

# A Rare Case Of Retroperitoneal Extra-Adrenal Paraganglioma Presented With Stroke: Case Report And Review Of Literature

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## Citation

R Pedamallu, S Subramanian, D Rambabu, N Kumar, B Satyanarayana, N Rukmangadha. *A Rare Case Of Retroperitoneal Extra-Adrenal Paraganglioma Presented With Stroke: Case Report And Review Of Literature*. The Internet Journal of Urology. 2012 Volume 9 Number 3.

## Abstract

Paragangliomas of the retroperitoneum arise from specialized neural crest cells distributed along the aorta in association with the sympathetic chain. We report a rare case of extra-adrenal paraganglioma between inferior vena cava and aorta in suprarenal portion with lateral displacement of inferior vena cava. This patient presented with left upper motor neuron facial palsy and left hemiparesis secondary to high blood pressure. The rarest presentation of paraganglioma with stroke has never been described to the best of our knowledge in the literature.

## CASE REPORT

A 19 year old man presented with sudden onset of weakness and parasthesia on the left side of body. He complained of severe pulsatile holocranial headache not associated with sweating, palpitations and visual symptoms. He was also recently diagnosed with hypertension.

On examination his blood pressure was 200/108 mm Hg. His neurological examination confirmed left upper motor neuron facial palsy and left hemiparesis. His carotid doppler was reported as normal. There were no masses palpable per abdomen and auscultation did not disclose any bruits.

Laboratory studies showed persistently elevated urinary normetanephrine levels.

An ultrasound performed to rule out adrenal masses showed a well defined hypoechoic lesion between inferior vena cava and aorta in suprarenal portion just above origin of right renal artery, displacing inferior vena cava laterally with no evidence of calcification.

CT scan abdomen revealed a 5.4 x 4 cm sized well defined heterogeneously enhancing lesion posterior to IVC at level of superior mesenteric artery. Mass effect was noted on IVC and right renal vein with no free fluid, no lymph nodes and normal right adrenal gland. These features were suggestive of paraganglioma in retroperitoneum (Fig1, 3).

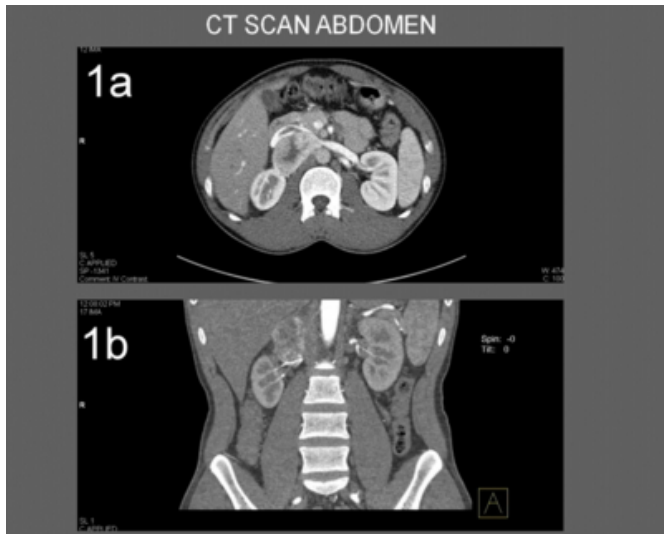
MRI brain showed right thalamic infarct and left frontal lobe infarct (Fig2).

He underwent laparotomy and excision of the lump under general anaesthesia

12 weeks after his initial admission (Fig 4). Histopathological diagnosis is consistent with paraganglioma (Fig 5, 6). His convalescence was uneventful.

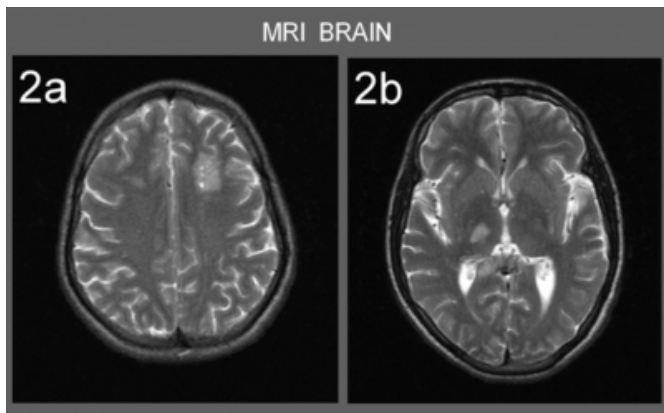
**Figure 1**

CT Scan Abdomen showing a well defined heterogeneously enhancing lesion 3-4cm posterior to IVC at level of superior mesenteric artery.



