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Malignant Paraganglioma Presenting as an Abdominal Lump: A Case Report

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Abstract: ***Tumours that arise from the neuroectodermal tissue of the adrenal medulla are termed phaeochromocytomas (PCCs) and those arising from the extra-adrenal parasympathetic and sympathetic ganglia are termed paragangliomas (PGLs). PCCs and PGLs are collectively abbreviated to PPGL. PGLs are either parasympathetic or sympathetic. Parasympathetic PGLs are sited mainly in the head and neck (HNPGLs) and 95% do not secrete catecholamines or other hormones. The common types of HNPGL are carotid body, vagal and jugulotympanic. Sympathetic PGLs usually secrete catecholamines. A 26 years old female patient presented to us with the complaint of pain in abdomen and a palpable lump extending from the left lumbar region to the umbilical and left iliac fossa region. Clinical, radiological and histopathological investigations pointed to the differential diagnoses of Desmoid Tumor, Neuroendocrine Tumor and Spindle Cell Tumor. Surgical intervention was done and there were intraoperative BP fluctuations and Adrenergic crisis, managed by Cardio-regulatory drugs. The post-operative histopathology report confirmed the presence of an aggressive Malignant Paraganglioma with lymph node metastases. The surgical and peri-operative events and care involved in the successful management of this case have been described in this case report. Further, it is important to consider possibilities like Paragangliomas/NETs when dealing with suspicious lesions/masses and to keep a Multi-Discipliary team with Emergency management and Critical Care on Standby.***

Keywords: Paraganglioma, neuroectodermal, catecholamines, histopathology, surgical intervention, multi-disciplinary team

1. Introduction

Retroperitoneal paragangliomas are rare tumours of the neural crest cells along the autonomic nervous system, extending from the head and neck to the pelvis, at any location where paraganglia are present; the superior para-aortic region being the most common (46%), followed by the inferior para-aortic region (29%), the bladder (10%) and the thoracic region (10%). Cases of PGLs have also been reported in other abdominopelvic sites as well as in craniocervical regions. These tumours are often difficult to diagnose as the non-specific complaints and wide spectrum of presentations combined with the close proximity to vital structures poses significant danger during surgical resection and makes the procedure demanding.[1]

PGLs can be further divided into sympathetic and parasympathetic types owing to their clinical and biological characteristics. Sympathetic PGLs account for 80% and parasympathetic PGLs account for 20% of PGLs. Sympathetic PGLs arise from the sympathetic nerves in the thorax and abdomen. 85% are located below the diaphragm, especially in the retroperitoneum around the organ of Zuckerkandl, which is a kind of chromaffin tissue located around the abdominal aorta, the inferior mesenteric artery, the beginning of the renal artery and the bifurcation of the abdominal aorta. Parasympathetic PGLs originate from the parasympathetic nervous ganglions, usually in the head and neck region.[2] The treatment of choice is radical resection. In advanced cases, surgery may be possible following chemotherapeutic debulking with cyclophosphamide, vincristine, and dacarbazine. 131I-MIBG radiotherapy has proved to be increasingly useful in reducing the pain associated with disseminated disease and also in facilitating surgical resection in previously inoperable cases. Conventional radiotherapy is purely palliative and it is used to reduce pain, especially that of bone metastases.[3]

2. Case Presentation

A 26 years old female presented to General Surgery OPD with pain and lump in the left lower quadrant of abdomen for 4 months. The pain was on and off, dull, aching in character and was aggravated with vigorous physical activity and partially relieved with oral analgesics. There was no history of palpitations, sweating or a noticeable fluctuation in or rise of blood pressure, accounted for in multiple physician visits. There was no history of weight loss and the lump had gradually increased in size over the last four months. On examination, there was a non-tender, hard retroperitoneal lump extending from the umbilicus to the left lumbar and left iliac fossa region measuring 4x5 cm. in size. It had smooth edges, regular margins and was partially movable along horizontal and vertical directions. The patient had normal vitals and no co-morbidities were present. USG and CECT reports suggested a highly vascular mass in the mid/left abdomen or mesentery, displacing the bowel loops towards midline and multiple enhancing mesenteric lymph nodes were present. The mass itself was reported as having solid and cystic areas with areas of calcification and necrosis. The differentials of GIST, Desmoid Tumor or a large Lymph Node were

