

A Rare Case Report: Retroperitoneal Paraganglioma

Dr Jay Chokshi¹, Dr Sandeep Kansal², Dr Mohammed Motiwala^{3*}

¹Assistant Professor, Department of General Surgery, Government Medical College, Surat, India

²Additional Professor, Department of General Surgery, Government Medical College, Surat, India

³rd Year Resident, Department of General Surgery, Government Medical College, Surat, India
Corresponding Author Email: [motiwalamohammed97\[at\]gmail.com](mailto:motiwalamohammed97[at]gmail.com)

Abstract: ***Background:** Paragangliomas are extra adrenal pheochromocytomas arising from chromaffin cells (embryonal derivatives of neural crest cells), able to produce catecholamines. This case report aims at analysing the clinical presentation, diagnosis and treatment outcome of this rare entity. **Case and Discussion:** We encountered a case of young lady with atypical presentation, diagnosed as this rare entity of paraganglioma, arising from paraganglia, similar to pheochromocytoma, extra adrenal in origin, located most commonly in abdomen which creates intraoperative and post operative life - threatening complications if not managed cautiously. **Conclusion:** With time, cautious and appropriate pre operative preparation, this pathology can be treated along with post operative management.*

Keywords: Pheochromocytoma, Retroperitoneal Paraganglioma, Surgery

1. Introduction

Paragangliomas are extra adrenal pheochromocytomas arising from **chromaffin cells** (embryonal derivatives of neural crest cells) which are able to produce catecholamines (epinephrine and norepinephrine).

Extremely Rare (2 - 8 cases/million), mostly benign but may be malignant, arising from paraganglia, similar to pheochromocytoma, more common in male, extra adrenal in location present in **abdomen (>95%)** most commonly, **often** in the **retroperitoneum**, but they can also present in head, face and neck regions as well, notorious to produce symptomatic crisis frequently.

This case report aims at discussing the clinical presentation, diagnosis and treatment outcome of this rare entity.

Case

A 30year old young lady with chief complain of abdominal pain - intermittent, dull aching, over the left side of abdomen since 2 months with normal General Physical Examination. On Per Abdomen examination there was a lump over the abdomen primarily in the left lumbar region, extending in to umbilicus. Her routine blood investigations were normal. Ultrasonography round, hypoechoic, irregular mass of 6*7*6 cm³ suspected to be arising from retroperitoneal region with solid and cystic component.

CECT (A+P): 9*8*7 cm³ sized lobulated soft tissue density lesion with subtle calcification with central 6*5 cm² cystic component extending from inferior pole of left kidney up to aortic bifurcation, abutting IMA and Aorta, with involving proximal 1/3rd of left ureter with moderate hydronephrosis (diagnosed as non - functioning left kidney). [Fig.1&2]; [2]

Biochemical investigations were done in form of plasma epinephrine, norepinephrine (>4000 pg/ml), plasma free

normetanephrine (>3600pg/ml) and urinary VMA (>2000 ml/24h) level which were **elevated**. [3]Preoperative preparation was done in form of Alpha blockers for 15 days followed by Beta blockers for 8 days followed by surgery. [4, 5]

Laparotomy was done and tumor of size 10*10*5 cm³ was found to be extending from DJ junction to inferior pole of left kidney encasing completely - proximal part of left ureter. Tumor was removed along with ureter and due to non - functioning kidney, left nephrectomy was done. [Fig.3]; [7]

Intraoperative duration was **uneventful**. In **Post operative** period patient had **2 episodes of hypoglycemia** and frequent **tachycardia** which was managed with 5% dextrose and cardio - selective beta blocker respectively. [4, 5]

HPE: Gross - combination of cystic & solid components. [Fig.4] Microscopic neoplastic cells nests intervened by highly vascular fibrous septa with Vascular and Capsular Invasion & margins positive. IHC reactive for NSE & S - 100. GAPP score 7/10. [Fig.5 & 6]

2. Discussion

Paragangliomas are extremely rare, rarer in females than males, common in abdominal origin i. e. retroperitoneum is the commonest site in abdomen in between Inferior Mesenteric Artery and Aortic bifurcation known as organ of **Zuckerkundl**.

These rare vascular neuro - endocrine tumor arise due to hereditary genetic (syndromic) or sporadic mutation. [1]Classical symptom triad (headache, sweating, palpitation) not found always as this patient had abdominal pain.

High risk of metastasis such as young age, bilaterality, multifocality, with past or family history; genetic evaluation should be considered. Thorough Biochemical and

Radiological (Anatomical and Functional Imaging) Investigations are the key for diagnosis.

