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SP 6. AN UNEXPECTED SECOND PHEOCHROMOCYTOMA IN PREGNANCY

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Background: Pheochromocytoma in pregnancy is rare with reported maternal and fetal mortality rates of 8% and 17%.

Methods: Herein, we report a recurrent pheochromocytoma presenting in pregnancy and review the presentation, the multidisciplinary medical management and surgical management of the patient.

Results: A 30 year-old woman G2P0 with Multiple Endocrine Neoplasia type 2A secondary to a C634R mutation in the RET proto-oncogene presented for a routine prenatal visit at 15 weeks of pregnancy. She reported mild headache, nausea, and anxiety that were attributed to pregnancy. One year prior to her visit she had a laparoscopic left adrenalectomy for multiple pheochromocytomas. Her plasma metanephrines postoperatively were normal. She has also had a total thyroidectomy with central neck dissection for medullary thyroid cancer and a total parathyroidectomy

with parathyroid autotransplantation for hyperparathyroidism nine years prior. Her BP was 130/80 and HR 90. Her obstetrician ordered screening plasma metanephrine and normetanephrine, which were 79 (< 57pg/ml) and 209 (< 147pg/ml). A follow-up 24-hour urine metanephrine and normetanephrine were 448 (36-190mcg/24 hr.) and 663 (35-482mcg/24hr.). MRI revealed a 1.5 cm nodule with mild T2 hyperintensity in the right adrenal gland, which was not present on an MRI from 1-year prior. The patient was started on doxazosin and was hydrated. She underwent a laparoscopic right adrenalectomy at 17 weeks of pregnancy. Fetal heart tones were normal before and after surgery. Pathology demonstrated 3 pheochromocytomas (1.5 cm, 1.5 cm, .7 cm) in the right adrenal gland. She was started on fluorinef and hydrocortisone and was

discharged on postoperative day 1. Her plasma metanephrines were normal at her 3-week follow up visit.

Conclusion: The C634R mutation in the RET proto-oncogene is a high-risk mutation associated with a higher penetrance and younger age of presentation for pheochromocytoma, which can be multiple, bilateral and extradrenal. This is especially important in patients contemplating pregnancy because of the high risk of maternal and fetal mortality associated with pheochromocytoma in pregnancy.