Pituitary adenoma and orbital haemangioma: a case report

R. J. GALVIN AND B. E. KENDALL
From the Maida Vale Hospital, London, and the National Hospitals for Nervous Diseases, Queen Square, London

SUMMARY A 43-year-old woman with a pituitary tumour had unexplained right proptosis. Orbital x-rays and ultrasound were normal, but computed tomography showed an intracanal tumour, which had typical features of a cavernous haemangioma on carotid angiography. Both tumours were resected and confirmed histologically.

About 8% of most series of intracranial tumours are pituitary adenomata (Zülch, 1965), the majority of these being chromophobe. The most usual presentation is with bitemporal field defects and visual failure (Martins, 1974). Cavernous haemangiomas are the commonest type of orbital tumour (Reese, 1964). Their cardinal symptom is proptosis, but they may also produce papilloedema, optic atrophy, and diplopia (Duke-Elder, 1974). We report here the rare, and presumably chance, association of both these tumours in the same patient. Although clinically a difficult problem, the patient displayed classical radiological features of both these tumours. The value of computed tomography is stressed in this situation.

Case report

The patient, a 43-year-old woman, had been noted by her friends to have right proptosis 1 year before being referred to hospital. This had slowly progressed. Three months later she had developed slight intermittent diplopia, mainly on upgaze, with vertical displacement of the images. For 6 months she noticed a faint ‘haziness’ of vision bilaterally, more marked in her right eye.

She had had decreased libido and secondary amenorrhoea for 26 years after a normal menarche, and for many years she had tended to gain weight and suffered from cold intolerance. She had been unsuccessfully treated with ‘thyroid tablets’ 10 years ago. For several years she had worn a wig because of sparse capital hair.

She was initially assessed ophthalmologically and was referred to the Maida Vale Hospital after a normal orbital ultrasound study and with a skull x-ray showing an enlarged pituitary fossa.

Examination showed a moderately overweight, normotensive woman. Scalp hair was thin and dry, and pubic and axillary hair, though sparse, was of normal distribution. There was 5 mm of axial proptosis on the right (Fig. 1). Elevation of the right
eye was restricted to 50%, and this was more marked on adduction. There were no orbital bruits. Visual acuity corrected to 6/7.5 right and 6/6 left. There was hereditary deuteranomaly. Initial Goldmann and tangent screen fields were full, but repeat testing after 6 weeks revealed a subtle right acocentral defect and left upper temporal restriction. The right disc was slightly hyperaemic; there were choroidal folds super temporally on the right.

Investigations. The erythrocyte sedimentation rate was raised at 37 mm in 1 hour. The remaining haematological and biochemical indices, including plasma and urinary osmolalities, were normal. Endocrinological studies showed a euthyroid and eu cortisol state, and an insulin tolerance test was normal. There was marked elevation of serum prolactin to greater than 4900 mU/l (normal 0 to 360 mU/l). Fluorescein angiography confirmed the right disc oedema and choroidal folds (Fig. 2). Pattern visual evoked responses were bilaterally abnormal in wave form, more so on the right, and half field responses were consistent with early compression involving the decussating fibres at the chiasm.

Radiological investigations. Plain x-rays and tomodograms of the skull showed asymmetrical ballooning of the sella turcica typical of a pituitary tumour (Fig. 3). There was no calcification or other abnormality. In particular the orbits appeared normal on polytomography. Orbital B-scan ultrasound was normal. Computed tomography (Fig. 4) showed a mass of increased attenuation, without calcification, which enhanced homogeneously, filling the ballooned sella and suprasellar cisterns. There was an even dense, rounded, intraconal mass with clear-cut edges displacing the right globe and obviously causing the proptosis. In common with most orbital masses it enhanced considerably after intravenous contrast medium.

Right carotid angiography showed a diffuse capillary blush supplied by hypophyseal arteries outlining the pituitary and suprasellar tumour. There was no displacement of any vessels by this tumour. The superior ophtalmic vein was deviated and curved superiorly and was somewhat narrowed but not obstructed by the intraconal mass. Lakes of contrast filled within part of the tumour in the venous phase (Fig. 5).

