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RESEARCH ARTICLE

INVASIVE ADRENAL PHEOCHROMOCYTOMA IN A CHILD REQUIRING TOTAL NEPHRECTOMY: ANAESTHETIC CHALLENGES

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Abstract

Pheochromocytoma is a rare catecholamine-secreting neuroendocrine tumor in children and is associated with major perioperative anaesthetic risk. We report a pediatric case of invasive adrenal pheochromocy toma associated with ipsilateral renal artery occlusion and requiring adrenalec tomy with total nephrectomy.The patient was a 13- year-old girl referred for surgical management of a large hypervascular adrenal mass closely related to the abdominal aorta. Preoperative management included alpha-adrenergic blockade and volume optimization before surgery. General anaesthesia was performed with invasive arterial pressure monitoring, central venous access, careful titration of induction, and thoraco-lumbar epidural analgesia placed before induction under full monitoring. Tumor manipulation was associated with major blood pressure variability requiring nicardipine. After tumor devascularization and removal, hypotension required norepinephrine support. Surgical dissection was complicated by major bleeding and haemorrhagic shock, treated with rapid transfusion and fibrinogen supplementation for acquired coagulopathy. Because of vascular involvement and renal artery occlusion, complete resection required adrenalectomy associated with total nephrectomy. The patient was transferred to the intensive care unit, stabilized, extubated safely, and subsequently transferred back to the referring ward without early major postoperative complications. This case highlights the need for multidisciplinary planning, preoperative adrenergic blockade, invasive monitoring, anticipation of both hypertensive and hypotensive crises, and preparedness for massive bleeding during pediatric pheochromocytoma surgery

Introduction:-

Pheochromocytoma and paraganglioma are rare neuroendocrine tumors arising from chromaffin tissue. Although most cases are reported in adults, pediatric cases require particular attention because childhood and adolescent pheochromocytoma/paraganglioma may be associated with hereditary predisposition, multifocality, and long-term recurrence risk. Excess catecholamine release may produce hypertension, headache, palpitations, diaphoresis, and life-threatening perioperative cardiovascular instability. Surgical resection remains the definitive treatment, but perioperative management is complex because tumor manipulation may trigger abrupt hypertensive crises, whereas venous ligation and tumor removal may be followed by vasoplegia, hypovolemia, and severe hypotension.

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International recommendations emphasize biochemical confirmation, appropriate imaging, preoperative alpha-adrenergic blockade for functional tumors, liberal salt and fluid intake when appropriate, and multidisciplinary management. These principles are particularly important in children with large invasive tumors or tumors closely related to major vascular structures. We report a rare pediatric case of invasive adrenal pheochromocytoma associated with ipsilateral renal artery occlusion, requiring adrenalectomy with total nephrectomy, and discuss the anaesthetic and haemostatic challenges.

Case Presentation:-

A 13-year-old girl was referred for surgical management of an adrenal pheochromocytoma. The exact body weight was not available in the anonymized submission file. The clinical history was suggestive of catecholamine excess, with severe arterial hypertension associated with headache, sweating, and palpitations. Preoperative cardiovascular assessment did not reveal overt heart failure. Baseline renal function was assessed before surgery because imaging showed ipsilateral renal artery occlusion. Contrast-enhanced abdominopelvic computed tomography demonstrated a large heterogeneous hypervascular adrenal mass with intense enhancement after contrast administration. The lesion was closely related to major vascular structures, particularly the abdominal aorta, and was associated with occlusion of the ipsilateral renal artery, indicating major vascular involvement and explaining the need to anticipate difficult dissection, renal devascularization, and significant haemorrhagic risk. Because the kidney was vascularly compromised and safe tumor clearance was considered difficult without en bloc resection, adrenalectomy associated with total nephrectomy was planned after multidisciplinary discussion involving pediatric surgery, anaesthesia, intensive care, radiology, and oncology/endocrinology.

