Unusual Suspect: Diarrhea and Shock Secondary to Pheochromocytoma and VIPoma

Douen, A[1]., Siu, W.[2,] Kaldas, F[1]., Wagner, J.[1,] Tan, S.[3,] ,Choudhry, M. [1], & Trillo, J [1]
1. Department of Internal Medicine, NYCHHC Coney Island Hospital, Brooklyn, USA; 2. St George's University, Grenada, Weill Cornell Medicine. 3. Department of Internal Medicine, Mount Sinai Beth Israel, New York, USA

INTRODUCTION

- Pheochromocytoma is a catecholamine secreting tumor that is found at an incidence of 2-8 per 1 million people[1] annually in the general population.
- Pheochromocytomas classically present with episodic headache, diaphoresis, and tachycardia.
- Vasoactive intestinal peptide(VIP) is a peptide hormone that enhances cardiac contractility, produce vasodilation, increases glycogenolysis, reduces arterial blood pressure, and relaxes the smooth muscle of the trachea, stomach, and gallbladder[2].
- VIP may lead to the secretion of water and electrolytes, hypokalemia, and flushing[2].
- This case presentation involved a patient with an atypical pheochromocytoma, suspected of also producing VIP.

CASE PRESENTATION

A 51-year-old male presented with episodic palpitations, diaphoresis, nausea, and intermittent substernal chest pain that exacerbated when he lied on his right side. Hypertension and left adrenal mass are among his medical history. During his admission, blood pressure fluctuated between 89-272 systolic and 42-152 diastolic. Vasopressors and IV labetalol were given for blood pressure management

He developed stomach pain, distention, and profuse secretory diarrhea. On day four of his hospital stay, this resulted in hypovolemic shock due to a 5L/day output. He further developed metabolic acidosis, hypokalemia, and his EKG revealed intermittent ventricular tachycardia and T wave inversion.

Endocrine evaluation revealed plasma metanephrines of 5679 pg/mL, VIP was 239 pg/mL, and gastrin was 313 pg/mL (normal ranges: metanephrines 12-60 pg/mL, VIP <70 pg/mL, and gastrin <100 pg/mL)

CT scan revealed a 5 cm adrenal mass in the gastro-entero-pancreatic area; pheochromocytoma was then confirmed with the positive metanephrines.

Patient finally eventually underwent an adrenalectomy and his symptoms resolved.

DISCUSSION

- Pheochromocytoma is a rarely identified and diagnosed tumor as in the majority of patient as up to 50% of patients are asymptomatic.[1]
- The classical presentation of pheochromocytoma is typically an episodic headache, diaphoresis, and tachycardia.
- Classic Symptoms of VIP secretion include watery diarrhea, hypokalemia and achlorhydria (WDHA) [2]
- VIP tumors are found in the pancreases 80% of the time[3]. But in our patient there had only been an adrenal mass making it the most likely source of VIP.
- On rare occasions, these tumors may also VIP. VIP can result in the relaxation of smooth muscles of the GI tract along with peripheral vasodilation and hypovolemia explaining this patients symptoms.
- This patient presented with atypical pheochromocytoma, suspected of secreting VIP. Due to the VIP secretion, the patient developed copious secretory diarrhea, leading to hypovolemia and in turn leading to hypokalemia. The hypokalemia resulted in the development of Ventricular tachycardia.

CONCLUSION

- This presentation leads to the conclusion that it is imperative to test metanephrines and image catecholamine-secreting-tumors if suspected in a patient.
- Pancreatic VIPomas must also be ruled out as elevated VIP levels may be due to pheochromocytomas and be detrimental to the patient's health.

Contact

Wenchy Siu
St. George's University / Weill Cornell School of Medicine
Email: wys4001@med.cornell.edu
Website: www.linkedin.com/in/wenchy-siu

References

1. *Pheochromocytoma and Paraganglioma Treatment (PDQ®)–Health Professional Version - NCI*. 22 Oct. 2010, https://www.cancer.gov/types/pheochromocytoma/hp/pheochromocytoma-treatment-pdq. nciglobal,ncienterprise.
2. Abdullayeva, Leila. "VIPoma: mechanisms, clinical presentation, diagnosis and treatment." World Academy of Sciences Journal 1.5 (2019): 229-235.
3. Reisch, Nicole, et al. "Pheochromocytoma: presentation, diagnosis and treatment." *Journal of hypertension* 24.12 (2006): 2331-2339.
4. Onozawa, Masahiro, et al. "Hypokalemic rhabdomyolysis due to WDHA syndrome caused by VIP-producing composite pheochromocytoma: a case in neurofibromatosis type 1." *Japanese journal of clinical oncology* 35.9 (2005): 559-563