"Doctor, does it look dangerous?"

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Bill, 40, presents with right anterior neck swelling, which has been present for the last eight months (Figure 1). The mass does not cause him any pain.

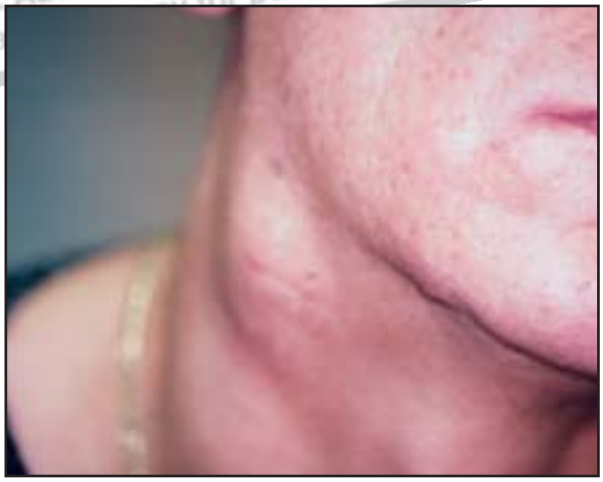

Figure 1. Patient with right anterior neck swelling.

Patient statistics

- Appears healthy
- Palpitations of the head and neck reveal a 5-cm, firm mass at the level of the thyroid cartilage; slightly tender and not compressible
- Cranial nerve exam is normal

Medical history

- Positive for hypertension
- Back surgery post-motor vehicle accident four years ago
- Taking atenolol for blood pressure, Tylenol®#3 and oxycodone for back pain
- Smokes one-half pack of cigarettes per day; has been doing so for 20 years
- Mother has history of breast cancer
- Bill has two healthy children

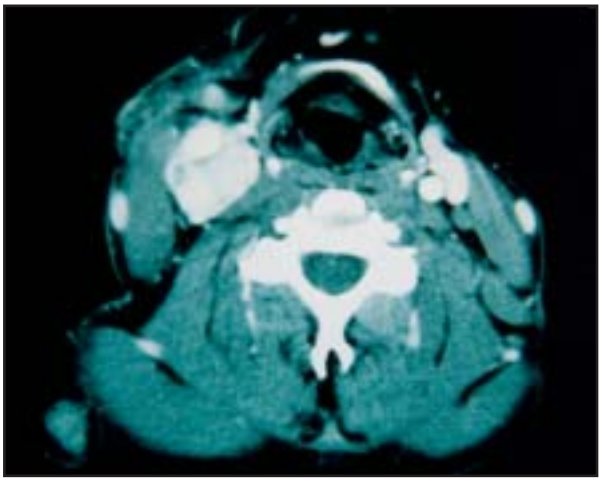

Figure 2. CT scan of the neck (infused).

What's your diagnosis?

a) Schwannoma
b) Carotid body tumour
c) Metastatic carcinoma
d) Lymphoma

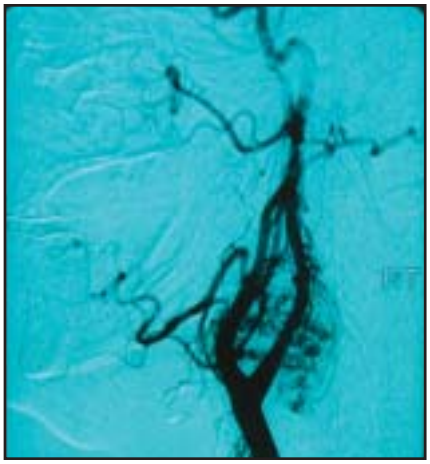

Figure 3. Carotid angiography.

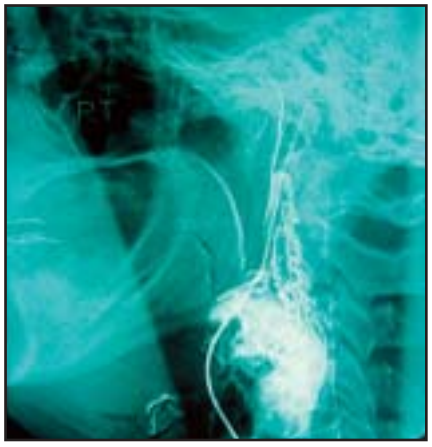

Figure 4. Carotid angiography.

Clinical investigations

- Blood investigations, including complete blood cell count, liver function tests, electrolytes, glucose, thyroid function tests and coagulation profile are normal
- 24-hour urine test for metanephrine is normal
- Excision biopsy for right cervical mass is discontinued because of brisk bleeding
- Computed tomography scan of the head and neck (infused) (Figure 2) shows a 3-cm, intensely enhancing mass in the right neck, centred at the carotid bifurcation and splaying the bifurcation
- The lesion is well-circumscribed
- Carotid angiography is performed (Figures 3 and 4)

Answer:
Carotid body tumour (CBT)

About CBTs

The most common nonadrenal paraganglioma to present as a neck mass at the carotid bifurcation is the CBT. Initially, CBTs appear as slow-growing, asymptomatic, palpable masses in the anterior triangle of the neck. Untreated, these tumours tend to surround the external and internal carotid arteries without any significant compression. Continued growth leads to cranial nerve involvement, pharyngeal compression and skull base and intracranial invasion.

The sympathetic chain and internal jugular vein may also be involved by a larger tumour. In addition to a painless mass, patients may present with:

- pain,
- hoarseness,
- dysphagia,
- Horner's syndrome,
- tongue paresis and
- vertigo.

Functional catecholamine secretory carotid body paragangliomas are rare and can produce paroxysmal hypertension, mimicking pheochromocytoma.

CBTs occur more frequently in adults between the ages of 45 and 50; they are uncommon in children. While most of these tumours are benign, 2% to 13% pursue a malignant course, with metastases to regional lymph nodes, lungs and bones.

What tests are needed?

Biopsy, including fine-needle aspiration, is unnecessary, dangerous and contraindicated in the evaluation of paragangliomas.

Ultrasonography, computed tomography and magnetic resonance imaging are very effective, noninvasive investigations. However, although it is an invasive test, carotid angiography is by far the most useful diagnostic test for paragangliomas. This modality can establish the diagnosis, demonstrate multiple lesions, determine the size and vascularity and evaluate the tumour blood supply.

While most CBTs are benign, 2% to 13% are malignant and metastasize to regional lymph nodes, lungs and bones.

Routine screening for urinary metanephrines, vanillylmandelic acid and serun catecholamines is probably only indicated for multiple or familial paragangliomas or in the presence of catecholamine-related symptoms.

What is the treatment?

The treatment of choice for most CBTs is surgical excision. However, because they are close to important vessels and nerves, there is a 3% to 9% risk of morbidity and mortality. Tumour size is important, as those > 5 cm in diameter have a markedly higher incidence of complications.

An extensive and complete preoperative workup is essential for safe resection. A vascular surgeon should be able to assist, if necessary. Perioperative alpha and beta andrenergic blockers should be given for all catecholamine-producing carotid paragangliomas. Some physicians recommend angiographic embolization preoperatively; others discourage this because it seems to be associated with inflammatory phase that makes the subadvential dissection more difficult.

Radiotherapy, either alone or in conjunction with surgery, is a second consideration and an area of some controversy. Historically, paragangliomas were considered radioresistant. Recent studies indicate good responses to supervoltage radiation, including one complete response. Most authors still recommend radiotherapy only for very large tumours, recurrent tumours or for patients who are poor surgical candidates. Dx

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