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CASE REPORT

Para-aortic paraganglioma and its surgical management: A case report

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ABSTRACT

Background: Paragangliomas are rare neuroendocrine tumors arising from chromaffin cells of the autonomic nervous system. They are challenging in children due to rarity and variable presentation. **Case Report** We report a 15-year-old boy with headache, palpitations, sweating, and persistent hypertension. Plasma and urine catecholamines were markedly elevated. CT and MIBG imaging localized a functioning para-aortic mass. Preoperative alpha blockade followed by beta blockade was given. He underwent midline laparotomy with complete excision of a highly vascular tumor abutting the abdominal aorta. Histopathology confirmed a benign paraganglioma without malignant features. Recovery was uneventful, and both blood pressure and catecholamine levels normalized. At three months he remained asymptomatic with normal laboratory values on follow-up. **Conclusion** This case supports consideration of paraganglioma in adolescents with unexplained hypertension, emphasizes multidisciplinary care, and highlights the need for long-term surveillance for recurrence.

Keywords: Functional Neuroendocrine Tumor, Para-aortic Paraganglioma, Pediatric Paraganglioma, Surgical Excision

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Introduction

Paragangliomas are extremely uncommon neuroendocrine tumors arising from autonomic nervous system chromaffin cells beyond the adrenal medulla. Their incidence has been found to be as low as 2–8 per million in the overall population, making them a diagnostic challenge especially in pediatric and adolescent populations where they are exceedingly rare (1,2). Para-aortic paragangliomas are specifically derived from sympathetic ganglion cells along the paravertebral sympathetic chain in close proximity to the abdominal aorta, and thus their anatomical position is surgically challenging due to their proximity to large vascular structures (2,3,4). Para-aortic paragangliomas represent about 15–20% of all pheochromocytomas and are either functional (catecholamine-secreting) or non-functional, with functional tumors being more frequently associated with classic sympathetic hyperactivity symptoms (5,6).

In adolescents, paragangliomas represent an exceptionally rare entity with distinct clinical implications. The rarity of these tumors in adolescents contributes to diagnostic delays

and may lead to prolonged exposure to uncontrolled catecholamine excess, potentially resulting in end organ damage, including catecholamine induced cardiomyopathy (5). Paragangliomas in the pediatric and adolescent population also demonstrate a stronger genetic predisposition compared to adult cases, with up to 77% of cases associated with germline mutations in susceptibility genes. The most commonly affected genes include succinate dehydrogenase subunits (particularly SDHB), VHL, RET, and HIF2A (7).

Clinically, functional paragangliomas typically present with the classic triad of headache, sweating, and palpitations, accompanied by sustained or paroxysmal hypertension that may be difficult to control with conventional antihypertensive medications (5,8). Biochemical diagnosis includes elevated plasma and urinary metanephrines and catecholamines, while anatomical localization requires cross-sectional imaging through CT, MRI, and functional imaging such as MIBG scintigraphy or positron emission tomography (4,8).

The optimal management of paragangliomas involves surgical excision, though this presents significant challenges due to the tumor's rich blood supply and tendency to release catecholamines when manipulated during surgery (2,8). Careful preoperative preparation is essential, including alpha-adrenergic blockade, beta-blockade, and volume expansion to prevent dangerous blood pressure spikes during surgery and hypotension after tumor removal (5,8). Surgical approaches have evolved from conventional open procedures to less invasive techniques, such as laparoscopic and robotic-assisted operations, which provide better precision and visualization, particularly for tumors in difficult anatomical locations. (2,4)

This case demonstrates the successful surgical treatment of a para-aortic paraganglioma in a teenage boy. We offer here practical insights into effective treatment strategies that may benefit the management of similar cases in the future.

Case Presentation

A 15-year-old male patient came to the outpatient department of Lady Reading Hospital having complaints of headache, sweating, palpitations, and persistently elevated blood pressure. He was having frontal, throbbing headache for about 3 months, and sweating and palpitations for 2 months. His hypertension was incidentally detected one month before presentation, which led to referral to our center.

The patient had no significant past medical history, with no previous hospitalizations or surgeries, and no history of medication use. Family history was negative for early-onset hypertension, endocrine disorders, tumors, or sudden cardiac death. He was a secondary school student with good academic performance and no history of substance use.

Physical examination revealed an alert, mildly anxious adolescent with blood pressure of 170/100 mmHg confirmed on multiple measurements, heart rate of 110 beats per minute, respiratory rate of 18 breaths per minute, and temperature of 37.0°C. He weighed 58 kg with a height of 167 cm (BMI 20.8 kg/m²). Cardiovascular examination showed regular rhythm with tachycardia but no murmurs. Respiratory and neurological examinations were unremarkable, and his abdomen was soft with no palpable masses.

Initial laboratory investigations, including complete blood count, renal and liver function tests, serum electrolytes, urinalysis, and glucose studies were within normal limits.