To avoid hypertensive crisis during surgery, **preoperative preparation** is **utmost** in these patients.

Though Surgery is the primary leading treatment option in these patients, but radiotherapy and chemotherapy may play role as palliative or adjuvant therapies due to its malignant and aggressive behavior.

The Grading system for Adrenal Pheochromocytoma and Paraganglioma (GAPP) [6] depending on the patient's points scored by their tumor characters, indicates metastatic potential and 5 - year survival rates.

3. Conclusion

Retroperitoneal Paraganglioma is a Primary Neural crest cell origin tumor which is a **"time bomb"** with high morbidity and mortality, if not treated timely. With clinical suspicion and detailed investigations diagnosis is possible. **Multi - disciplinary** approach is required to manage these patients to avoid complications, but **surgery is key treatment**.

4. Acknowledgements

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5. Declarations

- Funding: None
- Conflict of interest: None
- Ethical Approval: Not required

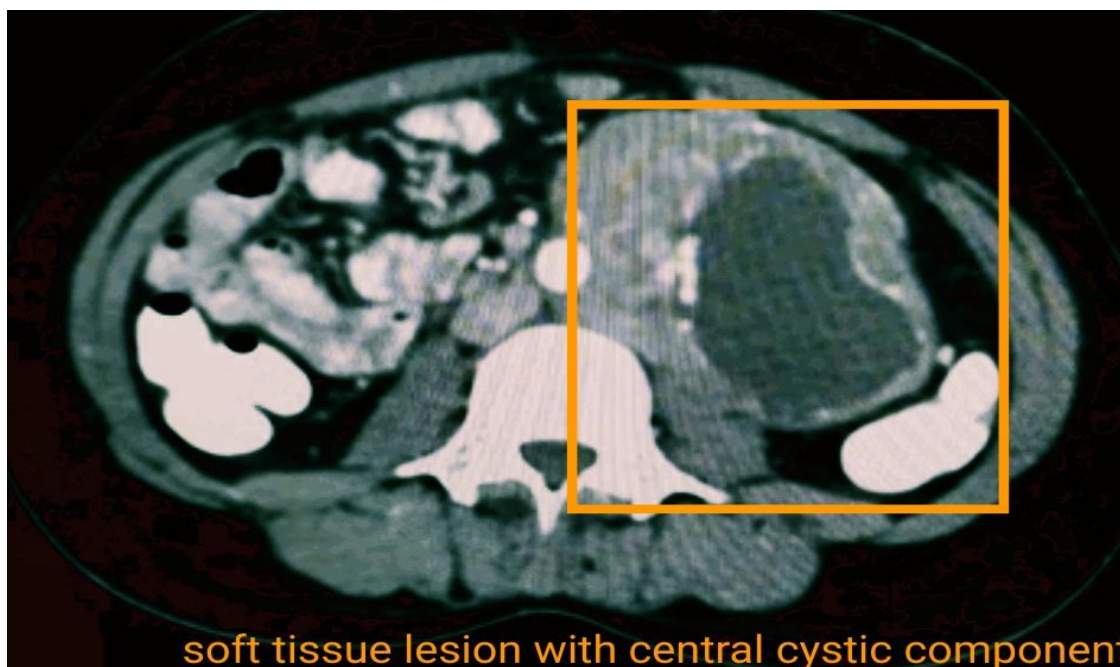


Figure 1: CE - CT (A+P): Suggestive of Soft tissue lesion with solid and cystic components

