Pheochromocytoma & Paraganglioma

- The adrenal glands are like “top hats” on the kidneys, their job is to produce different kinds of steroid hormones, especially cortisol & adrenaline.
    - These hormones are **necessary for life** because they regulate stress responses that control blood pressure and how our cells use glucose.
- Because we do so many CAT scans for other reasons, we often find nodules in glands “by accident” and these are called incidentalomas. Very rarely this nodule may be a pheochromocytoma.
- What is a pheochromocytoma?
    - This is a rare adrenal tumor (4-7% of adrenal tumors), and it is almost always benign. These tumors can unpredictably release large amounts of stress hormones. Patients may report spiking blood pressures, palpitations, anxiety attacks. Sometimes patients don’t have any symptoms at all.
        - When the tumor makes enough stress hormones, the adrenal gland on the other side “goes to sleep,” because there is already so much hormone around.
- How do I know if my adrenal tumor is a pheochromocytoma?
    - We screen every adrenal nodule for a pheochromocytoma, even if it doesn’t really “look” like one. This is usually done by collecting urine for 24 hours and checking it for compounds called metanephrines.
        - For certain cases, we may check a blood test instead of the urine test.
- I don’t have symptoms, so why should I even care about a pheochromocytoma?
    - Even when this kind of tumor doesn’t produce noticeable symptoms, it is taking its toll on your body. We want to remove these tumors to reduce your risk of long term heart problems or stroke.
- How is a paraganglioma different from a pheochromocytoma?
    - A paraganglioma is a “cousin” of a pheochromocytoma. They both can unpredictably make large amounts of hormones and they both cause long term problems if they are not removed. The only difference is that a paraganglioma isn’t in the adrenal gland, it’s found in certain nerves in the body.
- I have a diagnosis of pheochromocytoma or paraganglioma. Now what?
    - Now we get ready for surgery. The surgeon must be very experienced with taking out these specialized tumors, so frequently this will be done at a large university hospital. It’s a process that takes a long time:
        - Phase 1: We start you on medications called alpha-blockers to start getting you ready. This can take several days to several weeks.
        - Phase 2: Once you have gotten to the optimal dose of alpha blockers, you will start taking beta blockers as well. We use both drugs to prevent your body from reacting to the extra stress hormones this tumor makes.

        - Phase 3: When you are admitted to the hospital prior to your surgery, you will receive lots of IV fluids and a high salt diet. We want you to have strong blood pressures before you go to surgery.
        - Phase 4: the surgeon tries not to disturb the tumor too much while it is being removed, but sometimes the tumor can “panic” and flood the body with huge amounts of stress hormones and adrenaline. The anesthesia team will control your blood pressure and heart rate during the surgery.
        - Phase 5: After the tumor is removed, the body suddenly doesn’t have enough of the stress hormones or adrenaline, and blood pressure can drop. Usually patients need to recover in the intensive care unit, where they can get specialized medications to keep the blood pressure up while they wait for the other adrenal gland to “wake up.”
- What happens after surgery?
    - Patients can return to their normal life. Usually after everything is over, they feel much better, they may need fewer medications and some will say they didn’t realize how badly they felt until they discovered what “normal” felt like!
    - You will see your endocrinologist periodically to observe your recovery.

Resources

- Columbia Adrenal Center:
  http://columbiasurgery.org/conditions-and-treatments/pheochromocytoma
  http://columbiasurgery.org/conditions-and-treatments/paraganglioma
- Endocrine Surgeons:
  http://endocrinediseases.org/adrenal/pheochromocytoma_symptoms.shtml
  http://endocrinediseases.org/adrenal/paraganglioma.shtml
- Mayo Clinic www.mayoclinic.org
    - Benign adrenal tumors
    - Pheochromocytoma
    - Neuroendocrine tumors
    - Paraganglioma