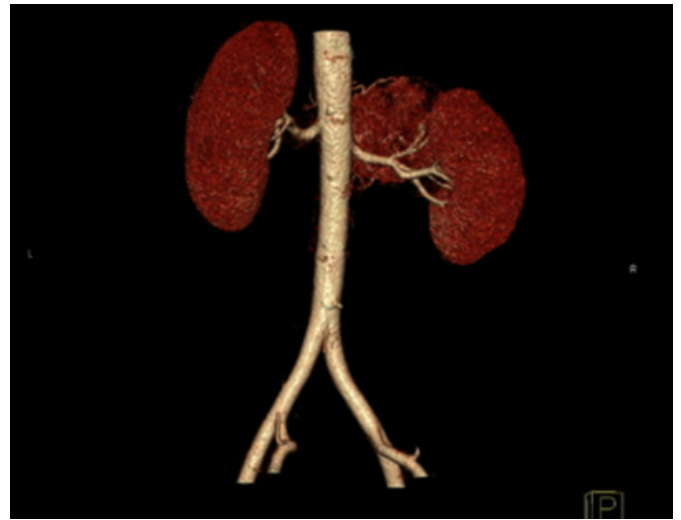
**Figure 2**

MRI Brain: T2 weighted images showing altered signal intensity which is hyperintense in left frontal area and in right thalamic area



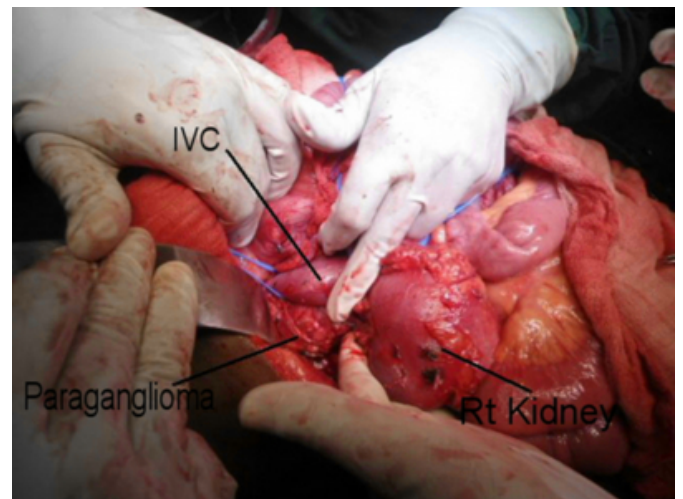
**Figure 3**

3D reconstruction image showing tumour relations to the right renal vessels.



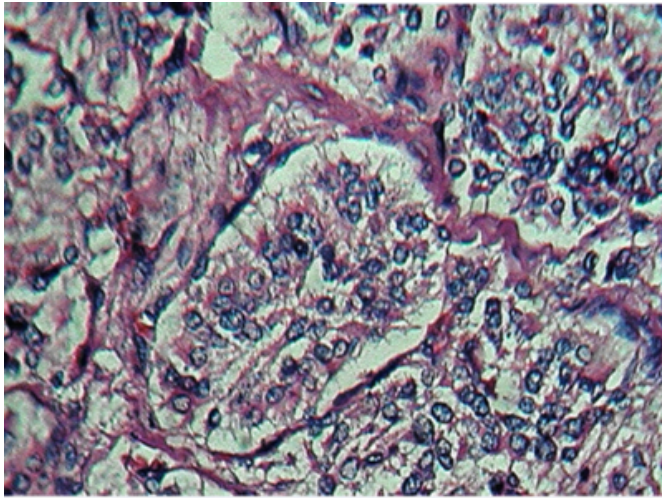
**Figure 4**

Intraoperative photograph demonstrates an extraadrenal paraganglioma located between inferior vena cava and aorta in suprarenal portion just above the origin of right renal artery extending behind inferior vena cava, adherent to it, and stretching it. Right adrenal gland was found to be normal.



