The Gulf Journal of Oncology

Indexed By PubMed and Medline Database

Issue 18, May 2015
ISSN No. 2078-2101

Cooperation between the Gulf Centre for Cancer Control and the Gulf Federation for Cancer Control
GCC Annual Cancer Awareness Week (Feb 1-7)

The Official Journal of the Gulf Federation For Cancer Control
Table of Contents

**Case Reports /Review Articles**

**An Unusual Breast Malignancy** .....................................................................................................................................................................07  

**Nuchal Fibroma: A rare entity of neck masses** .............................................................................................................................................10  
N. Alsaleh, H. Amanguno

**Epinephrine–secreting large incidental pheochromocytoma in a normotensive male with stormy intraoperative hemodynamics** ......13  
O. Nazir, T. Sharma, M. Maqsood, A. Khatuja, R. Misra

**Everolimus induced Pneumonitis** ..................................................................................................................................................................18  
Q. Badar, N. Masood, A. N. Abbasi

**Primary Mantle Cell Lymphoma of Appendix** ..................................................................................................................................................25  
VL Gaopande, SD Deshmukh, VC Shinde

**A Rare Variant of Multiple Myeloma: Non–Secretory Myeloma with diffuse Osteolytic Lesions** ...........................................................28  
S. Sultan, S.M. Irfan

**Pain and Cancer: a systematic review** .........................................................................................................................................................32  

**Original Articles**

**Tumor Thickness: A predictor of nodal disease in early squamous cell carcinomas of buccal mucosa** ..................................................38  
G. Deshpande, S. Das

**Hypofractionated Simultaneous Integrated Boost (SiB) versus Conventional Fractionation in Localized Prostate Cancer: A Randomized Pilot Study** ..........................................................................................................................................................44  

**Quantitative evaluation of the dosimetric effects of balloon deformation and source position in high–dose rate mammosite breast brachytherapy** ..............................................................................................................................................................................54  
I. Ali, S. Negusse, S. Ahmad, O. Algan

**Spectrum of Ovarian Tumors: Histopathological study of 218 cases** .........................................................................................................64  
N.A. Mansoor, H.S. Jezan

**Rare Chromosome Structural Aberration Characterizing Oncology Malignancy** ..................................................................................71  
A. Movafagh, A. Sayad, M. Hashemi, H. Darvish, D. Zare–Abdollahi, B. Emamalizadeh, F. Shahvaisizadeh, N. Mansouri, S. A. Mortazavi–Tabatabaei

**Infectious complications after allogeneic bone marrow transplantation: Sheikh Badryia Center, Kuwait** ............................................79  
S. AlShemmari, S. Refaat, A. A. Abdullah, M.A. Abul

**Cancer News and Scientific Events in the Arab Region**

- **News Notes** ..............................................................................................................................................................................................87
- **Advertisements** ........................................................................................................................................................................................90
- **Scientific events in the GCC and the Arab World for 2015** ..................................................................................................................91
Case Report

**Epinephrine-secreting large incidental pheochromocytoma in a normotensive male with stormy intraoperative hemodynamics**

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**Abstract**

Pheochromocytoma is a rare tumour which is usually suggested by sustained or paroxysmal hypertension however the spectrum of the presentation of pheochromocytoma continues to expand and hypertension may be absent despite excess catecholamine secretion. The normotensive pheochromocytoma is a distinct entity and as in the case we report the presentation was quite unique as well as the intraoperative behaviour was stormy.

**Keywords**

Incidentaloma, Pheochromocytoma, epinephrine

**Introduction**

Pheochromocytoma is a rare catecholamine secreting tumors arising commonly from adrenal medulla and to some extent from other paraganglia of the sympathetic chain with an incidence of 1.55–2.1 per million populations per year(1). Although hypertension, sustained or paroxysmal, is usually a cardinal feature of pheochromocytoma, normotensive presentation is unusual(2). Pheochromocytoma whether it is secreting or not does not change anesthetic care as it may become intraoperatively secreting; even clinically silent pheochromocytoma can be lethal (3).

**Case report**

A 52-year-old previously healthy male presented to our hospital as a case of hepatic mass diagnosed on routine medical checkup and USG. He was having chief complaints of dragging sensation in right upper abdomen from past 3 months and history of weight loss noticed by his family members from past 2 months. There was no history of hypertension, palpitations, chronic illness, hirsutism, polyuria, nocturia, loss of appetite, jaundice, melena, hematemesis, except occasional sweating on exertion.

