Invasive Adrenal Pheochromocytoma in a Child Requiring Total Nephrectomy: Anaesthetic Challenges.

Abstract

Pheochromocytoma is a rare neuroendocrine tumor in children and represents a major anaesthetic challenge because of abrupt catecholamine-related haemodynamic instability during surgery. Large invasive tumors closely related to major vessels are particularly high-risk because they combine hypertensive crises during tumor manipulation with severe hypotension and possible major bleeding after tumor devascularization. We report the case of a child with an invasive adrenal pheochromocytoma abutting the abdominal aorta and associated with ipsilateral renal artery occlusion. Preoperative alpha-adrenergic blockade and volume optimization were achieved before surgery. General anaesthesia with invasive monitoring, central venous access, careful induction, and thoraco-lumbar epidural analgesia was used. Marked blood pressure fluctuations occurred during tumor handling and were controlled with short-acting antihypertensive agents, mainly nicardipine. After tumor excision, transient hypotension required norepinephrine support. Surgical dissection proved difficult because of vascular adherence, and massive intraoperative bleeding led to haemorrhagic shock requiring red blood cell transfusion and targeted fibrinogen administration for acquired coagulopathy. Complete excision required adrenalectomy associated with total nephrectomy. Postoperative recovery in the intensive care unit was favourable, with haemodynamic stabilization, extubation in safe conditions, effective epidural analgesia, and transfer back to the referring ward after 48 hours. This case highlights the importance of meticulous preoperative preparation, invasive haemodynamic monitoring, anticipation of both catecholamine surges and haemorrhagic complications, and close multidisciplinary coordination in pediatric pheochromocytoma surgery.

Keywords

Pheochromocytoma; child; anaesthesia; adrenal tumor; nephrectomy; haemorrhagic shock; perioperative management

Introduction

Pheochromocytoma is a rare neuroendocrine tumor arising from chromaffin cells of the adrenal medulla. In children, it accounts for less than 10% of reported cases and is characterized by excessive catecholamine secretion, which may lead to severe hypertension, headache, diaphoresis, palpitations, and major perioperative haemodynamic instability.

Surgery is the definitive treatment. However, anesthetic management remains challenging because tumor manipulation may precipitate abrupt hypertensive crises, whereas venous ligation and tumor removal may be followed by profound vasoplegia and hypotension. The risk is even greater in large invasive tumors adherent to major vascular structures.

We report a case of invasive adrenal pheochromocytoma in a child, closely related to the abdominal aorta and associated with ipsilateral renal artery occlusion, requiring adrenalectomy and total nephrectomy. The case underlines the anaesthetic and haemostatic challenges of this rare high-risk situation.

Case Presentation

A child was referred for surgical management of an adrenal pheochromocytoma. Abdominopelvic contrast-enhanced computed tomography demonstrated a large heterogeneous hypervascular adrenal mass with intense enhancement after contrast injection. The tumor was closely related to major vascular structures, particularly the abdominal aorta, and was associated with occlusion of the ipsilateral renal artery, indicating significant renal vascular involvement and an invasive pattern.

These anatomical findings suggested complex surgical dissection with a high risk of major bleeding and led to the decision to perform tumor excision associated with total nephrectomy.

Perioperative and Anaesthetic Management

Preoperative antihypertensive preparation was instituted to reduce the risk of perioperative haemodynamic complications. The patient received alpha-adrenergic blockade, allowing improved blood pressure control and preoperative volume optimization. Haemodynamic balance was considered satisfactory at the time of surgery.

Given the anticipated instability related to catecholamine release, general anaesthesia with invasive monitoring was performed. An arterial line was inserted for continuous blood pressure monitoring and a central venous catheter was placed for rapid administration of vasoactive drugs and blood products. Anaesthetic induction was progressive in order to avoid excessive sympathetic stimulation. Anaesthesia was maintained with strict control of anaesthetic depth and ventilation.

Thoraco-lumbar epidural analgesia was added as part of a multimodal analgesic strategy to reduce opioid requirements and contribute to perioperative haemodynamic stability while facilitating postoperative pain control.

During tumor manipulation, major blood pressure fluctuations occurred and were treated with short-acting antihypertensive agents, particularly nicardipine. After tumor excision, arterial hypotension required transient vasopressor support with norepinephrine.

Intraoperative Course

Surgical dissection proved particularly difficult because of the invasive nature of the tumor, its close adherence to the abdominal aorta and other major vascular structures, and the occlusion of the ipsilateral renal artery. These anatomical constraints rendered tumor mobilization hazardous and exposed the patient to a high risk of bleeding.