Pneumoencephalography confirmed the outline of the suprasellar mass and showed the anterior recesses of the third ventricle to be elevated and splayed apart by it.

Operation. A right frontal craniotomy was performed for excision of the pituitary tumour (Mr L. Symon). The tumour proved difficult to detach from the undersurface of the chiasm, but it was excised fairly completely with its capsule. The
craniotomy was extended into the orbit, and a
black rounded mass $16 \times 10$ mm was found in the
lateral part of the orbit, running from behind the
globe almost to the apex of the orbit. It was easily
removed in its entirety. Postoperatively the patient
had a dense bitemporal hemianopia but with
macular sparing. She otherwise made an uneventful
recovery.

**Histology.** The tumours showed classical histo-
logical features of chromophobe adenoma and
cavernous haemangioma.

**Discussion**

The ballooned sella turcica and the capillary blush
supplied by hypophyseal arteries to a tumour
extending from the sella through the suprasellar
cisterns are the classical radiological features of a
pituitary adenoma. The computed tomographic
features are also those commonly found in pituitary
adenomas, 75% of which show increased attenu-
ation on computed tomography and most of
which show homogeneous enhancement after
intravenous contrast medium. Pneumoencephalo-
graphy, which is performed routinely in our unit

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**Fig. 4** Computed tomography before (above) and
after intravenous contrast medium (below). The scans were
taken at the level of the orbit and sella (window level
$= -6$, window width $= 100$) and suprasella (window
level $+ 18$, width $40$). There is a spherical intraconal
mass with clear-cut edges in the right orbit displacing the
globe anteriorly. It is of high attenuation and homogeneous
but without evidence of calcification. There is another
mass of greater-than-brain attenuation without calcification
in the pituitary fossa extending into the suprasellar
cistern. The right side of the dorsum is deviated
posteriorly. Both tumours show homogeneous enhancement
after contrast medium.
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Fig. 5 Right internal carotid angiotomogram. Venous phase. There is a cluster of dilated veins (see arrows) in the middle and posterior thirds of the right orbit within the mass. These appearances are typical of a cavernous haemangioma.

to show the relationship of a pituitary mass to optic chiasm and nerves prior to surgery, further confirmed that the tumour extended from the pituitary fossa into contact with the optic chiasm.

In the presence of a pituitary tumour and with a normal orbital ultrasound the possibility of an endocrine exophthalmos was considered clinically. It seemed unlikely with choroidal folds, pronounced unilateral involvement, and absence of thyrotrrophic abnormalities. Computed tomography left no doubt that the proptosis was due to an intraconal orbital tumour and showed no evidence of thickening of the rectus muscles such as occurs in thyrotrrophic exophthalmos. Computed tomography shows the localisation and extent of intraorbital tumours in the great majority of cases (Lloyd and Ambrose, 1977). It is usually possible to distinguish benign intraconal tumours, which typically show clearly demarcated edges from infiltrating orbital masses, which are much less well defined and may obliterate the outlines of normal structures within the orbit. A haemangioma is the commonest cause of a rounded, well-demarcated, homogeneous, high-attenuation mass, though it is precisely similar to the less-common neurofibroma and to some optic sheath meningiomas. The latter, however, may contain calcification. There is generally no difficulty in distinguishing an optic nerve glioma, which tends to be orientated in the axis of the nerve and inseparable from it. Since there is no blood-brain barrier in the orbit, all intraorbital tumours may enhance and cannot be distinguished by using this technique.

Most orbital haemangiomas show abnormal circulation. This is sometimes rapid, with arteriovenous shunting, but the appearances shown in our patient of cavernous blood spaces outlining with contrast medium in the venous phase are typical of this type of lesion. There is, in addition, a variable amount of solid tumour which causes a mass effect. The deviation of the ophthalmic veins without obstruction is helpful in distinguishing localised benign masses from infiltrating orbital lesions.

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References


