Abdominal Mass Suspicious for Pheochromocytoma

Dung D. Nguyen, M.D., Aru Reddy, M.B.B.S., Edwin A. Bowe, M.D.

University of Kentucky Department of Anesthesiology, Lexington, KY

Introduction

Our patient was a 16 year old, 56 kg female who presented with right flank pain. She was previously healthy until four months prior, when she developed intermittent right flank pain associated with hematuria. When initially seen in the emergency department at another hospital she was diagnosed with a urinary tract infection. Due to ongoing flank pain, she was referred to our facility. Evaluation showed a 15cm x10cm x8cm right adrenal mass which was highly concerning for a pheochromocytoma.

Work-up revealed elevated plasma free normetanephrines of 13 (normal <0.9) and urine normetanephrines of 2441 (normal 0-278). Urine VMA and HMA were also elevated at 61(normal 0-9) and 31 (normal 0-15). MIBG scan demonstrated a large MIBG-avid right adrenal mass, consistent with a neurogenic tumor. Even though her labs and imaging were diagnostic of a pheochromocytoma, she did not have the classic clinical picture of episodic palpitations, diaphoresis and headaches. She was scheduled for surgical resection of the right adrenal mass and started on pre-operative alpha and beta blockade.

Intra-operative Management

Patient received two weeks of alpha blockade prior to surgery for resection of a pheochromocytoma. Even prior to alpha blockade, outpatient 24 hour ambulatory blood pressure monitoring showed no systolic blood pressure over 120 mmHg. Pre-induction blood pressure was 123/65 with a heart rate of 80. Anesthesia was induced with propofol, lidocaine, fentanyl and rocuronium. Arterial and central venous catheters were placed. Sevoflurane and an infusion of remifentanil were used for maintenance. Vasoactive infusions were prepared. Tumor manipulation during resection did not result in hypertension as would be expected with a pheochromocytoma. Neither did the patient exhibit hypotension following interruption of venous drainage from the tumor.

The most challenging part of the seven hour case was replacing intraoperative blood loss, which was approximately 3.2 liters. The surgical procedure included a radical right adrenalectomy with en bloc resection of kidney and ureter.(Figure 1).

Discussion/Conclusion

Pathology demonstrated a ganglio-neuroblastoma, a very rare, more poorly differentiated, catecholamine producing tumor usually found in children. The annual incidence is reported to be 7.6 per 1 million in the USA. It occurs equally in both genders and most commonly in babies and in young children, with occurrence after 10 years of age being extremely rare.

Ganglio-neuroblastomas have intermediate malignant potential, between that of neuroblastomas and ganglioneuromas and are composed of mature ganglion cells and primitive neuroblasts.

Patients usually present clinically with pain caused by the primary tumor or by metastatic disease. If it metastasizes, it is usually to bone. Complete excision combined with radiation and chemotherapy remains the mainstay of therapy. Even though these tumors produce catecholamines, they are usually not associated with hemodynamic consequences. However, as an anesthesia provider for resection of an undiagnosed adrenal mass with elevated catecholamines, proper precautions should always be exercised.

References

- Young et al. Clinical Presentation and diagnosis of pheochromocytoma. UpToDate
- Fatimi et al. Ganglioneuroblastoma of the posterior mediastinum: a case report Journal of Medical Case Reports 2011, 5:322