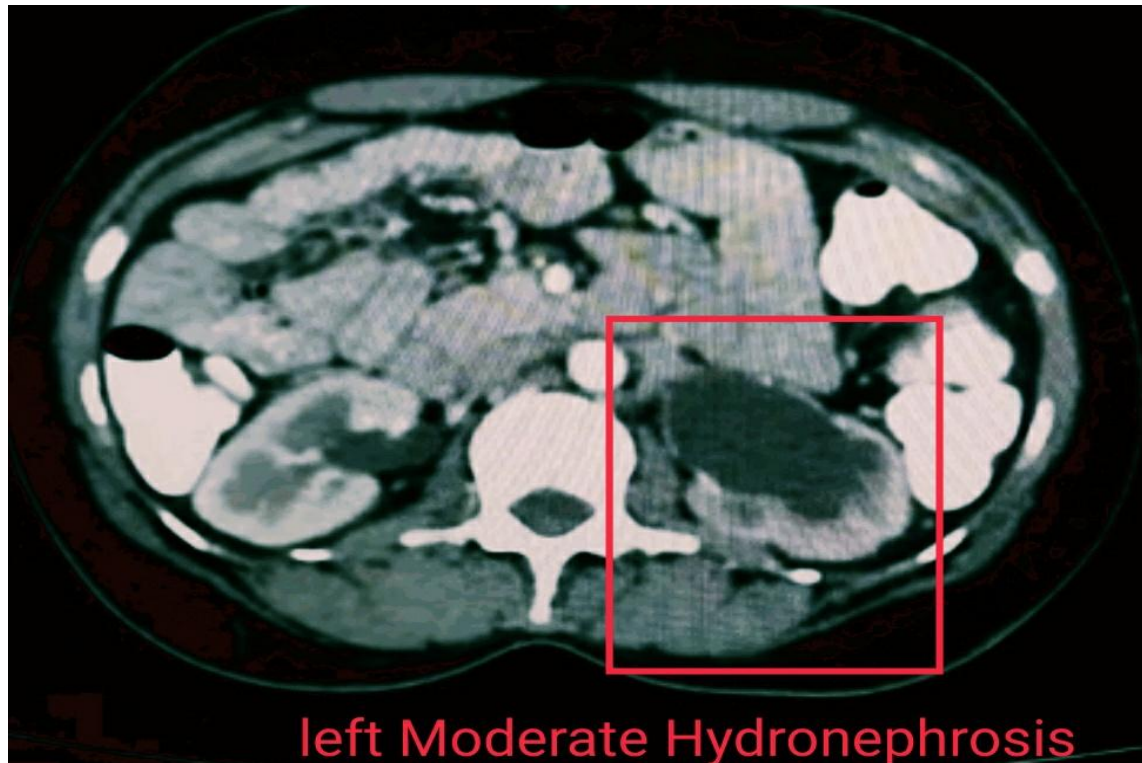


Figure 2: CE - CT (A+P): Tumour involving proximal part of left ureter with left moderate hydronephrosis (with no excretion of dye in left kidney till 48 hours - non functioning left kidney).

