Case Report

An unusual case of paraganglioma

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ABSTRACT

Pheochromocytomas or Paragangliomas are neroendocrine tumors elaborating catecholamines. They may not present always in a classic manner and a high degree of suspicion is important in diagnosing them early. They can grow large enough to impinge on the great vessels and yet symptoms may not indicate the severity until late. A young African 32 year old Male presented with occasional headaches, moderate weight loss, abdominal pain and hypertension for three years. On investigation for hypertension a large paraganglioma was discovered wrapped around the Aorta and vena cava. He was operated successfully after preoperative preparation and was discharged normotensive. **Conclusion:** Young hypertensive should always be worked up fully including checking for abdominal masses. The classic paroxysm was absent in this case. After surgical removal the catecholamines elaborated by the tumor also take time to be washed out of the circulation thus the blood pressure will need careful monitoring. Conventional alpha blockage first and beta blockage later was the algorithm followed but choice of agent was also discussed with anesthethesiologist. Though classically taught phenoxy benzamine is used, in this case it was not used and a combination of Prazosin and Propranolol was used successfully.

Key words: Hypertension, paraganglioma, pheochromocytoma

Introduction

A 32 year old African male presented with history of Weight loss, pain in the abdomen and episodes of sweating and palpitations. There was no relief from sugar intake. He had hypertension which was being treated by multiple drugs over a course of three years. As a consequence of the episodes of the aforesaid symptoms he had been forced to take extended leave of absence and as a result lost his job as an accounting clerk. He was evaluated in our Hospital as a case of young hypertension and found to have a large palpable mass in the epigastrium that Magnetic resonance imaging (MRI) showed as a large tumor adherent to the great vessels. Biochemical tests supported a diagnosis of a paraganglioma/neurendocrine tumor. Patient was prepared for surgery and the operation lasted 9hrs. Postoperatively he was still hypertensive but the anti hypertensives were tapered over 10 days. He recovered completely and eight months later on follow up had regained some of his weight, was off anti hypertensives and had also been successfully reemployed.

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Case Report

A 32 year old African male an accounting clerk by profession presented with history of Weight loss of around 6 kilograms over a period of two years and episodes of Palpitations accompanied by Headaches. He also had episodes of perspiration and sometimes Giddiness. On several occasions he had been admitted in a local hospital with the above Symptoms. He however had no loss of vision at any time and there was no relief by the intake of sugar. A few times there was pain in the abdomen during such episodes which though he says was severe but short lasting. There were no bowel disturbances. He was taking multiple anti hypertensives and also Tab Morphine for pain during these episodes.

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Clinical examination

On initial presentation the patient was a thinly built, anxious looking individual and his palms were moist to touch. His thyroid was not enlarged and there were no unusual skin findings. The blood pressure recorded both in the ward and by us averaged 150/96mmHg. He had tachycardia to the tune of around 100/mins. Other than that the lungs, Central and peripheral nervous system examinations were normal. On examining his abdomen a vague, ill defined, mildly tender mass was felt in right hypochondrium and extending up to the epigastric region. Based on the symptomatology and general physical examination of this patient; a battery of investigations were ordered for establishing the cause of the hypertension. Blood sugars Serum electrolytes, Calcium, Phosphorous etc., all were normal. The Liver, Kidney parameters were also normal. The Renal Artery Doppler was normal. The ECG showed a Left Ventricular Hypertrophy pattern and the Echocardiogram revealed a concentric Left Ventricular Hypertrophy consistent with long standing hypertension. A 24hrs Urinary Metanephrines and normetanephrines study was ordered and an MRI abdomen scheduled.

MRI scans

A large mass found indenting the great vessels. Figure 1 and Figure 2.

Clinically correlating with the symptomatology a catecholamine producing lesion was suspected and the Urinary Normetanephrines-55217 nmol/L (normal 72-505) were grossly elevated. The Urinary Metanephrines 698 nmol/L (normal 9.3-122) too were very high. To finally confirm that the mass in abdomen was indeed responsible, an meta iodo benzyl guanidine (MIBG) scan was done Figure 3.