During dissection, abundant intraoperative haemorrhage occurred, leading to rapid haemodynamic deterioration with arterial hypotension and increasing requirements for fluid resuscitation and vasopressors. The clinical picture was consistent with acute haemorrhagic shock, prompting immediate transfusion support.

Packed red blood cells were transfused to restore intravascular volume and oxygen-carrying capacity. In addition, acquired coagulopathy related to massive bleeding was treated with targeted fibrinogen administration, resulting in progressive improvement in haemostasis.

Because of the invasive nature of the mass, its intimate vascular adhesions, and the occluded ipsilateral renal artery, complete excision ultimately required adrenalectomy associated with total nephrectomy in order to achieve safe oncologic resection while limiting vascular complications.

Postoperative Outcome

At the end of surgery, the patient was transferred to the intensive care unit for close postoperative surveillance. Extubation was performed under safe conditions after haemodynamic stabilization. The postoperative course in the intensive care unit was favourable, with no recurrence of major haemodynamic instability.

Analgesia was effective, notably through epidural analgesia maintained for 48 hours. In the absence of postoperative complications, the patient was transferred back to the originating ward for continued care.

Discussion

Pheochromocytoma is uncommon in paediatric practice, but it carries major anaesthetic risk because catecholamine release is often excessive and unpredictable. Management therefore relies on rigorous preoperative preparation, anticipatory anaesthetic strategy, and close coordination between anaesthesiologists, surgeons, and intensivists.

In the present observation, the clinical context of severe hypertension with headache, sweating, and palpitations was highly suggestive of pheochromocytoma. However, the particular complexity of the case stemmed from the invasive character of the tumor, its abutment to the abdominal aorta, and the occlusion of the ipsilateral renal artery. These features explain why total nephrectomy had to be combined with adrenalectomy, a strategy reported only in rare comparable paediatric situations.

The literature emphasizes the importance of adequate alpha-adrenergic blockade before surgery, as this significantly reduces perioperative complications related to hypertensive surges. Despite appropriate preparation, important blood pressure fluctuations frequently occur during tumor handling, followed by severe hypotension after venous ligation and tumor removal. In our patient, these expected phenomena were observed and controlled with short-acting vasoactive agents in line with current practice.

Another noteworthy aspect was the occurrence of major intraoperative haemorrhage and haemorrhagic shock, complications only rarely highlighted in paediatric pheochromocytoma surgery. In this case, extensive vascular involvement and difficult dissection near the aorta probably increased bleeding risk. Early transfusion therapy combined with targeted fibrinogen supplementation enabled restoration of haemodynamic stability and haemostasis.

Thoraco-lumbar epidural analgesia was also of interest. Although epidural techniques may be debated in pheochromocytoma because of vasodilatory effects, careful titration can improve analgesia, reduce opioid exposure, and contribute to perioperative stability, as observed here.

Finally, the favourable postoperative course, with intensive care monitoring, safe extubation, effective analgesia, and rapid return to the referring ward, underlines the benefit of a structured multidisciplinary approach in such rare high-risk surgery.

Conclusion

Anaesthetic management of pheochromocytoma in children remains a major challenge, especially when the tumor is invasive and closely related to major vascular structures. This case emphasizes the importance of meticulous preoperative preparation, invasive monitoring, and anticipation of both haemodynamic and haemorrhagic events. A multidisciplinary strategy involving anaesthesia, surgery, and intensive care is essential to achieve a favourable outcome in this rare and high-risk setting.

Learning Points

- Paediatric pheochromocytoma should be considered a high-risk anaesthetic condition because of unpredictable catecholamine surges.
- Preoperative alpha-adrenergic blockade and volume optimization remain essential before tumor resection.
- Invasive haemodynamic monitoring and immediate access to short-acting vasoactive agents and blood products are crucial during surgery.
- Invasive tumors adherent to major vessels may expose to massive bleeding and can necessitate extended resection such as nephrectomy.
- Structured multidisciplinary management is key to favourable postoperative recovery.

Declarations

Funding: None.

Conflicts of interest: The authors declare no conflicts of interest.

Ethics approval: Institutional requirements should be adapted according to local policy.

Consent for publication: Written informed consent from the patient's legal guardian should be confirmed before submission.

Figure Legends

Figure 1. Contrast-enhanced abdominopelvic CT scan showing a large heterogeneous hypervascular adrenal mass closely related to the abdominal aorta and associated with ipsilateral renal artery occlusion.

Figure 2. Intraoperative view showing difficult dissection of the invasive adrenal tumor in close contact with major vascular structures.

Figure 3. Surgical specimen after adrenalectomy associated with total nephrectomy.

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