Preoperative Preparation:-

Preoperative antihypertensive preparation was performed with alpha-adrenergic blockade and volume optimization. The aim was to reduce the frequency and severity of catecholamine-related hypertensive episodes and to limit post-resection hypotension. Beta-blockade was avoided before adequate alpha-blockade. Blood pressure control was considered acceptable before surgery, without clinical signs of uncontrolled heart failure or persistent hypovolemia. Because the submitted anonymized file did not contain the exact alpha-blocker name, dose, or duration, these details were not invented in the revised manuscript. If available in the medical record, they should be added by the authors before resubmission.

Anaesthetic Management:-

The anticipated anaesthetic difficulties were catecholamine surges during laryngoscopy and tumor manipulation, post-resection vasoplegia, major bleeding related to vascular adhesions, and postoperative intensive care requirements. Standard monitoring was supplemented by invasive arterial blood pressure monitoring before induction, central venous access for vasoactive drugs and transfusion, urinary catheterization, temperature monitoring, and serial blood gas/laboratory assessment according to intraoperative evolution. Thoraco-lumbar epidural analgesia was inserted before induction under full monitoring and after verification that there was no contraindication to neuraxial blockade. General anaesthesia was induced gradually to avoid sympathetic stimulation and maintained with adequate anaesthetic depth, controlled ventilation, careful fluid administration, and readiness for rapid vasoactive treatment. Hypertension during tumor handling was treated mainly with nicardipine titration. After tumor devascularization and removal, hypotension was treated with fluid resuscitation and norepinephrine support. This biphasic haemodynamic profile is typical of functional pheochromocytoma surgery and requires continuous anticipation rather than delayed reaction.

Intraoperative Course:-

Surgical dissection was difficult because of the close relationship between the tumor and major vascular structures, especially the abdominal aorta, and because of ipsilateral renal artery occlusion. During dissection, abundant bleeding occurred, followed by haemodynamic deterioration compatible with haemorrhagic shock. Management included rapid volume resuscitation, packed red blood cell transfusion, correction of anaemia and coagulopathy, and fibrinogen supplementation. Point-of-care viscoelastic haemostatic testing was not documented as available in the submitted file; therefore, the indication for fibrinogen was described as based on the clinical context of major bleeding and suspected acquired coagulopathy. Where available, ROTEM or TEG can help guide targeted haemostatic therapy in major bleeding. Complete surgical management required adrenalectomy associated with total nephrectomy. The rationale for nephrectomy was the combination of ipsilateral renal artery occlusion, vascular compromise of the kidney, and the need for safe en bloc resection of an invasive adrenal tumor. No intraoperative arrhythmia or cardiac arrest was reported in the submitted anonymized information.

Postoperative Outcome:-
At the end of surgery, the patient was transferred to the intensive care unit for close monitoring. Extubation was performed after haemodynamic stabilization. Early postoperative management focused on detection of recurrent hypotension or hypertension, pain control, renal function after nephrectomy, urine output, haemoglobin, coagulation parameters, and signs of bleeding. Epidural analgesia provided effective postoperative analgesia and was maintained for 48 hours. The early postoperative course was favourable, with no recurrent major haemodynamic instability reported, and the patient was transferred back to the referring ward for continued care. Long-term oncologic and endocrine follow-up was recommended because pediatric pheochromocytoma/paraganglioma may be associated with hereditary disease and recurrence risk.

Histopathology:-
Histopathological examination confirmed adrenal pheochromocytoma. Immunohistochemistry details, proliferative index, PASS/GAPP score, capsular or vascular invasion status, and genetic testing results were not available in the anonymized submission file. The absence of these details is a limitation. For a final publication version, the authors should add the pathology report if accessible, including tumor size, margin status, Ki-67 index, chromogranin A, synaptophysin, S100 sustentacular staining, and any available malignancy risk assessment.