Endocrine workup showed significantly elevated 24-hour urinary catecholamines (norepinephrine 620 µg/24h, epinephrine 110 µg/24h) and metanephrines (normetanephrine 1450 µg/24h, metanephrine 480 µg/24h). Plasma metanephrines were similarly elevated (normetanephrine 1.2 nmol/L, metanephrine 0.8 nmol/L).

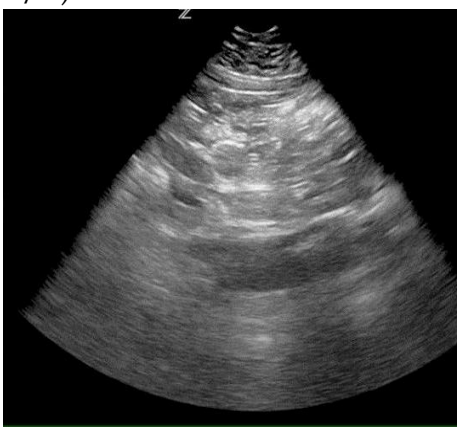

Figure 1: Transverse abdominal ultrasound image

Abdominal ultrasound was inconclusive. CT abdomen with contrast demonstrated a well-defined, enhancing mass (6.4 x 3.9 cm) adjacent to the abdominal aorta at the level of the renal arteries.

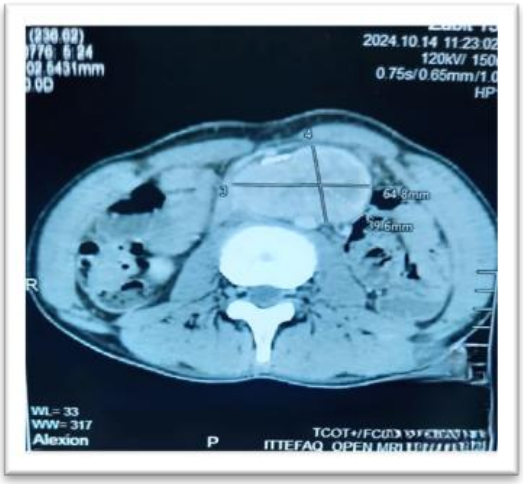

Figure 2: Contrast-enhanced CT showing a vascular para-aortic mass measuring 64.8 × 39.6 mm in 15-year-old male, consistent with functional paraganglioma.

Pre-operative management included alpha-blockade with phenoxybenzamine (gradually increased to 10 mg three times daily), followed by beta-blockade with propranolol 20 mg twice daily for tachycardia control. The patient was encouraged to maintain liberal salt and fluid intake.

A multidisciplinary surgical team performed the operation with invasive hemodynamic monitoring. Through a midline laparotomy approach, they accessed the retroperitoneum and identified a highly vascular, reddish-brown tumor (Figure 3) intimately associated with the abdominal aorta. The tumor received multiple feeding vessels directly from the aorta, which were meticulously ligated. Complete resection was achieved with preservation of aortic wall integrity. Intraoperative blood pressure fluctuations were managed with short-acting agents. The estimated blood loss was 600 ml, requiring transfusion of 2 units of packed red blood cells.

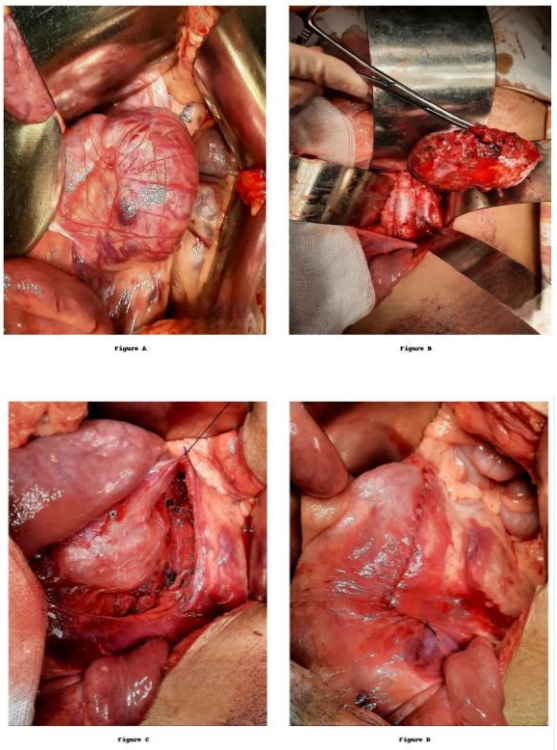

Figure 3(A–D): Intraoperative findings of a para-aortic paraganglioma showing encapsulated mass

(A), excised tumor (B), exposed surgical bed post-resection (C), and final closure after hemostasis (D). Histopathological examination confirmed the diagnosis of paraganglioma with no evidence of malignancy (Figure 4).

