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PHAEOCHROMOCYTOMA-TUMOR OF ADRENAL MEDULLA

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ABSTRACT- It is a tumor of sympathetic nervous system and rare neuro endocrine tumors. 90% are present in adrenal medulla. 10% are extra adrenal called as PARAGANGLIOMAS. They are catecholamine secreting tumors and account for less than 0.1% cases of hypertension. Most of them are benign but about 15% show malignant transformation.

1. INTRODUCTION

They arise from chromaffin cells. Phaeo means Dusky and Chromo means Color. Right adrenal is involved more than left adrenal gland.

Adrenal tumors secrete more of epinephrine and Extra adrenal tumors secrete norepinephrine.

2. CLINICAL FEATURES

Hypertension; Headache; Palpitation; Perspiration; Pallor; Paroxysms of orthostatic hypotension; Anxiety; Angina(catecholamine induced); CHF; Catecholamine induced Dilated cardiomyopathy; cardiac arrhythmias;

Signs- Hypercalcemia; Glucose intolerance; Lipolysis

PHEO-RULE OF 10-

10% Extra adrenal

10% occur in children

10% familial

10% bilateral



10% recur

10% malignant

10% metastasize

10% non-secreting

10% discovered incidentally

10% present in head and neck

3. ASSOCIATION

MEN2a; MEN2b; Von-Hippel Lindau; Neurofibromatosis; Struge Weber Syndrome

4. DIAGNOSIS

Biochemical-24hrs urine catechols; 24hrs urine metanephrine; plasma catecholamines and metanephrine

Localization- CT abdomen; MRI; MIBG scan

24hrs urine collection-Creatinine; Catecholamines; Metanephrines; VanylMandellic acid (VMA); dopamine

MIBG scan-123 I/131 I labeled meta iodo benzyl guanidine is injected into a vein; MIBG is taken up by tumor cells; Scan taken at 24hrs; 48hrs; 72hrs

5. MANAGEMENT

Medical Therapy- Started 6weeks prior to surgery to allow restroration of normal plasma volume

Start with α -blocker Phenoxy benzamine(10-20 mg orally TID) If α -blocker produces tachycardia; β -blocker can be given orelse combined α + β blocker (labetalol) can be given.

B-blocker should not be given before α -blocker as it causes paradoxical rise in BP due to unopposed α -mediated vasoconstriction

Surgery- Intra op problems can occur at the time of handling and removal of tumor due to outpouring of catecholamines in blood.

Have at hand α and β -blocker loaded I.v; phentolamine 50 mg in 500 ml normal saline; sodium nitroprusside 50 mg in 250 ml of 5% dextrose.



6. CONCLUSION

Prognosis is good if treated early; 10% can recur.

Repeated follow up for every 5 years necessary.

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