suggested. USG-guided FNAC suggested an Intermediate to High grade Spindle Cell Neoplasm. A confirmatory diagnosis could not be established. Midline exploratory laparotomy was done and on coming in contact with the tumor, there was sudden overshoot of BP (180-220 mm Hg Systolic and 110-140 mm Hg diastolic) and Pulse Rate (130-150 bpm). Surgery was halted and Cardiology call was sent. Central Venous Canulation and Peripheral Arterial Canulation were done and the patient was started on iv Labetalol, Nitroglycerine, Esmolol and Dexmedetomidine. Surgery was resumed after stabilization of vitals and a mass ~6x7 cm in size was isolated near the bifurcation of Abdominal Aorta between the Right and Left Common Iliac Vessels. Enlarged lymph nodes were seen in clusters and dissected from above the right and left iliac arteries. In the post-operative period, the patient was managed on Prazosin, Bisoprolol, Ivabradine and Magnesium Sulphate tablets for controlling BP and pulse rate. Post-operatively, urine free Metanephrines and Normetanephrines were raised, i.e., 2 nmol/L and 3 nmol/L, respectively. Histopathological report revealed the presence of a tumor with epithelioid chief cells arranged in clusters (Zellballen pattern) separated by fibrovascular stroma, features suggestive of high-grade aggressive Paraganglioma with 17 lymph nodes isolated, 7 of which showed involvement by the tumor. IHC reporting showed CD56+, Synaptophysin+, Chromogranin+, S100+ and Ki67 1-2%. The diagnosis of malignant metastatic PGL was confirmed.