Vitals recorded were blood pressure:—124/86mmHg, pulse rate:—76/min regular, respiratory rate:—14/min. His body weight:—73kg, height:—1.64mt and BMI:—27.1kg/m². Systemic examination was inconclusive except mass noted in right upper abdomen on palpation.

Laboratory data showed blood sugar (R):—124mg/dl, Hb:—13.2g/dl, platelet count = 4lakh/, Haematocrit = 42.7%, TLC = 8000/microl, total bilirubin = 0.5mg/dl, AST= 22.6iu/l, ALT = 19.3iu/l, Alkaline phosphatase = 110 iu/l, serum.Na+:—137meq/l, Serum.K+:—4.7meq/l, urea:—13.2mg/dl, creatinine:—0.9mg/dl, coagulation profile: Apltt= 30.2sec, prothrombin time = 10.7sec, INR = 1.05, thyroid profile TSH= 3.57

CT scan abdomen (Figure 1) showed a large 18cm (ap) x 15cm (trans) size solid cystic mass with heterogeneous enhancement lying in posterior hepato—renal plane displacing right kidney inferiorly and ivc rt. renal vein and portal vein anteriorly and

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superiorly reaching up to diaphragm. Right adrenal gland not visualized. Left adrenal is normal. CT findings were suggestive of right adrenal neoplasm of pheochromocytoma.

Figure 1: CT abdomen showing 186mm × 156mm solid mass in right posterolateral hepato-renal region.

24–hr urine catecholamines showed VMA = 26.48 mg/g creat (1.60 – 4.20), epinephrine = 233.38 mcg/g creat (1.30–10.70), norepinephrine = 227.03 mcg/g creat (8.90 – 61.6), dopamine = 471.09 mcg/g creat (40.00 – 390).

With all these findings, the patient was diagnosed as a case of right adrenal pheochromocytoma and planned for right adrenalectomy.

Preoperative workup involved ECG – normal sinus rhythm, echocardiography – normal study with LVEF 55%. Consultation from endocrinologist for adequate preoperative blockade of catecholamine effect involved to tab. prazosin for 14 days followed by tab. propranolol (after confirming adequate α blockade). Arrangement for necessary blood and blood products were done since the tumor was large.

Prior to induction under light sedation epidural catheter at L1, L2 (for intra and post op analgesia) and arterial catheter were placed in radial artery.

Intraoperative course: Induction of anesthesia was done with fentanyl 2mcg/kg, propofol 2mg/kg, vecuronium 0.1mg/kg and then central venous catheter was placed in right internal jugular vein. Anesthesia was maintained with oxygen, nitrous oxide and isoflurane. Intraoperatively massive hemodynamic fluctuations were noted while the tumor was being handled and were out of proportion to their preoperative stable values (Figure 2).

Episodes of hypertension were managed with infusion of sodium nitroprusside and labetalol boluses while intraoperative hypotension was managed with noradrenaline infusion and phenylephrine boluses.

Figure 2: Wide variations in intraoperative systolic blood pressure (SBP) and diastolic blood pressure (DBP) (x-axis shows time in minutes and y axis BP in mm Hg).

Figure 3: Gross specimen of the pheochromocytoma which measured 25× 15× 8 cm in dimensions.

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Total intraoperative blood loss was about 4000ml. Fluid replacements involved crystalloids 9000ml, colloids 1500ml, packed RBC 10 units, fresh frozen plasma 4 units. Intraoperative serial arterial blood gas monitoring was done and necessary corrections were done accordingly. Postoperatively patient was shifted to ICU for elective postoperative ventilation with noradrenaline 0.1mcg/kg/min. Patient was progressively weaned off from inotropic support in next 24 hours and was extubated next day. He was discharged one week after surgery.

**Discussion**

Pheochromocytoma are rare neuroendocrine tumors that produce, metabolize, and usually secrete catecholamines. Although hypertension is a common presenting feature of pheochromocytoma, the tumors occur (or are present) in only 0.1% of patients with hypertension. The variability of symptoms and rarity of occurrence render these tumors difficult to diagnose; many are discovered incidentally during radiological examination or at autopsy (4, 5). The greatest frequency occurs in the fourth and fifth decades of life, with slightly higher female preponderance (60%). Pheochromocytoma release large amounts of catecholamines (cca) (adrenaline, noradrenaline and dopamine), and various peptides and ectopic hormones (enkephalins, somatostatin, calcitonin, oxytocin, vasopressin, insulin and adrenocorticotropic hormones) (6). A diagnosis is established by measuring the levels of metanephrines in the urine or blood (7). Numerous anesthetic techniques have been used from time to time but among these the use of a combined epidural and general anesthesia is considered to be the most suitable and the preferred technique (8, 9). The regional anesthetic technique though suppresses the stress response of surgical incision but cannot control the quantum of catecholamines released during the tumor handling (10).