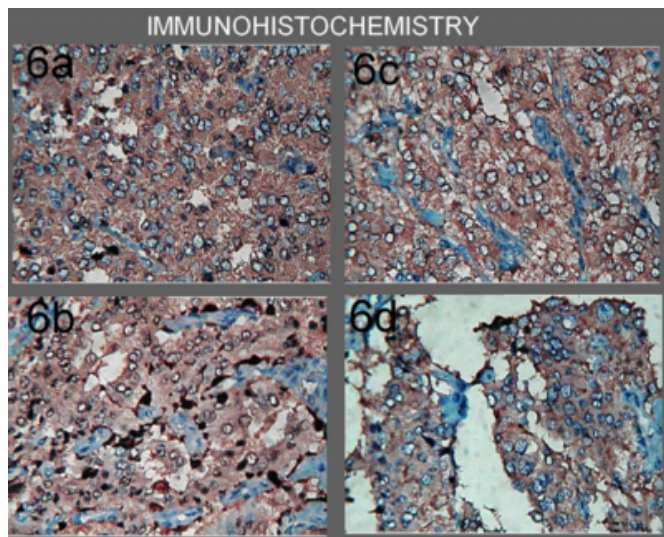
**Figure 5**

H&E stain showing characteristic nesting pattern of cells - Zellballen pattern.



**Figure 6**

Immunostaining with chromogranin neuron specific enolase



(Fig 6c), synaptophysin (Fig 6d) and S-100 for sustentacular cells (Fig 6b) confirmed neuroendocrine origin of the tumour cells.

## DISCUSSION

Paragangliomas of the retroperitoneum are extra-adrenal neuroendocrine tumours arising from sympathetic and parasympathetic components of the autonomic nervous system. Males are affected more frequently than females. Extraadrenal paragangliomas affect patients in the 2nd or 3rd decade of life. It has been reported that 10% to 40% of extra-adrenal paragangliomas are malignant (4). Distant metastasis is the only reliable criteria for confirming malignancy. Local

tissue invasion or pathological evidence of nuclear pleomorphism or mitotic activity does not necessarily imply malignancy (1, 2). Most tumours are hormonally active (functional) and cause clinical findings associated with the excess secretion of catecholamine. These include headaches, sweating, palpitations and hypertension. Paragangliomas can occur anywhere from the base of the brain to the urinary bladder. The common locations for extra-adrenal paragangliomas include the organ of zuckerkanndl, bladder wall, retroperitoneum, heart, mediastinum, carotid bodies and glomus jugulare bodies (1, 2, 3). The majority of paragangliomas present in the head and neck region are nonfunctioning tumours of the parasympathetic system, while those below the neck are frequently functional and associated with the sympathetic system. Approximately, 10% of paragangliomas are multiple. Rarely, a continuous chain of paragangliomas occur in a paraspinal location, a condition known as paragangliomatosis. Multiple endocrine neoplasia types IIa and IIb and Carney's syndrome (defined as the presence of two of the: (1) extra-adrenal paraganglioma, (2) pulmonary chondroma and (3) gastric GIST need to be considered when a diagnosis of extra-adrenal paraganglioma is made (5).

Our case is a unique presentation of retroperitoneal extra-adrenal paraganglioma presented with stroke. Uncontrolled blood pressure despite prompt medication treatment in young male patient with stroke had led to further investigations. Surgical removal of this tumour has normalized his blood pressure and significantly reduced the possibility of further strokes.

Young male patient with uncontrollable blood pressure presented with cerebrovascular accident should arouse the suspicion of retroperitoneal extra-adrenal paraganglioma.

## CONCLUSION

Clinical suspicion of retroperitoneal extra-adrenal paraganglioma in all cases of uncontrollable blood pressure presented as stroke in young male and surgical excision of these tumours can prevent further episodes.

## ACKNOWLEDGEMENTS

Written consent was obtained from the patient for the publication of the case report.

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