Ulbright TM. The 2016 WHO Classification of Tumours of the Urinary System and Male Genital Organs-Part A: Renal, Penile, and Testicular Tumours. Eur Urol. 2016;70(1):93-105.
[6]. Korpershoek E, Koffy D, Eussen BH, et al. Complex MAX Rearrangement in a Family With Malignant Pheochromocytoma, Renal Oncocytoma, and Erythrocytosis. J ClinEndocrinolMetab. 2016;101(2):453-460
[7]. Shuch B, Ricketts CJ, Metwalli AR, Pacak K, Linehan WM. The genetic basis of pheochromocytoma and paraganglioma: implications for management. Urology. 2014 Jun;83(6):1225-32. doi: 10.1016/j.urology.2014.01.007. Epub 2014 Mar 15. Review. PubMed PMID: 24642075; PubMed Central PMCID: PMC4572836.
[8]. Comino-Méndez I, Gracia-Aznárez FJ, Schiavi F, et al. Exome sequencing identifies MAX mutations as a cause of hereditary pheochromocytoma. Nat Genet. 2011;43(7):663-667.
[9]. Conzo G, Musella M, Corcione F, De Palma M, Ferraro F, Palazzo A, Napolitano S, Milone M, Pasquali D, Sinisi AA, Colantuoni V, Santini L. Laparoscopic adrenalectomy, a safe procedure for pheochromocytoma. A retrospective review of clinical series. Int J Surg. 2013;11(2):152-6. doi: 10.1016/j.ijsu.2012.12.007. Epub 2012 Dec 23. PubMed PMID: 23267853.

[10]. Eisenhofer G, Peitzsch M. Laboratory evaluation of pheochromocytoma and paraganglioma. Clin Chem. 2014 Dec;60(12):1486-99. doi: 10.1373/clinchem.2014.224832. Epub 2014 Oct 20. Review. PubMed PMID: 25332315.
[11]. van der Zee PA, de Boer A. Pheochromocytoma: a review on preoperative treatment with phenoxybenzamine or doxazosin. Neth J Med. 2014 May;72(4):190-201. Review. PubMed PMID: 24829175.
[12]. Asher KP, Gupta GN, Boris RS, Pinto PA, Linehan WM, Bratslavsky G. Robot-assisted laparoscopic partial adrenalectomy for pheochromocytoma: the National Cancer Institute technique. Eur Urol. 2011 Jul;60(1):118- 24. doi:10.1016/j.eururo.2011.03.046. Epub 2011 Apr 9. PubMed PMID: 21507561; PubMed Central PMCID: PMC3109214.
[13]. Boris RS, Gupta G, Linehan WM, Pinto PA, Bratslavsky G. Robot-assisted laparoscopic partial adrenalectomy: initial experience. Urology. 2011 Apr;77(4):775-80. doi: 10.1016/j.urology.2010.07.501. Epub 2010 Dec 3. PubMed PMID: 21122898; PubMed Central PMCID: PMC3074102.

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