Discussion:-
This case illustrates several anaesthetic challenges specific to pediatric pheochromocytoma surgery. First, functional catecholamine-secreting tumors require preoperative alpha-adrenergic blockade and volume optimization to reduce perioperative cardiovascular instability. International guidance recommends preoperative blockade for hormonally functional pheochromocytoma/paraganglioma and careful multidisciplinary management. Invasive arterial pressure monitoring and immediate access to short-acting vasoactive drugs are essential, particularly in children with large tumors. Second, the case was unusual because of vascular involvement and ipsilateral renal artery occlusion. The requirement for nephrectomy in pediatric pheochromocytoma is uncommon and increases the educational value of the case. The renal artery occlusion suggested a compromised kidney and made en bloc surgical resection more likely. This anatomical pattern also increased the risk of difficult dissection and major bleeding. Third, the haemodynamic profile was typical but severe: hypertension during tumor manipulation followed by hypotension after devascularization and tumor removal.

Nicardipine was appropriate for short-acting control of hypertension, while norepinephrine was used after resection to counter vasoplegia and hypovolemia. Careful titration is important because both undertreatment and overtreatment can be harmful in children. Fourth, major bleeding was a central feature of the case. Invasive adrenal tumors abutting major vessels can cause haemorrhagic shock during dissection. Modern perioperative bleeding management favors structured protocols, early recognition of coagulopathy, rational transfusion, and, when available, viscoelastic testing such as ROTEM or TEG to guide fibrinogen, plasma, platelet, and factor therapy. In this case, fibrinogen was administered for acquired coagulopathy in the context of massive bleeding; however, absence of documented fibrinogen level or ROTEM/TEG data limits the precision of interpretation. Finally, this report underlines the importance of postoperative intensive care. After pheochromocytoma resection, patients may develop hypotension, rebound hypertension, hypoglycemia, bleeding, renal impairment, and pain- related sympathetic activation. ICU monitoring allowed safe extubation, haemodynamic observation, continuation of epidural analgesia, and early detection of complications.

Limitations:-
This report is limited by the absence of several patient-specific details in the anonymized submission material, including exact body weight, complete biochemical values, detailed alpha-blocker dosing, precise intraoperative blood loss, exact transfusion volumes, viscoelastic coagulation testing, and complete pathology/immunohistochemist ry. These omissions were specifically addressed by avoiding unsupported numerical claims. The manuscript would be strengthened substantially if the treating team added verified data from the medical record before final resubmission.

Conclusion:-
Pediatric invasive adrenal pheochromocytoma with renal artery occlusion requiring nephrectomy is rare and represents a high-risk anaesthetic scenario. Management requires preoperative adrenergic preparation, invasive haemodynamic monitoring, rapid access to antihypertensive and vasopressor therapy, preparedness for major

haemorrhage, and postoperative intensive care. The novelty of this case lies in the combination of pediatric pheochromocytoma, major vascular involvement, ipsilateral renal artery occlusion, nephrectomy, catecholamine-related haemodynamic instability, and haemorrhagic shock.

Learning Points:-

- Pediatric pheochromocytoma surgery requires multidisciplinary planning and preoperative alpha-adrenergic preparation.
- Invasive tumors close to the aorta or associated with renal artery occlusion carry a high risk of difficult dissection and major bleeding.
- Continuous invasive arterial pressure monitoring and short-acting vasoactive drugs are essential.
- Post-resection hypotension should be anticipated and treated rapidly with fluid resuscitation and vasopressors when needed.
- Major bleeding should be managed with structured transfusion and coagulation support; ROTEM/TEG is useful when available.
- Postoperative ICU monitoring is important after high-risk pediatric pheochromocytoma resection.

Declarations:-

Ethics approval: According to the authors, formal ethics committee approval was not required by local institutional policy for this anonymized single-patient case report.

Consent for publication: No patient photographs, radiologic images, operative images, or identifying details are included in this revised version. According to the authors, written parental consent for publication was not required under local policy in the absence of identifiable material. Patient confidentiality has been preserved.

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