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Paraganglioma behaving like a Pheochromocytoma? -A rare intraoperative finding

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I. Introduction

Paragangliomas are catecholamine-secreting neuroendocrine tumors that originate from the chromaffin cells of the sympathetic ganglia. Approximately 10% of paragangliomas maybe malignant, resulting in a rare occurrence of 90-95 cases per 400 million people.1 Paraganglioma is a type of neuroendocrine tumor that forms near certain blood vessels and nerves outside of the adrenal glands. These tumors can also be called extra-adrenal pheochromocytomas.

PGs share an almost identical profile with PCs, with the anatomical location of the tumors, PGs being extra-adrenal tumors and PCs growing inside the adrenal gland, being the only differentiation between the two[2]. Approximately 35-50% of paragangliomas may spread to other parts of the body.

II. Case Report

Herein, we report a case of a 16-year-old female patient who presented with nausea, vomiting, pain abdomen and bloating since last few months and was found to have a large left retroperitoneal tumor upon imaging. We shall discuss about how and what all happened just as the surgeons explored and reached the tumor and started manipulating it. The blood pressures started to rise and then fall dramatically and there were many other intra operative vital fluctuations noted as the case went through. Finally the tumor was successfully removed, and subsequent histological analysis was compatible with the presence of a paraganglioma.

The diagnostical findings and presentation were very much similar to that of a Pheochromocytoma but this case serves as a reminder that despite its rarity, Pheochromocytoma should never be dismissed as a differential diagnosis if correlating symptoms and diagnostic findings are consistent with that of paraganglioma etiology. Rare thing about this case that we shall be discussing later in our report is that how this Paraganglioma behaved and touted as a Pheochromocytoma intra-operatively.

III. Conclusion

Paraganglioma diagnosis, peri-operative anaesthesia management and surgical resection is a tough challenge for the anesthesiologists, this clinical case may prove to be a thoughtful experience for anesthetic management in the successful resection of giant paragangliomas as well as their intra operative managment.

Introduction

- Paragangliomas are catecholamine-secreting neuroendocrine tumors that originate from the chromaffin cells of the sympathetic ganglia.
- Approximately 10% of paragangliomas maybe malignant, resulting in a rare occurrence of 90-95 cases per 400 million people
- Paraganglioma is a type of neuroendocrine tumor that forms near certain blood vessels and nerves outside of the adrenal glands.
- These tumors can also be called extra-adrenal pheochromocytomas.
- Paraganglioma share an almost identical profile with Pheochromocytoma,with the anatomical location of the tumors, PGs being extra-adrenal tumors and PCs growing inside the adrenal gland, being the only differentiation between the two.
- Approximately 35-50% of paragangliomas may spread to other parts of the body.

Case report

Herein, we report a case of a 16-year-old female patient who presented with nausea, vomiting, pain abdomen and bloating since last few months and was found to have a large left retroperitoneal mass upon imaging. The pre-anaesthetic check up revealed no significant findings that would raise a suspicion of this case to be a pheochromocytoma but for any chances of incidental findings all possible preparation were done.

We shall discuss about how and what all happened just as the surgeons explored and reached the tumour and started manipulating it.
An epidural followed by general anesthesia was planned for this case. Arterial line, central line (IJV), peripheral line were accessed under all aseptic precaution.
Sometime after induction when the surgeons started manipulating the tumour the blood pressures started to raise significantly and the maxiumum value noted was 257/141 mmhg raising doubts and making it more challenging task for the team to manage as it started to micmick as a pheochromocytoma. There were many fluctuations noted as the case went through which was well managed by our team using IV dexmeditomedine, Nitroglycerine, beta blockers, di-uretics and other drugs in divided doses. As soon as the tumour was resected the blood pressures fell down significantly for which inotropes had to be started.
Seeing the extensive duration of surgery and significant intra operative events the patient was later shifted to the ICU on inotropes for further management and observation.
Vitals after shifting the patient to the ICU:
IBP 73/46mmhg (on inotropes) , PR 90/min, SPo2 100% on mechanical ventilation.
Patient was handed over to the ICU/Critical care team and regular follow up was done by the team anaesthesia.
On 2/9/23 that is just about 48 hours post surgery patient had an episode of monomorphic ventricular tachycardia which was noted by the ICU team and urgent DC shock was delivered and the patient again regained normal sinus rhythm. Multiple PVC's were noted post VT, a bedside 2D Echo and cardiology reference were done which was normal and stated no urgent intervention respectively.
Regular follow up was done by the team anaesthesia and critical care and the patient was weaned off and got extubated on 4/9/23.