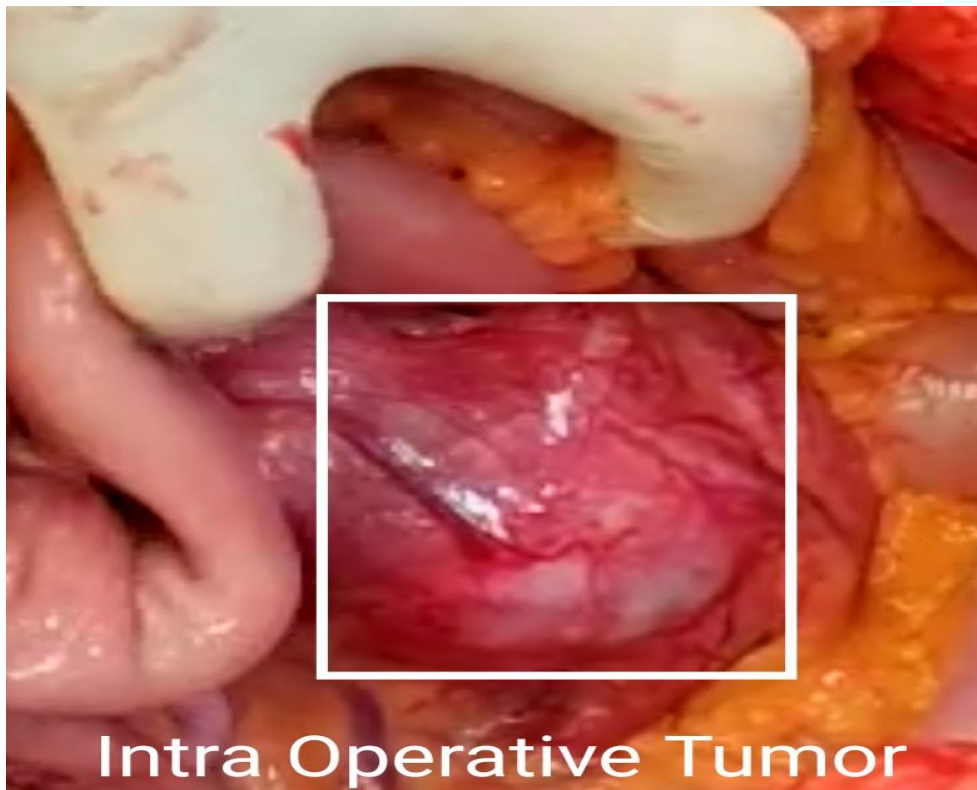


Figure 3: Intra Operative picture of tumour

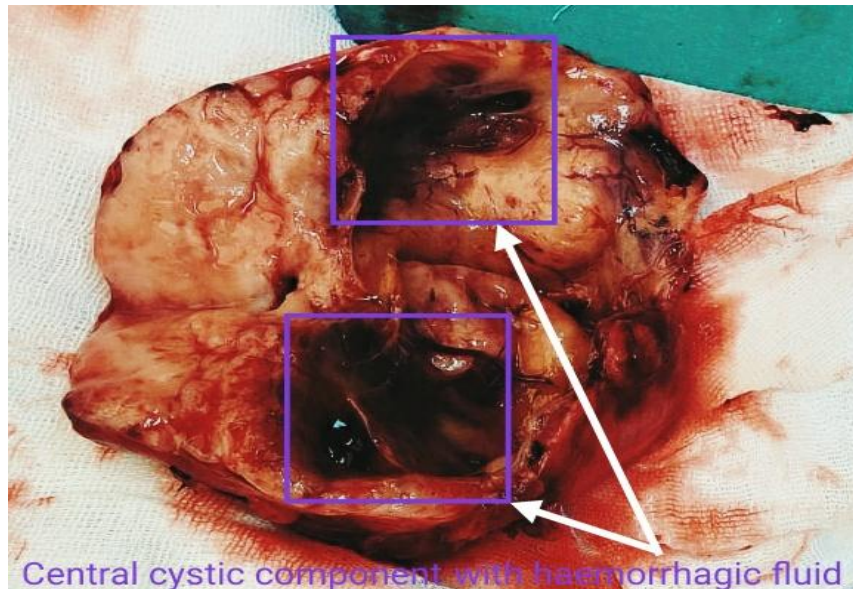


Figure 4: Gross Picture of Tumour showing solid with cystic component

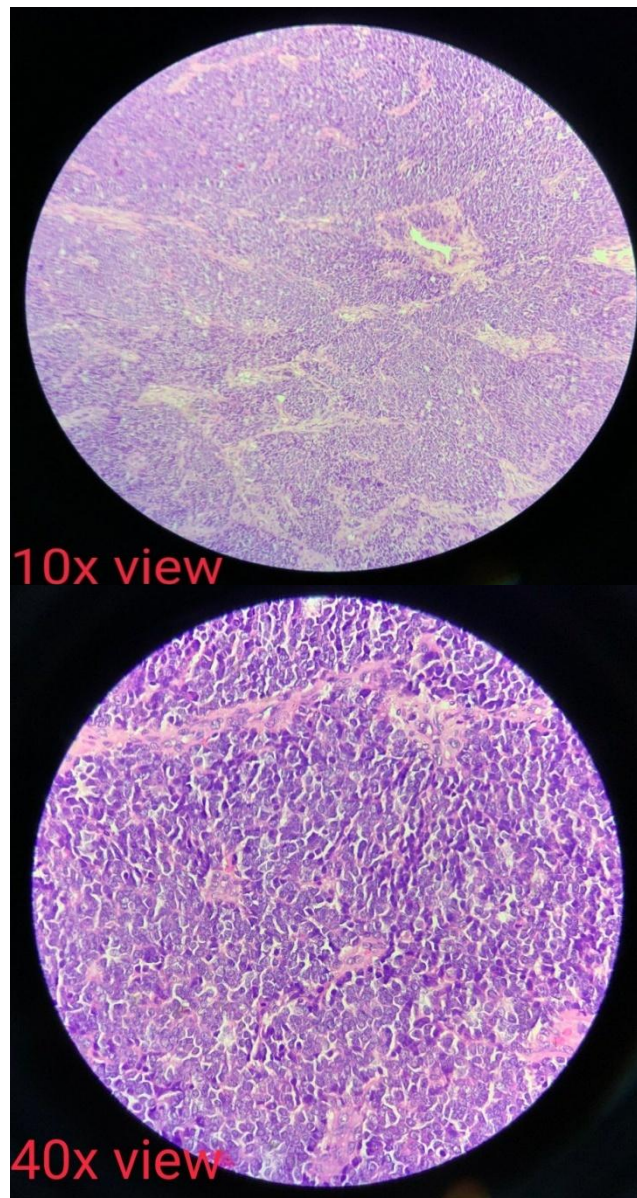


Figure 5 & 6: Microscopic images of Tumour: Suggestive of neoplastic cells separated by highly vascular fibrous septa. Mild to moderate pleomorphism, granular eosinophilic cytoplasm, atypical mitotic figures and areas of hemorrhage seen.