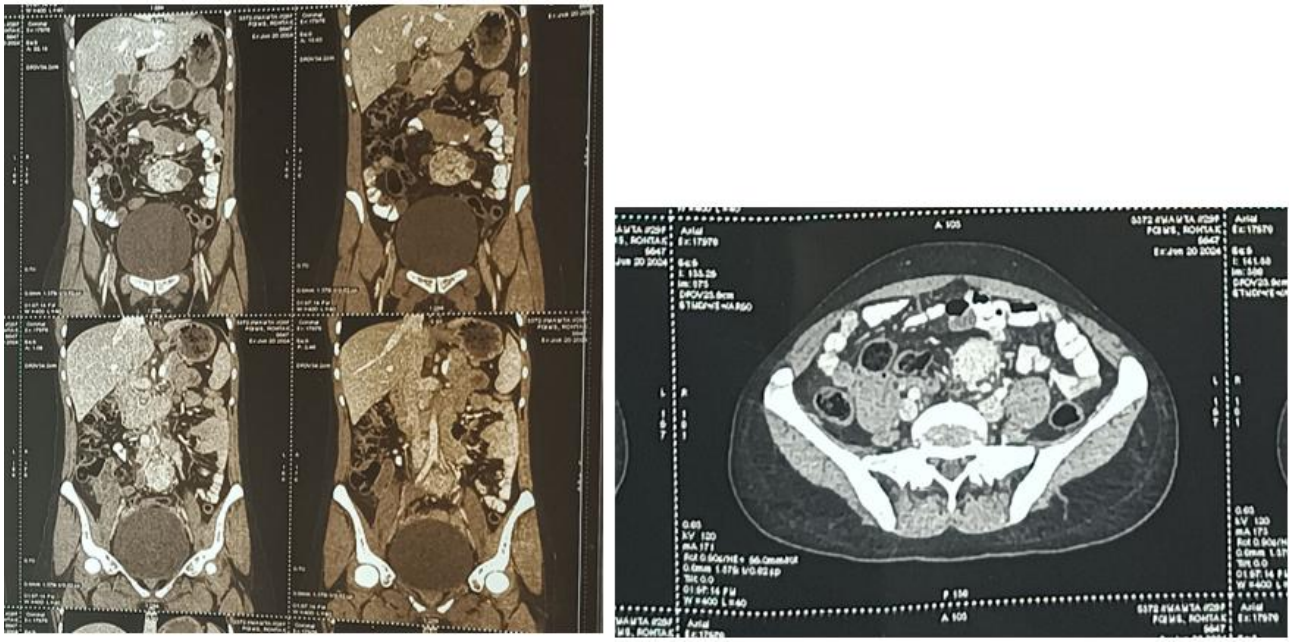

Figure 1: CT sections showing retroperitoneal mass near aortic bifurcation.

3. Discussion

The retroperitoneum can host a wide spectrum of rare pathologies, including benign and malignant tumours. Tumors usually present late and cause symptoms or become palpable once they have reached a significant size. These tumours are best evaluated with good quality cross-sectional imaging and preoperative histology by core needle biopsy is required when imaging is non-diagnostic.[4]

PGLs are less common than Pheochromocytomas and are an unusual cause of retroperitoneal masses. They may not present with the classical features like hypertension, headache, sweating, palpitations, etc. Imaging might show a vascular tumor with cystic areas or central necrosis and location near the great vessels should alert the clinician and the surgeon about the possibility of a PGL. Depending on feasibility, I-MIBG Scan or a 18-Fluorodopamine PET Scan should be carried out to clinch the diagnosis due to its various genetic associations and the option of Gene Testing should be provided for the benefit of future generations. Complete surgical resection followed by radiotherapy is the mainstay of treatment for resectable tumours and palliative radiotherapy or neoadjuvant chemotherapy can be tested out for unresectable tumours.

For patients exhibiting functional PGLs, the most common type of presenting symptom was the classic triad of catecholamine-secreting PGLs: episodic headache, diaphoresis and tachycardia. Recent studies and case series have identified that functional tumors occur in approximately 59% patients, of which only 60% patients exhibited hypertension preoperatively. The remaining 40% patients with no preoperative hypertension exhibited a fluctuation in blood pressure during dissection of the tumor intraoperatively. Additionally, for patients with non-functional PGLs, the most common type of presenting symptom was abdominal mass (46%). Non-functional retroperitoneal paraganglioma, which lacks symptoms at the early stage, is identified to be markedly larger compared with the functional tumor, mostly due to their late discovery. Further, a number of previous studies have revealed that patients with PGLs additionally also show a variety of uncommon non-specific symptoms, including palpitations, panic attacks, abdominal discomfort and dyspnea.[5]

