Lesson of the Week

Late diagnosis of pituitary and parapituitary lesions causing visual failure

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It is well recognised that pituitary or parapituitary lesions may compress the optic chiasm, causing visual field defects and ultimately optic atrophy and even blindness. Unfortunately, however, these tumours are still occasionally missed entirely or diagnosed late, so that vision is less likely to recover.¹ We report on five patients seen during the past three years at this hospital in whom the delay between initial presentation and correct diagnosis and referral was up to seven years; this was probably a factor in the persistence of visual impairment after treatment in some of the patients.

Case reports

The table shows the results of pituitary tests and tests of vision before and after treatment in all five patients.

Case 1—A 69 year old man presented to the ophthalmology department of his local hospital in 1981 with blurred vision in his right eye. Central serous retinopathy was diagnosed. Although a right temporal scotoma was noted a year later, he was followed up clinically for a further 18 months, during which time his vision gradually deteriorated. Four years after the initial presentation formal visual field testing by Goldmann perimetry was performed because of further failure of vision; this showed upper bitemporal field defects, and he was referred for assessment. When seen in our unit because of this field defect and because by then a pituitary tumour was suspected he gave an additional history of impotence of 10 years' duration, lethargy of four years' duration, and dull pain behind his right eye. Examination showed that he was hypogonadal with thin, dry skin, and perimetry showed only small nasal fields of vision remaining. Computed tomography of the head confirmed the presence of a large pituitary tumour with suprasellar extension, and endocrine assessment showed it to be a non-functioning adenoma; it was removed transphenoidally with improvement both in his visual fields and in his visual acuity.

Case 2—A 44 year old woman presented to her local hospital in 1979, having awoken with blurred vision in both eyes. Retrobulbar neuritis was diagnosed, and she was given a two year course of injections of adrenocorticotrophic hormone, which resulted in weight gain of 32 kg but little improvement in her vision. In 1982 she was referred to our unit for a second opinion about the cause of her obesity and visual loss. When seen she also gave a history of abrupt cessation of menstruation nine years previously, when she was 35, and a four year history of lethargy and intermittent headaches. Examination showed that she was almost blind, with bilateral optic atrophy. Goldmann perimetry showed sparing of only the left and right nasal fields. Her visual acuity was 6/0 bilaterally. In addition she had mild galactorrhoea. Computed tomography showed a large suprasellar enhancing mass. Endocrine investigations showed hyperprolactinaemia (prolactin concentration 2871 mU/l), pituitary depend-

In patients with unexplained visual impairment a careful endocrine history should be taken with formal plotting of visual fields, plain skull radiography, and, if indicated, computed tomography to exclude a lesion in the pituitary or parapituitary region

ent hypothyroidism, and gonadotrophin deficiency. A craniopharyngioma was removed transfrontally, and although vision improved in the right eye, the left remained blind.

Case 3—A 69 year old woman first presented to an eye department in September 1976 with a one year history of deteriorating vision in her left eye. Bilateral cataracts were diagnosed. Two years later her sight had deteriorated and there was early optic atrophy on the left. At this time she started to complain of headaches behind her eyes, which were occasionally present on waking. Goldmann perimetry showed left temporal field loss. A skull x ray film showed a well defined frontoparietal sclerotic lesion, which was thought most likely to be an osteoma, but a normal pituitary fossa. In May 1980 early optic atrophy on the right was noted as well as established optic atrophy on the left. In November 1983, while she was attending the endocrine clinic for routine follow up of thyroid carcinoma that had been diagnosed in 1972, it was noted that her left eye was blind and there was a possible temporal field defect on the right. Goldmann perimetry showed normal visual fields on the right, but acuity was reduced to 6/18. Computed tomography showed a calcified mass above the pituitary fossa resembling a craniopharyngioma. This was removed transfrontally and found to be a meningioma. She subsequently remained well, but her vision did not improve.

Case 4—A 53 year old man presented to his local ophthalmology department in 1980 with loss of vision in his right eye. Right macular oedema and bilateral peripheral retinal degeneration were diagnosed, and he was treated with cryotherapy, with an initial but short lived improvement. A year later he was referred to our hospital for a second opinion because of continued deterioration of vision. He gave a 10 year history of complete impotence and a six month history of intermittent right sided headaches associated with photophobia. On examination there was disc pallor on the right. Goldmann perimetry showed a large nasal field defect on the right and left upper quadrant hemianopia. Computed tomography confirmed the presence of a large pituitary tumour with suprasellar extension, which was subsequently shown to be a prolactinoma (serum prolactin concentration 11 350 mU/l). He was treated with mesulergine, a dopamine agonist, and given testosterone replacement. The tumour rapidly shrank in size, the prolactin concentration fell to normal, and both his sexual function and vision substantially improved, with visual fields and acuity becoming normal.

Case 5—A 65 year old woman developed frontal headaches in 1984; on one occasion she awoke with severe head pain and was seen in the local hospital, where nothing abnormal was found. Later in 1984 she developed intermittent diplopia and was seen in the local eye hospital, where no abnormality was detected. She was referred for a medical opinion six months after her initial presentation. Clinical examination of her visual fields with a red pin showed bitemporal hemianopia, which was confirmed by Goldmann perimetry; visual acuities were 6/9 on the left and 6/6 on the right. Computed tomography confirmed the presence of a pituitary tumour with suprasellar extension. Endocrine investigations showed this to be a non-functioning tumour, and it was subsequently removed transphenoidally. Postoperatively her visual fields returned to normal and her visual acuity became normal bilaterally.

