Case Based Urology Learning Program

Resident's Corner: *UROLOGY*

Case Number 15

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A 33-year-old woman presents with right sided upper abdominal pain and headache. Her blood pressure is 210/110 and difficult to control in the emergency room. She is stabilized and admitted for further evaluation. CT reveals a right upper quadrant mass that is definitely invading into the lower aspect of the liver. Serum epinephrine and norepinephrine are both well over 1000 pg/mL.

Should a biopsy be considered?
What is the most likely diagnosis?

Should a biopsy be considered? What is the most likely diagnosis?

The most likely diagnosis is malignant pheochromocytoma. A biopsy would be dangerous in this setting.

What percentage of pheochromocytomas are malignant?

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Approximately 10-20%.

What are the criteria for malignancy for pheochromocytoma?

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Similar to most endocrine tumors, malignancy is clearly established only by the demonstration of metastasis or local invasion into adjacent organs or grossly into the venous system. Histologic criteria to establish malignancy are not particularly reliable, so the role of biopsy is very limited for this type of neoplasm.

What is thought to be the best single test to establish a diagnosis of pheochromocytoma?

What is thought to be the best single test to establish a diagnosis of pheochromocytoma?

Plasma metanephrines have been shown to have a sensitivity of 99% and a specificity of 89% for this purpose. However, the accuracy of any endocrine test for pheochromocytoma may be lower in malignant cases because poorly differentiated tumors may downregulate catecholamine production. Dependent on tumor size and relative catabolic vs. metabolic activity, the accuracy of endocrine tests of blood or urine can vary in different circumstances. A combination of plasma metanephrines and urine metanephrines is a reasonable approach for most patients suspected of having a pheochromocytoma. An endocrine consult is always a reasonable consideration.

What are the classic imaging findings for pheochromocytoma, and what additional imaging should be considered for this patient?

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The classic finding is a “light bulb” image on the T2 phases of MRI.

For this patient with locally invasive, malignant pheochromocytoma a metastatic evaluation should be pursued, typically using the ^{131}I -MIBG scan which allows for whole body scanning.

The metastatic evaluation is otherwise negative.

What is the optimal treatment for this patient?

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Malignant pheochromocytoma is primarily a surgical disease, and the primary treatment is surgical excision with negative margins. In this case, an en bloc partial hepatectomy will be required.

What will be required to prepare this patient for surgery?

What will be required to prepare this patient for surgery?

Alpha blockade followed by beta blockade if necessary to further control the BP and pulse. Remember, always “alpha first”. Alternatively, calcium channel blockers can also be used and are equally effective.

Preoperative anesthesia consult should also be obtained, and preoperative hydration to restore blood volume should be pursued.

What are important intraoperative considerations for this patient?

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Anesthesia will obtain appropriate lines to monitor and correct blood pressure as needed, primarily using short acting agents such as nitroprusside, which allow for minute to minute correction of the blood pressure as needed

From a surgical standpoint, manipulation of the tumor should be minimized and the adrenal vein, and other venous outflows, should be clipped or ligated early in the procedure, after warning the anesthesiologist that this is going to occur, as it may be associated with a rapid fall in blood pressure.

For patients with metastatic pheochromocytoma with difficult to manage hypertension, what treatments can be considered?

For patients with metastatic pheochromocytoma with difficult to manage hypertension, what treatments can be considered?

Palliation is a top priority for many patients with metastatic pheochromocytoma due to the endocrinologic manifestations. For this surgical debulking with metastasectomy should be considered if substantial debulking can be achieved. Beyond this, radiopharmaceutical agents such as ^{131}I -MIBG can be considered and can palliate symptoms in some patients. Chemotherapy and radiation therapy can also be considered but have a limited role in this malignancy

What is the prognosis for patients with metastatic pheochromocytoma?

What is the prognosis for patients with metastatic pheochromocytoma?

Most series quote 5 year survival rates of approximately 30-50%. The disease tends to be indolent in some patients, but highly progressive in the majority.

How is surveillance performed?

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In addition to history, physical exam and conventional imaging, surveillance should include endocrine testing reflecting the profile at the time of presentation. PET and ^{131}I –MIBG can also be considered, although their role is not well defined.

Selected Reading

Novakovic K, Ersahin C, Picken M, Campbell SC: Malignant Adrenal Tumors, *AUA Update Series* 2004;23(30):242-7.

Topic:

Oncology: Adrenal Tumors

Subtopics:

Malignant Pheochromocytoma