When presenting within the abdomen, they may arise as a primary retroperitoneal neoplasm and can be mistaken for other retroperitoneal tumors, including those arising from the pancreas.[6] For retroperitoneal PGLs, the median diagnostic age is 37-43 years with roughly similar incidence among females and males (1.3:1). The average size, rate of metastasis (i.e., malignancy), and rate of function is 7.4 cm, 9.5%, and 57.1%, respectively. In some case series, tumors greater than 7 cm are more likely to require resection of adjacent organs(P=0.01) and the overall 5-year survival has been seen to be approximately 73%. Survival was significantly worse after metastasis (P=0.0023) but did not depend on the tumor diameter, the secreting function of the tumor, the status of surgical margins of resection, or status of the resected lymph nodes. They might initially present with non-specific complaints like lower back pain, abdominal heaviness, urinary symptoms or changes in general health. Imaging may show a solid-cystic mass with necrotic appearance or calcifications; however, a definitive diagnosis can be reached only via histological analysis. PGLs with chief cells and sustentacular cells grouped in clusters (Zellballen pattern) clubbed with immunohistochemical picture of synaptophysin, NSE and chromogranin positivity in chief cells and S100 positivity in sustentacular cells.[6,7] Complete surgical resection is the only potential curative treatment modality for retroperitoneal tumors as there are chances of malignancy and is best performed in high-volume centres by a multidisciplinary team, owing to the possibility of intraoperative cardiovascular catastrophe that might arise in cases of pheochromocytoma and paraganglioma. A team of experienced anaesthesiologists and cardiologists must be kept on stand-by in order to handle these situations. Further research is required to develop novel biological therapies to target the various molecular pathways resulting in this pathology.[8,9] In cases where the pre-operative diagnosis is clear, the surgical management of a hormonally functional paraganglioma must be preceded by 7 days of preoperative medication, that is, α blockers, while β-blockers and calcium channel blockers can also be prescribed, only in association with α-blockers. Prescribing β Blockers alone may cause to per-operative issues as it increases catecholamine-induced vasoconstriction. Some proposed protocols advise using a 3 days-infusion of Urapidil and Magnesium sulfate, having better treatment compliance. The challenge of the anaesthetist is to control blood pressure and volume and to prevent cardiac rhythm disorders, which might require invasive monitoring. Even if there are no recommendations for the induction agents, propofol remains the most indicated. Ketamine and phenothiazines should be avoided, as they indirectly increase catecholamine secretion. Etomidate has a beneficial effect in patients with hemodynamic instability. The major risk after tumor resection is circulatory collapse explained by vasomotor paralysis.[9,10]