The MIBG scan uses meta-iodobenzylguanidine[1] scintigraphy that has a specificity of almost 100%in localising a pheochromocytoma. As shown above the tumor is clearly visualized.

Pre and post-operative course

Alpha receptor blockade initiated first with Prazosin (minipres) a short acting alpha-one blocker. The Patient was given four pints of normal saline per day to prevent volume depletion and minimise the postural hypotension that usually accompanying alpha blockade. An Effective alpha blockade is signalled by >20mm fall in BP on upright posture. This was achieved. Beta blockade was then initiated with non-selective agent propranolol to control tachycardia due to catecholamines. His Blood pressure was maintained at approx. 110/78mm Hg. The Patient was operated successfully by the Oncosurgeon and intra operative pressure managed by the Anesthesiologist. Post operatively patient was monitored closely and propranolol restarted to cater for still circulating catecholamines. The Patient made a full and complete recovery and was eventually taken off anti hypertensives on discharge.

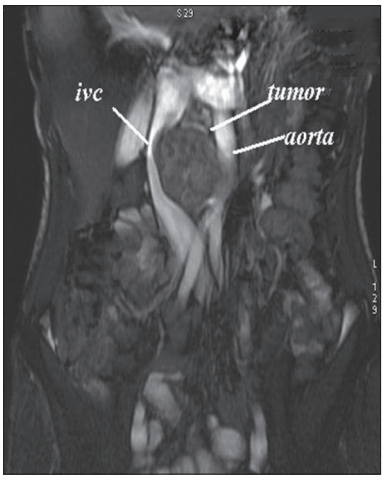

Figure 1: MRI scan abdomen coronal view

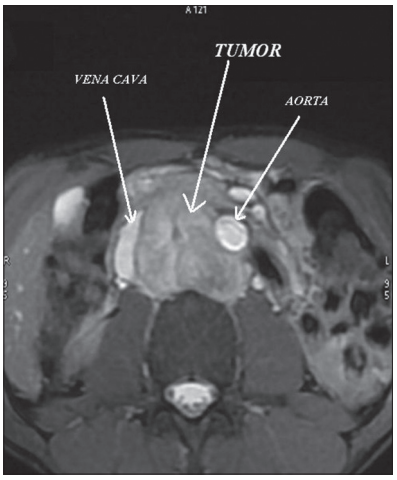

Figure 2: MRI scan transverse section abdomen

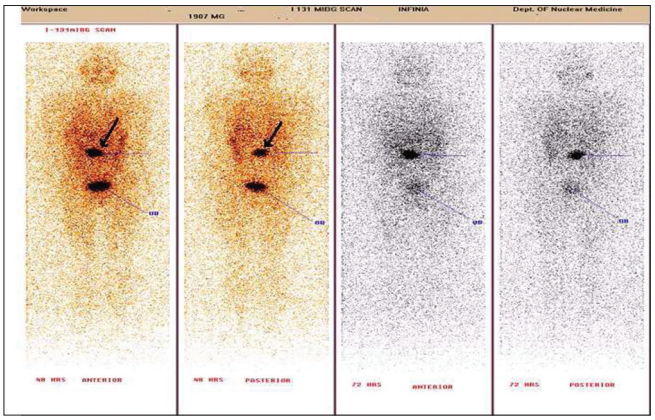

Figure 3: MIBG scan

Conclusion

A pheochromocytoma classically has the Five P's
Pressure (HTN) 90% of cases
Pain (Headache) 80% "
Perspiration 71% "
Palpitation 64% "
Pallor 42% ".

Paroxysms (the sixth P!) Of the above Pain (Headache), Perspiration, Palpitations are quite important and Lack of all three virtually excluded diagnosis. All young hypertensives must be evaluated fully keeping the above facts in mind. As far as the pre-operative preparation of a pheo is concerned; the anesthesiologist view point was that prazosin and propranolol was better than using phenoxybenzamine as phenoxybenzamine destroys the beta receptor permanently. In case the patient goes into shock the resuscitation becomes much more difficult if the inotropic receptors are compromised. Consent "Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal."

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