Patient was then kept on 100% oxygen support and was kept in the ICU for observation. After 24 hours of extubation the patient was shifted to the surgery ward after consulting the primary surgeon and his team.
The post op FNAC/biopsy sample of the specimen showed the diagnosis of Paranganglioma.

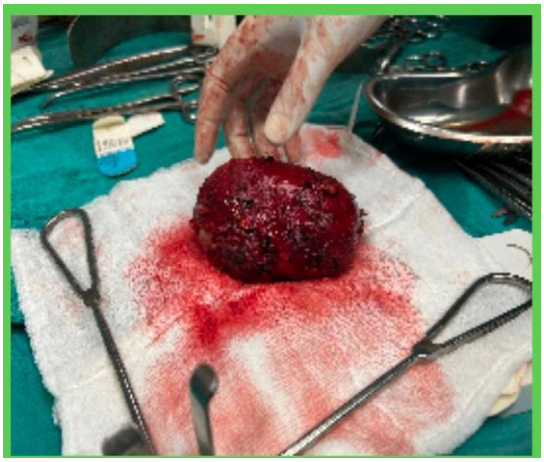
The diagnostical findings and presentation were very much similar to that of a Pheochromocytoma but this case serves as a reminder that despite its rarity, Pheochromocytoma should never be dismissed as a differential diagnosis if correlating symptoms and diagnostic findings are consistent with that of paraganglioma etiology.
Finally the tumor was successfully removed, and subsequent histological analysis was compatible with the presence of a paraganglioma.

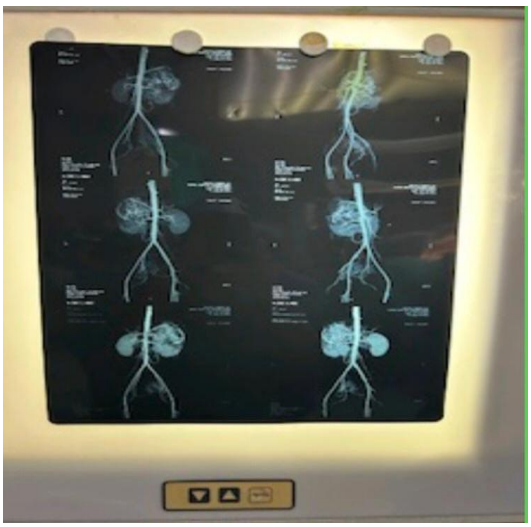
IV. Discussion

As we all know that Paraganlioma or Pheochromocytoma in itself is a big challenge for the Anaesthesiologist when it comes to managing the intraoperative events and in this case as per the physician's reference it was dismissed as innocent non-secreting paraganglioma as urinary VMA and metanephrines were near upper normal.
Subsequently she wasn't prescribed any preoperative alpha or beta blockers

V. Results

Patient was well managed intra operatively and then later in the ICU. Patient was successfully weaned off and was extubated after post operative day 2 and was later taken off from all inotropes support and was then shifted to the general surgery ward from where the patient got discharged on advice.





VI. Conclusion

- Paraganglioma diagnosis, peri-operative anaesthesia management and surgical resection is a tough challenge for the anesthesiologists.
- Pheochromocytoma should never be dismissed as a differential diagnosis if correlating symptoms and diagnostic findings are consistent with that of paraganglioma etiology.
- This clinical case may prove to be a thoughtful experience for anesthetic management in the successful resection of giant paragangliomas as well as their intra operative managment.

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