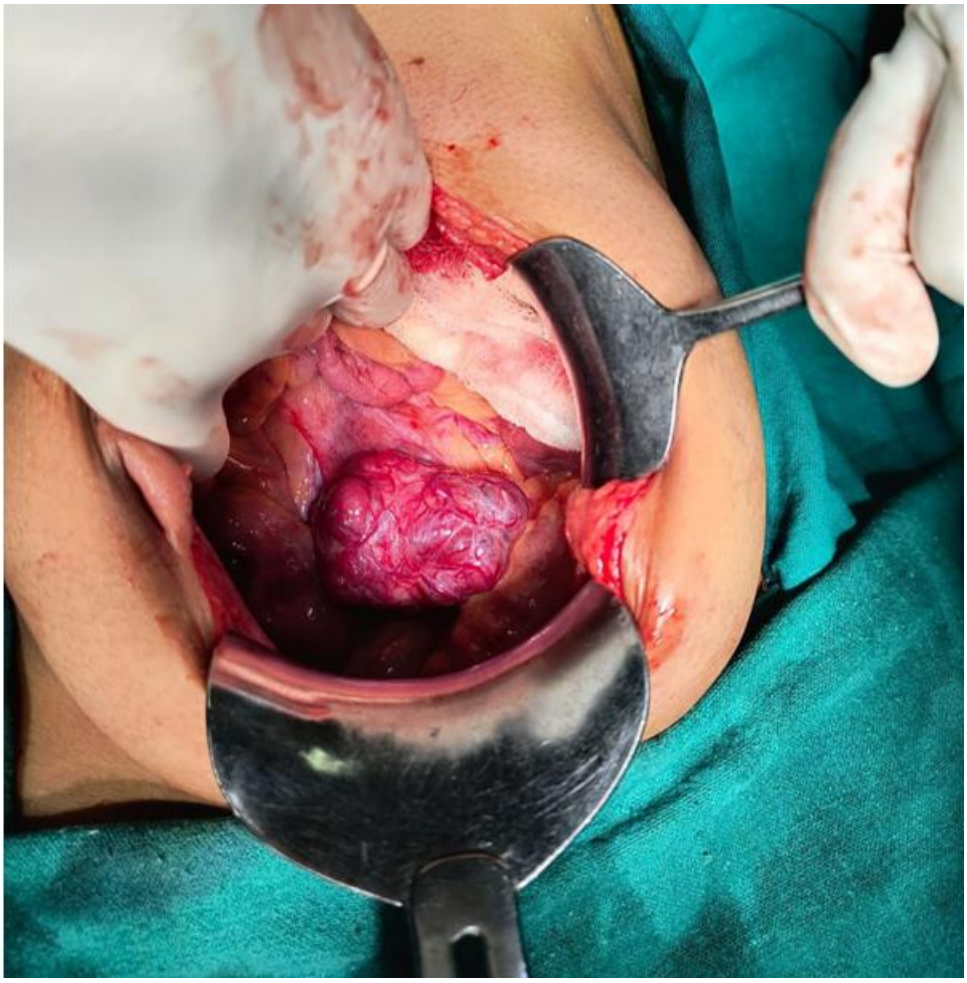

Figure 2: Retroperitoneal mass encountered during Exploratory Laparotomy

Figure 3: (a) Excised paraganglioma and clusters of lymph nodes. (b) Weight of excised main specimen.

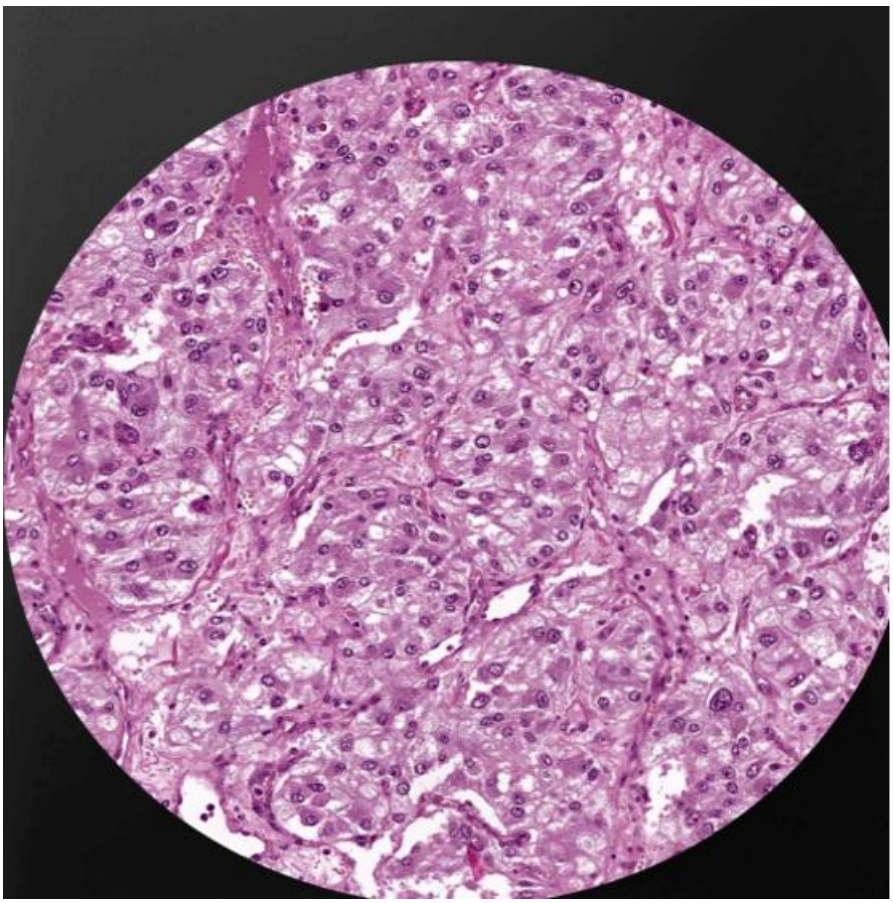

Figure 4: Histopathological image showing Zellballen pattern

4. Conclusion

In conclusion, retroperitoneal paragangliomas are rare tumors, mostly benign with good prognosis, but can be locally invasive and metastasize as well. They can present with pain, mass or hypertensive episodes, lr have varied non-specific signs and symptoms. Following imaging and histological analysis, patients should be initially evaluated with catecholamine levels, followed by tumor-specific imaging to locate the primary lesion and to understand its nature. Surgical excision remains the mainstay of treatment, although advanced disease and prominent vascularity can at times make excision difficult or impossible. An experienced and dedicated cardiology and anaesthesiology team should be available when dealing with tumors of unknown pathology and in diagnosed cases of PPGLs.

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