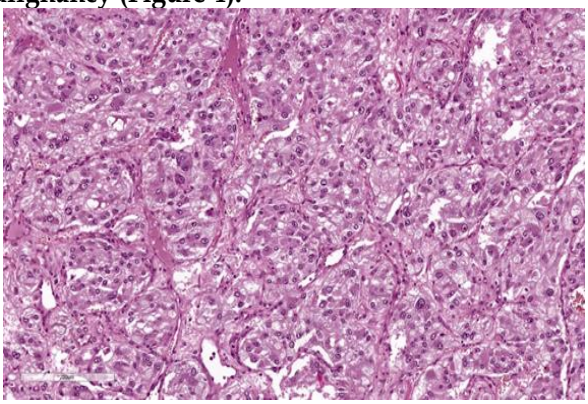

Figure 5: Histopathological section of the para-aortic paraganglioma showing spindle shaped cells.

The patient was monitored in the intensive care unit for 48 hours post-operation. Blood pressure normalized within 24 hours after surgery without antihypertensive medications. Plasma and urinary catecholamines returned to normal levels when checked one-week post-operation. Oral intake was resumed on postoperative day 2, and the patient was discharged on day 6 in stable condition.

At two-week follow-up, the patient maintained normal blood pressure (115/75 mmHg) without medications. One-month follow-up confirmed resolution of all presenting symptoms. Three-month follow-up showed plasma metanephrines remaining within normal limits. The patient was enrolled in a long-term surveillance plan with annual clinical assessment and biochemical testing. Genetic testing for hereditary paraganglioma syndromes was recommended and is pending.

Discussion

Paragangliomas in adolescents remain exceptionally rare, posing diagnostic challenges due to their non-specific initial symptoms and low clinical suspicion, potentially leading to significant delays in diagnosis. A comprehensive review by the French Society of Pediatric Oncology identified only 81 children with paragangliomas over a 16-year period (2000–2016), with 40 cases involving extra-adrenal tumors (7).

Our patient presented with the classic symptomatic triad of headache, sweating, and palpitations, accompanied by persistent hypertension. This clinical presentation aligns well with reports in pediatric literature, where up to 90% of pediatric patients exhibit sustained or paroxysmal hypertension, and symptomatic presentation frequently includes headaches, palpitations, and diaphoresis (9,10).

The biochemical diagnosis in our case was confirmed by significantly elevated urinary and plasma metanephrines and catecholamines, consistent with current guidelines advocating the use of these tests for accurate biochemical confirmation of functional paragangliomas (11,12). Anatomical localization was effectively achieved through multimodal imaging, including ultrasound, Contrast enhanced CT, and functional MIBG scintigraphy. Preoperative management is essential for reducing perioperative complications due to catecholamine surges. This approach is strongly recommended to mitigate intraoperative hemodynamic instability (11). The multidisciplinary surgical team successfully managed significant intraoperative blood pressure fluctuations with short-acting agents, underscoring the importance of vigilant hemodynamic monitoring.

The surgical approach via midline laparotomy allowed optimal exposure and facilitated meticulous vascular control of feeding vessels directly originating from the abdominal aorta. Complete tumor resection, as achieved in our patient, remains the cornerstone of definitive management, significantly reducing recurrence risk and improving long-term prognosis (9,10). Despite advances in minimally invasive

techniques, open surgery remains justified for larger and anatomically challenging tumors, as in our case (10). Histopathological evaluation confirmed the diagnosis of benign paraganglioma, which was consistent with the majority of cases (13). Nevertheless, paragangliomas exhibit a stronger genetic predisposition, necessitating genetic testing for hereditary syndromes, particularly SDHx mutations, which carry significant prognostic implications regarding recurrence and malignancy risk (12,14). Genetic testing was recommended and pending in our patient, influencing future surveillance strategies and familial screening recommendations. Post-surgical follow-up is critical for adolescent patients with paragangliomas due to the risk of recurrence or metachronous disease, particularly in those with underlying genetic syndromes.[6] A 10-year event-free survival rate of only 39% has been reported in pediatric paraganglioma cases, although overall survival remains excellent at 97% (6). Long-term surveillance is imperative, particularly in pediatric populations, due to a higher genetic predisposition and potential for late recurrences or metastases.[11] Our patient demonstrated excellent early postoperative outcomes, with normalization of blood pressure and biochemical markers, but rigorous annual surveillance remains necessary. Current guidelines recommend lifelong or at least 10-year follow-up involving clinical assessments, biochemical testing, and periodic imaging, tailored according to the genetic background and risk factors identified. (11)

Conclusion

In conclusion, this case underscores the critical importance of considering functional paragangliomas in the differential diagnosis of adolescent hypertension accompanied by catecholamine-excess symptoms. Timely diagnosis, multidisciplinary surgical management, effective preoperative preparation, and structured long-term surveillance significantly improve outcomes in these rare but potentially life-threatening tumors.

Conflict of Interest: The authors declare no conflict of interest.

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Manuscript writing and approval	MD, SA, MM, SU
All the authors agree to take responsibility for every facet of the work, making sure that any concerns about its integrity or veracity are thoroughly examined and addressed.	