VOLUME - 12, ISSUE - 06, JUNE - 2023 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Surgery

A RARE CASE REPORT: RETROPERITONEAL PARAGANGLIOMA

Dr Mohammed Motiwala*	Registrar-3rd year, Department of General Surgery, Government Medical College, Surat, India *Corresponding Auhtor
Dr Sandeep Kansal	Additional Professor, Department of General Surgery, Government Medical College, Surat, India
Dr Jay Chokshi	Assistant Professor, Department of General Surgery, Government Medical College, Surat, India

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

KEYWORDS : Pheochromocytoma, adrenal, retroperitoneal paraganglioma, hypertension

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 30 year old young lady with chief complain of abdominal pain-intermittent, dull aching, over the left side of abdomen since 2 months, 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 cm^3 suspected to be arising from retroperitoneal region with solid and cystic component.

CECT (A+P): 9*8*7 cm^3 sized lobulated soft tissue density lesion with subtle calcification with central 6*5 cm^2 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]

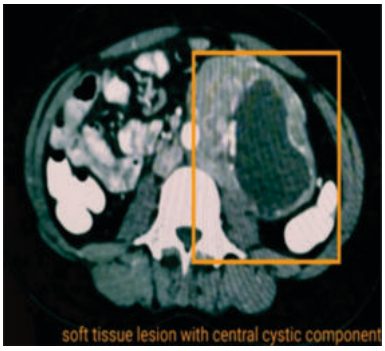

Fig. 1: CE-CT (A+P): Soft Tissue Lesion With Solid And Cystic Components

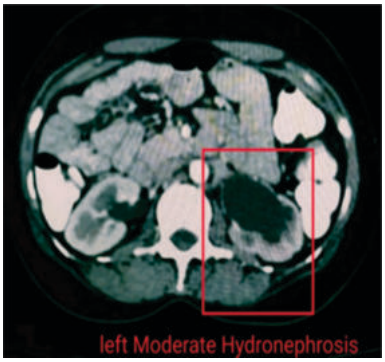

Fig. 2: CE-CT (A+P): Tumour Involving Proximal Part Of Left Ureter With Left Moderate Hydronephrosis (With No Excretion Of Dye In Left Kidney Till 48 Hours-non Functioning Left Kidney).

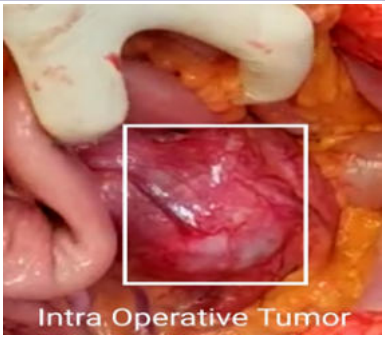

Fig. 3: Intra Operative Picture Of Tumour

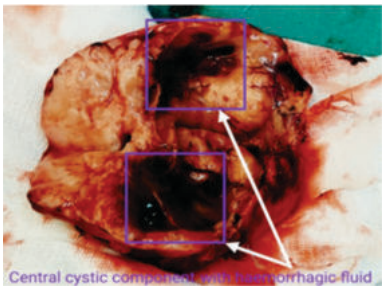

Fig. 4: Gross Picture Of Tumour Showing Solid With Cystic Component.

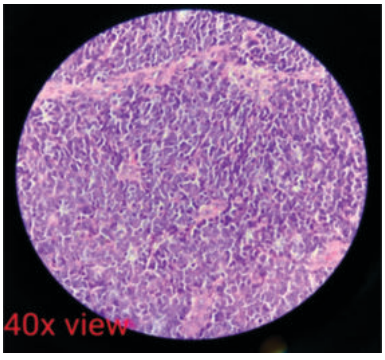

Fig. 5: Microscopic Images Of Tumour: Suggestive Of Neoplastic Cells Separated By Highly Vascular Fibrous Septa. Mild To Moderate Pleomorphism, Granular Eosinophilic Cytoplasm, Atypical Mitotic Figures And Areas Of Hemorrhage Seen.

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) is usual, but atypical features are not uncommon.

High risk of metastasis such as young age, bilaterality, multifocality, 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 must 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.

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 examination and detailed investigations diagnosis is possible. Multi-disciplinary approach is required to avoid complications, but surgery is key treatment.

REFERENCES

1. Bernardo Dias Pereira1 Henrique Vara Luiz2 Ana Gonçalves Ferreira2 Jorge Portugal2 Genetics of Pheochromocytoma and Paraganglioma, Paraganglioma: A Multidisciplinary Approach; pg 1-22
2. M. Nishino, K. Hayakawa, M. Minami, A. Yamamoto, H.Ueda, and K. Takasu, "Primary retroperitoneal neoplasms: CT and MRI imaging findings with anatomic and pathologic diagnostic clues," Radiographics, vol. 23, no. 1, pp. 45–57, 2003.
3. Ying Shen, Liming Cheng Biochemical Diagnosis of Pheochromocytoma and Paraganglioma, Paraganglioma: A Multidisciplinary Approach; pg 23-40
4. Ramachabdran R, Rewari V. Current perioperative management of pheochromoytomas. Indian J Urol. 2017; 33:19–25
5. Fishbein L, Orlowski R, Cohen D. Pheochromocytoma/paraganglioma: Review of perioperative management of blood pressure and update on genetic mutations associated with pheochromocytoma. J Clin Hypertens. 2013;15(6):428–34. http:// dx.doi.org/10.1111/jch.12084
6. Yong Wang, Minghao Li, Hao Deng, Yingxian Pang, Longfei Liu, and Xiao Guan 2020 Am j cancer Res, 2020 The systems of metastatic potential prediction in pheochromocytoma and paraganglioma
7. Baudin E, Habra MA, Deschamps F, Cote G, Dumont F, Cabanillas M, et al. Therapy of endocrine disease: Treatment of malignant pheochromocytoma and paraganglioma.Eur J Endocrinol.2014; 171(3):R111–22. http://dx.doi.org/10.1530/EJE-14-0113