

## CASE REPORT

# PHAEOCHROMOCYTOMA - An Incidental Finding on Autopsy

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### Abstract

Phaeochromocytoma is an uncommon tumour of the adrenal medulla. This report describes an autopsy case in which a large suprarenal mass was found incidentally<sup>1</sup> which was then confirmed histologically as Phaeochromocytoma.

### Case

A 44 year old male who had gone fishing became suddenly unconscious and was declared as 'dead on arrival' in the ER. He had no significant past history. The autopsy was performed. There were no significant external findings on autopsy. Internal examination revealed congested and oedematous brain with intracerebral haemorrhages in the right hemisphere and left ventricle. The heart showed left ventricular hypertrophy. The right kidney was normal. The left kidney was enlarged with a suprarenal mass.

### Pathology

#### Macroscopy

The specimen received was a capsulated grey-brown to yellow piece of tissue measuring 10x 8 x 2cms. The cut surface was homogeneous yellow with areas of haemorrhages.

#### Microscopy

Microscopic examination revealed a capsulated tumour composed of large polygonal cells arranged in well defined nests separated by a vascular network. (Fig.1) The cells contained finely granular eosinophilic cytoplasm and oval nuclei. The tumour cells revealed minimal cellular and nuclear pleomorphism. Many areas of haemorrhage are noted in the tumour substance. (Fig. 2)

### Discussion

The term phaeochromocytoma was first coined by Ludwig Pick, a pathologist, in 1912, although Frankel, in 1886, made the first description of a

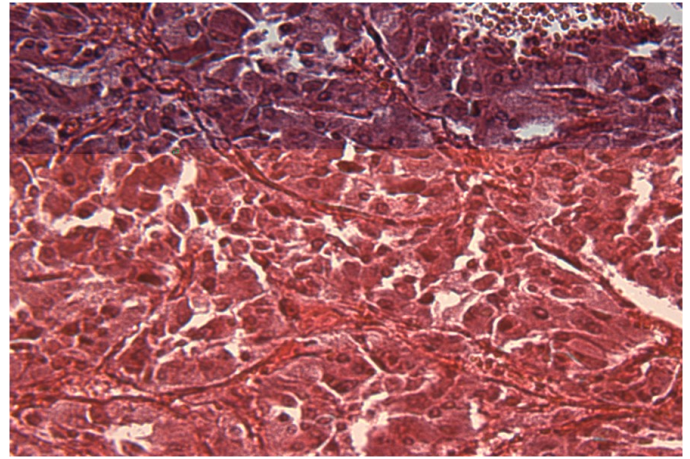


Fig 1. Histology section shows well defined nests of polygonal cells separated by capillary network. (Original magnification X 100)

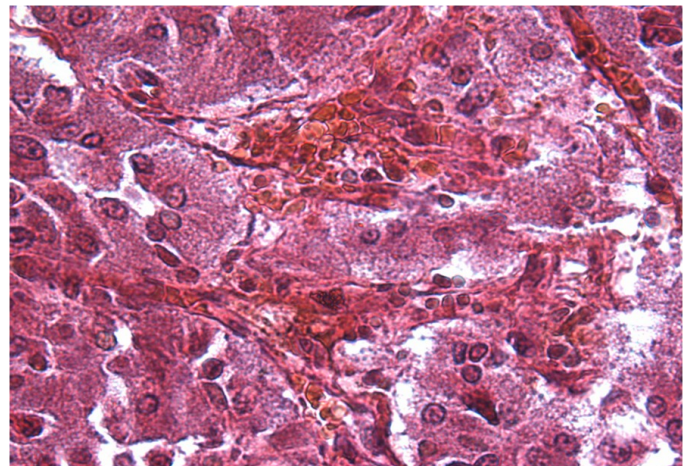


Fig. 2. Histology section shows areas of haemorrhages in the tumour substance. The tumour cells are large with granular eosinophilic cytoplasm. (Original magnification X 200)

patient with phaeochromocytoma. This is a neuroendocrine tumour of the adrenal medulla or extra-adrenal chromaffin tissue. It may produce, store and secrete nor-epinephrine, epinephrine or both.

This tumour is curable if diagnosed and treated on time, but may be fatal if remaining unrecognized or mistreated. This case report explains the similar outcome of an undiagnosed phaeochromocytoma leading to hypertensive cerebral haemorrhage which is an unusual but not unheard cause of death in forensic service. A similar case has been mentioned in the literature<sup>3</sup>.

The catecholamines secreted by the tumour caused repeated attacks of hypertension which ultimately led to rupture of intracranial vessels and fatal intracerebral and intra-ventricular bleed. Left ventricular

hypertrophy of the heart was also secondary to hypertension.

This tumour is called a '10% tumour' because 10% of them are bilateral, 10% are at extra-adrenal sites, 10% occur in children, 10% recur and 10% can turn into malignancy. The tumours in adults are seen in the age group of 40 - 60 years; 80% are unilateral; 97% are intra-abdominal, solitary, highly vascular, and weighing from 100 grams to 3Kg.

Mechanism of release of catecholamines from the tumour cells is yet unknown.

The patient usually presents with severe attacks of hypertension resistant to conventional treatment and or with symptoms of sympathetic stimulation<sup>2</sup>. On examination, severe and sustained rise in blood pressure is recorded with variable arrhythmias and ECG changes<sup>2</sup>.

Phaeochromocytoma sometimes occurs in the context of Multiple Endocrine Neoplasia (MEN type 2) together with medullary carcinoma of thyroid gland<sup>4</sup>. MEN type 2 is a rare familial syndrome caused by mutations in the RET proto-oncogene and is inherited as an autosomal dominant disorder. Diagnosis is made by demonstration of increased production of catecholamines or their metabolites. 24 hour urine sample is the most preferred specimen and free epinephrine is the more specific component for diagnostic purposes (>50 ug/day of free epinephrine suggests the presence of phaeochromocytoma). Measurement of plasma metanephrine is a highly sensitive (99%) technique for phaeochromocytoma but has limited value due to the requirement of high quality assays like HPLC (High Pressure Liquid Chromatography). Other modes of investigation like CAT scans and MRI scans are the preferred choices to localise the tumour<sup>5</sup>. Phaeochromocytoma is a surgically correctable cause of hypertension<sup>5</sup>. Unresectable tumours can be treated by long term use of adrenergic blocking drugs and inhibitors to catecholamine release. Patients with benign tumours localised to adrenals, when treated promptly, will have survival normal to age matched populations. With metastasis the overall 5 year survival reduces to 50%.

## Forensic Pathology Service in Samoa

The pathology department in Samoa is based in Tupua Tamasese Meaole Hospital under the National Health Services. The Department provides routine laboratory services as well as Forensic Services. Forensic autopsies which are referred from the Coroner<sup>6</sup> are performed by the Forensic Consultant Pathologist with the assistance of a mortician and a mortuary technician, but at times of necessity, the Forensic Pathologist asks for help from the Clinical Pathologist.

## Take Home Message

We describe a rare case with the incidental finding on autopsy of a supra-renal tumour. On histology, it was confirmed as phaeochromocytoma of the adrenal gland. This is an uncommon tumour with a fatal outcome if it goes unrecognised.

## References

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