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Results of tests of pituitary function and visual fields and acuity before and after treatment

	Case 1	Case 2	Case 3	Case 4	Case 5
Results of tests of pituitary function*:					
Follicle stimulating hormone	Decreased	Decreased	Normal	Decreased	Normal
Luteinising hormone	Decreased	Decreased	Normal	Decreased	Decreased
Growth hormone	Decreased	Decreased	Decreased	Decreased	Decreased
Prolactin	Normal	2871 mU/l	Normal	11 350 mU/l	504 mU/l
Thyroid stimulating hormone	Normal	Decreased	Normal	Normal	Normal
Adrenocorticotrophic hormone	Normal	Normal	Normal	Normal	Normal
Visual fields:					
Before treatment	{ Bitemporal hemianopia	Left: small nasal field Right: small nasal field	Left: nil Right: normal	Left: upper quadrant defect Right: nasal field defect	Left: normal Right: upper quadrant defect
After treatment	{ Upper temporal quadrant defect	Left: small nasal field Right: temporal haemianopia	Left: nil Right: temporal hemianopia	Normal	Normal
Visual acuity:					
Before treatment	{ Left: 6/9 Right: 6/18	Left: 6/0 Right: 6/0	Left: blind Right: 6/18	Left: 6/12 Right: 6/0	Left: 6/6 Right: 6/9
After treatment	{ Left: 6/9 Right: 6/9	Left: 6/0 Right: 6/9	Left: blind Right: 6/18	Left: 6/6 Right: 6/6	Left: 6/6 Right: 6/6

*As assessed by basal testosterone, oestradiol, thyroxine, prolactin, and combined pituitary function test with insulin, thyroid releasing hormone, and gonadotrophin releasing hormone.²

Discussion

Pituitary tumours are common: they represent 12% of all intracranial tumours that present clinically and are found in at least 8-10% of unselected patients undergoing postmortem examination.^{3,4} The incidence of pituitary tumours that present with visual symptoms or field defects is decreasing.³ Thus in a review of 1000 patients with pituitary tumours seen up to 1972 Hollenhorst and Younge found that 42% of patients presented with visual symptoms and 70% had field defects on examination.⁵ Cameron *et al* found that half of 44 patients with pituitary tumours seen from 1968 to 1978 had visual field defects.⁶ More recently, Anderson *et al* found that only 9% of 200 patients with pituitary tumours had field defects.³ The reasons for this trend are many, but in particular the advent of new reliable hormone assays, such as for serum prolactin, and the readiness with which patients now present with endocrine symptoms such as menstrual disorders, infertility, impotence, galactorrhoea, and loss of libido mean that many more tumours are being diagnosed and treated earlier than before.

The exclusion of a pituitary or parapituitary lesion in any patient with unexplained visual symptoms or signs is particularly important as treatment of such a lesion will prevent the visual loss that ultimately occurs; indeed, in the absence of optic atrophy, which may follow prolonged compression of the optic nerve, the likelihood of complete or near recovery is high.¹ It is therefore essential to be particularly diligent in seeking symptoms or signs of pituitary or parapituitary lesions so as not to miss or delay such a diagnosis.

Headache is a common symptom in both patients with poor vision and those with pituitary or parapituitary lesions, occurring in about half of such cases.⁶ It is probably too non-specific to be of much diagnostic use by itself, but headaches due to such tumours tend to be retro-orbital, dull, and intermittent, may be worse on waking, and may be associated with changes in posture, diplopia, and photophobia. Headaches were reported spontaneously in all the patients described here.

Loss of secretion of gonadotrophin is often one of the earliest of the endocrine abnormalities caused by pituitary tumours, and impotence in men or abrupt early cessation of menstruation may be the earliest indication of endocrinopathy. It is noteworthy that impotence preceded visual symptoms by several years in the two men described and that in one of the women her periods stopped abruptly

at the early age of 35. None of these symptoms relating to impaired gonadal function had been sought by the clinicians dealing with the patients at an early stage.

Generally, tumours in the region of the optic chiasm need to be large before they cause visual symptoms or signs, and thus most such pituitary tumours are associated with abnormalities of the sella turcica on lateral skull radiography.⁶ This was the case with all three of the patients with pituitary tumours reported on here. With parapituitary lesions a skull radiograph may be normal but a computed tomogram will show a mass lesion if it is present. More recently the visual evoked response has been used to show chiasmal compression or to check visual recovery postoperatively in patients with pituitary tumours. The visual evoked response has the advantage that it can show abnormalities when the visual fields are normal.^{1,7}

We conclude that a careful endocrine history should be taken in patients presenting with atypical or unexplained visual failure or in whom the ophthalmological diagnosis does not account for the pattern of visual loss and that these patients should undergo formal plotting of visual fields, plain radiography of the pituitary fossa, and, if indicated, computed tomography of the pituitary and parapituitary region. Early diagnosis of chiasmal compression due to parapituitary masses is likely to lead to complete recovery of the visual defects, but this may not be the outcome if early symptoms are ignored.

References

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