In our case there were no peculiar signs and symptoms of pheochromocytoma despite increased catecholamines (increased adrenaline, noradrenaline and dopamine levels, with predominant epinephrine secretion) and it was diagnosed incidentally on CT scan. It is an unusual presentation pheochromocytoma. Hinyokika Kiyo reported a case of 66-year-old female who had no symptoms and no episodes of

*Figure 4. Photomicrograph of the histopathology specimen showing cells arranged in nesting pattern, high power magnification shows abundant cytoplasm with fine granular nucleus.*
hypertension but abdominal CT scan showed a large mass, 9x8x6cm, in the right adrenal region and plasma and urinary catecholamine showed normal adrenaline and noradrenaline but elevated dopamine levels (11). Awada et al described a normotensive 26-year-old woman with a large pheochromocytoma that secreted multiple catecholamines, including dopamine. Some pheochromocytomas secrete dopamine in addition to or in the absence of other catecholamines. Patients with these tumors are frequently normotensive (12). Sukoret et al reported an epinephrine—secreting pheochromocytoma in a normotensive woman with adrenal incidentaloma that was discovered incidentally when she presented with abdominal pain and a normal blood pressure (13). Awoke et al reported a 41-year-old woman with pheochromocytoma in atrial wall with no symptoms of hypertension but had increased adrenaline, noradrenaline and dopamine levels (14). Smircić et al reported a patient with a pheochromocytoma that was unusual for two reasons: she was normotensive during 3 weeks of close observation in the hospital and the urinary adrenaline level was higher than the noradrenaline level (15).

Despite no peculiar symptoms of hypertension, palpitations and despite adequate α blockade in preoperative period the intraoperative haemodynamics course of our case, was stormy. Agarwal et al reported an audit during the period 1990–2003 of a total of 45 patients of pheochromocytoma out of which nine patients were diagnosed as having subclinical or normotensive. Seven have elevated urinary metanephrine levels, and one patient had normal values. After selective α—adrenergic blockade (prazosin), surgery was performed. Six patients required infusion of sodium nitroprusside intraoperatively (16).

Hypertension that accompanies pheochromocytoma was generally been ascribed to the excessive circulating catecholamines released from the tumor. Norepinephrine principally has (alpha)—excitatory effects and hypertensive manifestations which parallel its circulating level. Epinephrine has both (alpha)— and (beta)—excitatory effects. Its action at the level of the (alpha)—2 presynaptic receptors is perhaps responsible for the occasional functional dysautonomia with postural hypotension and collapse. Another explanation for normotension in pheochromocytoma is the fact that apart from catecholamine, these tumors secrete more than 10 other substances: calcitonin, somatostatin, neurotensin, adrenocorticotropin, (beta)—endorphin, lipotropin, metenkephalin, dopamine, neuropeptide and occasionally, vasoactive intestinal peptide (VIP). Calcitonin gene—related peptide and VIP are potent vasodilators and these may contribute to the normal blood pressure (17,18,19).

Pheochromocytoma is a rare tumor which is usually suggested by sustained or paroxysmal hypertension however the spectrum of the presentation of pheochromocytoma continues to expand and hypertension may be absent despite excess catecholamine secretion and normotensive pheochromocytomas are a distinct entity, and all adrenal incidentalomas should alert anesthesiologist of intraoperative hemodynamic fluctuations despite benign preoperative hemodynamics. Uniqueness of our case lies in the fact that:

1. It presented as upper abdominal mass and hepatic adenoma on routine USG and it was discovered as a huge adrenal mass on CT scan.
2. It showed increased catecholamines (with adrenaline levels higher than noradrenaline and dopamine) despite no history of palpitations and hypertension.
3. It showed huge fluctuations in haemodynamics intraoperatively despite adequate α—blockade and its dormant nature preoperatively.

**Conclusion**

Normotensive patients with large adrenal masses should alert the anesthetist of its stormy intraoperative course and hence adequate preparations should be made in advance.

**Abbreviations**

CT: Computed tomography; BMI: Body mass index; USG: Ultrasonography; VMA: Vanil mandilic acid (VMA); ECG: Electrocardiography
References