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Just epistaxis?

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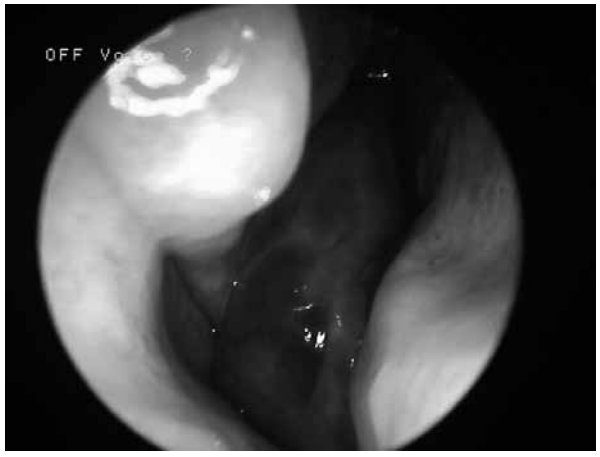
CASE REPORT

A 74-year-old woman with epistaxis at the right side of the nose was referred to the ENT department by the general practitioner. For six weeks she had experienced daily dropwise blood loss at the right side of the nose. She had no complaints of disturbed nasal breathing, rhinorrhoea, visual disturbances or headaches. She was taking anticoagulants for a cardiac condition. She had not suffered from epistaxis before.

Anterior rhinoscopy did not show any lacerations of the nasal mucosa in locus Kiesselbachi. Nasal endoscopy showed a smooth mass that almost completely obstructed the lumen in the right dorsal nasal cavity and nasopharynx. Small superficial veins were visible on the mass (*figure 1*). Palpation of the neck did not reveal any lymphoid nodules.

Figure 1. *Mass in dorsal part of nasal cavity and nasopharynx seen with nasal endoscopy. The mass is located in the choane. On the right side the nasal septum is visible. The left top shows the middle nasal turbinate*



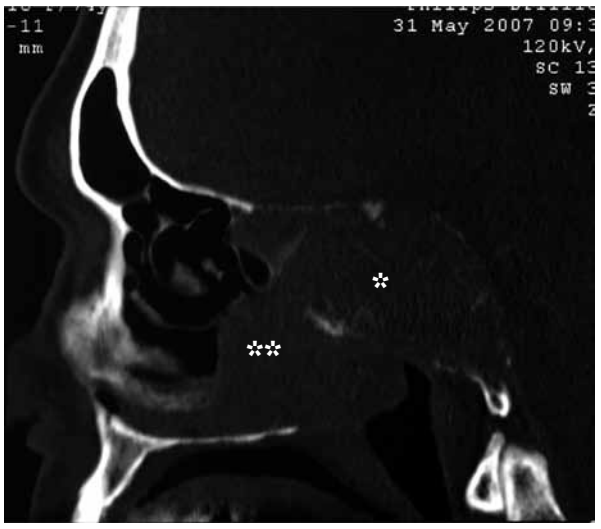
After cessation of the anticoagulants for a week a biopsy of the mass was taken under general anaesthesia. The resulting epistaxis was managed by tamponade of the right nasal cavity. Also magnetic resonance imaging (MRI) of the head was performed.

The scan showed a giant mass (>6 cm) destroying the sella region, invading the sphenoid sinus and nasopharynx. The mass was growing half way to the anterior skull base and passed the cerebral pons caudally. No compression of the optic chiasm was seen (*figure 2*).

WHAT IS YOUR DIAGNOSIS?

See page 230 for the answer to this photo quiz.

Figure 2. *Sagital CT scanning slide of the right nasal cavity showing a mass destroying the sella region, invading the sphenoid sinus and nasopharynx. *sphenoid sinus; **bulging of the mass in the nasal cavity and nasopharynx*



DIAGNOSIS

Histological examination of the biopsy showed pituitary adenoma. Accessory staining for prolactin was positive. Consultation with the endocrinologist revealed no symptoms related to hyperprolactinaemia or hypopituitarism. Hormonal function evaluation, however, showed extremely elevated serum prolactin levels: 697000 mU/l (normal: <500 mU/l) and a mild secondary hypothyroidism without any signs or symptoms (thyroid-stimulating hormone (TSH) 0.89 mU/l and free T4 9.8 pmol/l).

The patient was treated with a dopamine agonist; serum prolactin levels decreased to 4000 mU/l after six months. Control MRI was performed and showed shrinkage of the mass. She had no recurrence of epistaxis and her nasal airway was not obstructed.

Prolactinomas are pituitary adenomas that express and secrete prolactin (PRL) to variable degrees; they are almost invariably benign, but are nevertheless frequently clinically significant. Prolactinomas are generally classified according to size as microadenomas (less than 10 mm in diameter) or macroadenomas (more than 10 mm in diameter). Prolactinomas are called giant prolactinomas when they reach a size of >40 mm diameter and show invasive growth on neuroimaging.[1-3]

Epistaxis is a very rare primary presenting symptom of a giant prolactinoma. Ghannam *et al.*[4] described a case of a TSH-secreting pituitary adenoma first presenting with nasal bleeding. In 1985, Lessard *et al.*[5] described a unilateral intranasal extension of a pituitary adenoma. Epistaxis was not a presenting symptom.

The diagnosis of prolactin-producing pituitary adenoma is made on a combination of radiological (MRI) and histopathological findings and elevated prolactin levels. Symptoms of prolactinomas are closely related to hyperprolactinaemia (galactorrhoea and hypogonadism) and the location and size of the mass. In most cases mass effects are due to suprasellar extension with invasion of the optic chiasm which causes visual disturbance.

All patients with macroadenoma and most patients with microadenoma require treatment. Dopaminergic agonists such as bromocryptine and cabergoline are the first-line, preferred therapy. Transsphenoidal surgery is an option in individuals who cannot tolerate a dopamine agonist or in whom the drug is ineffective.[3,6] In macroadenomas medical treatment causes rapid shrinkage of the lesion and cessation of prolactin secretion.[5,6] Treatment of giant prolactinomas with dopamine agonists may result in cerebrospinal fluid liquorrhoea.[2]

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