Pheochromocytoma
- An experience from a single centre in South India

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Background

WHO (2004) define pheochromocytomas as an intra-adrenal paraganglioma, whereas closely related tumours of extra-adrenal sympathetic or parasympathetic paraganglia are classified as extra-adrenal paragangliomas. The prevalence of pheochromocytomas varies widely from 0.05% to 0.1% in patients with sustained hypertension. About 24% to 27% of patients are associated with known genetic mutations. Among the children, prevalence of mutation may be as high as 40%. Upto 20% of pheochromocytomas are extra-adrenal in nature, while 13-26% are malignant.

Study Design

Retrospective Data: 1994-2014 (21-years)

Outpatient and inpatient hospital records, discharge summaries, operation notes, laboratory reports and follow-up records.

203 patients with pheochromocytoma/paraganglioma

Patient profile: 1994-2014

1994-2006 (n=94) 2007-2014 (n=86)

Age (yrs) 35 (11- 71) 36 (18-72)

Male – n(%) 45 (48 %) 41 (48%) Female – n(%) 49 (52%) 45 (52%)

• Age of onset is earlier as compared to the Western literature (43.9 years)  
  • Male : female ratio = 1:1

Investigations profile: 1994 - 2014

INVESTIGATIONS POSITIVE

Urinary metanephrines/normetanephrines 119/136 (88 %)

Predominant metanephrines 24%

Predominant normetanephrines 12%

Urinary Vanillyl Mdaamic Acid (VMA) 32 / 44 (72%)

Predominant normetanephrine group

• Had significantly more normotensives than metanephrine group (42% vs 17%, p=0.001)

• Had more extra-adrenal tumour than the metanephrine group (59% vs 33%)

Spectrum of Genetic Mutation (N=50)

VHL 19% RET 19% SDHB 25% SDHD 37%

• 82% had germline mutations in any of the susceptibility genes

• VHL was more common in young age (< 22 yrs & b/l tumour)

• One of the malignant tumours had SDH-D positive and one had SDH-D mutation

Pre-operative antihypertensives

• 2-3 drugs – 80% of patients

• Phenoxybenzamine (27%)

• Phentolamine (26%)

• Phenoxybenzamine + phentolamine (13%)

Types of Treatment received

TREATMENT MODALITY DISTRIBUTION (180) N (%)

Surgery Completed successfully 176 (97%)

Recurrent initial operation elsewhere 26 (15%) 16 (8.8%)

Mortality 01 (0.5%)

MIBG therapy 01 (0.5%) 12 (7%)

Lutetium therapy Post-surgery 01

Follow-up

Median Duration (months) 52 months (06- 243 months)

Persistent hypertension 51(29%)

• 50% had reduction in antihypertensives post surgery

• Pre-operative duration of HTN longer persistent postoperative hypertension

Conclusion

• The median age of presentation of Pheo/PGL at diagnosis was 36 years (range: 16-72 years).

• Pheochromocytoma was Incidentally detected in 22- 24% patients.

• Malignancy was seen in 14% and was more common with larger tumours(>10 cm), extra adrenal tumours and in patients with higher normetanephrine levels.

• VHL was the most common genetic mutation, more common younger subjects and in patients with bilateral tumours.

References


