CLINICAL VIGNETTE

Paraganglioma

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A 58-year-old female presented to a local emergency room with upper abdominal pain and nausea with emesis. She was afebrile and had normal blood pressure, with heart rate of 90 beats/ minute. Initial blood work was normal except mildly elevated amylase at 102 units/ L (Normal 20-70). She had history of osteopenia and history of angioedema episodes in past possibly related to acetaminophen and ibuprofen. Her routine medications were aspirin at 81mg and Raloxifene. Family history was remarkable for colon cancer in mother.

Initial evaluation included ultrasound showing Cholelithiasis thickened gall bladder wall and peri cholic fluid. Murphy sign was positive. Subsequent HIDA scan was also consistent with acute cholecystitis and computerized tomography showed evidence of gall stones and acute cholecystitis. CT also noted a soft tissue heterogeneous 4 cm mass within the inferior root of the small bowel mesentery. She underwent gall bladder removal and concurrent resection of bluish mass at the mesenteric root of the mid to distal small bowel, which showed no evidence of gross invasion. Electrocautery was used to dissect the mass with 2-3 centimeter margins. The mass was retrieved into a separate endo retrieval bag. Her post-op course was uneventful. On pathology, the tumor was 4.5 cm tanned and hemorrhagic. It stained positive for synaptophysin and chromogranin and stained negative for S100, CD117, desmin, SMA, CD34 and AE1/AE3. Ki 67 was 1%, consistent with a paraganglioma.

Paragangliomas are uncommon neuroendocrine tumors from chromaffin cells from the embryonic neural crest. Abdominal Paragangliomas arise from sympathetic para ganglia. They generally arise at the junction of the renal veins at the inferior vena cava or near the origin of the inferior mesenteric artery from lower aorta. The majority of sympathetic Paragangliomas arise in abdomen. Catecholamine secretion has been reported in up to 85% of abdominal Paragangliomas so presentation can be similar to Pheochromocytoma.[1,2] They are usually benign but can have malignant behavior in up to 20% tumors. This is defined by presence of metastasis and not by invasion or histological criteria.[1-3] If there are often concurrent tumors present, the patient may have an inherited syndrome like MEN2 (multiple endocrine neoplasia), Neurofibromatosis type 1 or Von Hipple Lindau syndrome.

On imaging the location of the soft tissue mass and high degree of vascularity should raise suspicion. Besides CT scan and MRI, PET scans using FDG or I-123 MIBG as well as I-111 octreotide have been employed when malignancy or genetic syndromes are suspected.[4] Catecholamine secretion should be assessed even if there are no suggestive symptoms.[4,5] Patients can develop significant rise of blood pressure and related complications during surgery. If these complications occur during surgery of an unsuspected mass, the procedure should be stopped and the patient should undergo a full evaluation. Aggressive medical management including adrenergic blockade as well as proper monitoring during future surgery may be required.[4,5]

Preoperative biopsy is not recommended because of risk of bleeding and fibrosis may make subsequent surgical resection difficult.[3,4] Complete surgical en bloc resection without entry of tumor capsule with early division of outflow veins and inflow vessels should be performed.[4-6] Embolization, radiation, radio surgery have been used in difficult to resect Paragangliomas, usually of extra abdominal origin.[7,8]

Later recurrences have been noted. For nonfunctioning Paragangliomas, urine and blood tests for catecholamine as well as annual imaging are recommended initially and less frequently later. Biochemical monitoring is recommended lifelong.[4,5]

In this case diagnosis of Paraganglioma was not considered by radiologist or surgeon and the incidental mass was resected as unplanned surgery without evaluation. Fortunately her Paraganglioma did not secrete catecholamines and surgery was uneventful. Her post-op evaluation with whole body PET CT scan, as well as blood and urine evaluation for catecholamines were unremarkable.

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Submitted June 3, 2018