AORTOCAVAL EXTRA ADRENAL PHEOCHROMOCYTOMA.

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Manuscript Info

Abstract

We report a case of extra adrenal pheochromocytoma in a young adult male with hypertension having a tumour in inter aorto-caval region which was managed with excision after careful planning with adequate optimization and surgical expertise.

Key words: Catecholamines.

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Introduction:

Pheochromocytomas are catecholamine secreting neuroendocrine tumors that arise from chromaffin cells of sympathoadrenal system. Usually found in the adrenal gland, but also occur in extra adrenal sites. Most of the tumours secretes predominant noradrenaline than adrenaline. Though it’s a rare tumour accounting for 0.1% of hypertension it poses a great challenge for the management. Morbidity and mortality is quoted to be 50% in an unexpected emergency situation and less than 2% in a planned surgery. Management of aortocaval extra adrenal pheochromocytoma is highly challenging both in medical and surgical grounds. Medically because of high degree of hemodynamics changes associated during pre, intra and post operative periods. Surgically because of location of tumour near major vessels. Hence careful planning with adequate optimization and preoperative preparation, adequate intraoperative and postoperative management provides best successful result.

Case report:

25 year male admitted for complaints of pain abdomen and headache for past 1 month. He was diagnosed to have hypertension and was on antihypertensive drugs for past 6 months. No other significant history. General examination findings were normal. His BP- 160/110 mmhg. Abdomen soft, no mass palpable per abdomen. His basic blood investigations were normal. ECG showed features of LVH and Echo- conc. LVH, normal LV systolic function. Urinary VMA (vanillyl mandelic acid) found to be normal. Then plasma metanephrines was done which was elevated 147pg/ml (normal < 90 pg/ml). CECT abdomen showed enhancing retroperitoneal soft tissue lesion to rule out retroperitoneal sarcoma. MRI abdomen revealed 4 x 3 x 6 cm oval lesion noted between IVC and aorta, compressing the IVC and displacing aorta.
With above features patient was diagnosed to have functioning aortocaval paraganglioma (pheochromocytoma) and planned for surgical excision.

Patient was preoperatively optimized by following measures. he was started on t.prazosin 2mg bd and after 1 week was increased to 5mg bd until α blockade was attained. T. Propranolol 10 mg bd started later, T amlodipine 2.5 mg od was added. Pre op vitals were HR 74/min, BP – 130 / 80 mm hg, Spo2 – 100%, CVS / RS – NAD.

Intraoperatively: epidural with general anaesthesia was given with intraarterial continuous bp monitoring with cvp and peripheral lines. Duration: 4 hours, fluids: 3 litres of crystalloids, vital parameters: Hr: 48 – 110 / min, BP: 70 / 30 mm hg to 308 / 212 mm hg, Spo2: 99 to 100%, cvp: 10 to 18 cm h2o.

Drugs used intraoperatively: Sodium nitro prusside: 1 to 3 µg/ kg/min iv given through aluminium foil covered infusion in order to avoid light exposure, phenyl ephrine: 1 to 2 µ g/ kg / min iv, esmolol: 0.1 to 0.2 mg / kg / min iv, metoprolol: (1+1) mg iv bolus, norepinephrine: 0.1 to 0.2 µ g/ kg / min iv.
Procedure: midline vertical laparotomy incision made. Peritoneal cavity entered and bowels were pushed away from midline. Retroperitoneal cavity entered. Abdominal aorta, renal vessels and IVC exposed and clearly delineated from tumour mass. Left renal vein was found stretched over the lesion. Tumour of size 6x4 cm found at level of renal artery behind renal vessels between aorta and IVC.

Tumour dissected carefully from surrounding major vessels and structures and excised into without much handling, inorder to avoid systemic entry of large amount of catecholamines. Hemostasis secured.

Post operative: after completion of the procedure patient was shifted to ICU for elective mechanical ventilation with a supportive dose of phenylephrine infusion at a rate of 2 micro gm/kg/min. Later the dose of phenylephrine was tapered and weaned off. Patient was given epidural bupivacaine + fentanyl at the dose of 5ml/hr for pain relief. Patient was extubated the next day morning with normal vitals. T.prazosin was tapered gradually. Postoperative period was uneventful. His blood pressure normalized. Discharged 10th POD without any antihypertensives. HPE reported as paraganglioma – pheochromocytoma. Post operative urinary VMA and plasma metanephrines at 3 months were found to be normal. Patient on regular follow up without need of any antihypertensive drugs.
Conclusion:-
Management of aortocaval extra adrenal pheochromocytoma is highly challenging but with careful planning and adequate optimization and surgical expertise it can be successfully managed.

References:-