Pituitary adenoma presenting as the Foster-Kennedy syndrome

Simon Ruben, John Elston, Richard Hayward

Abstract

A 27-year-old man presented to the casualty department with visual failure. Clinically he demonstrated the Foster-Kennedy syndrome. Computed tomography revealed a large space-occupying lesion which was subsequently shown to be a pituitary adenoma. The literature is reviewed and possible mechanisms of the Foster-Kennedy syndrome are discussed.

The Foster-Kennedy syndrome although not a common phenomenon has been described in association with a wide variety of intracranial pathologies. We present here the first case of the Foster-Kennedy syndrome secondary to a pituitary adenoma.

Case report

A 27-year-old man presented to the casualty department of an ophthalmic hospital complaining of visual failure. The vision in his right eye had been declining slowly over the preceding 4 years but over the week prior to presentation it had fallen precipitously resulting in complete blindness in this eye. The vision in the left eye had also deteriorated over the preceding week together with the onset of frontal headaches associated with nausea and vomiting. There were no other symptoms but his general practitioner

had noted the presence of a right convergent squint.

On examination he was generally well, afebrile, alert, and orientated. Visual acuities were no perception of light on the right, hand movements on the left. There was a right relative afferent pupillary defect and fundal examination revealed optic atrophy on the right and marked papilloedema on the left (Fig 1). There was venous congestion and absent venous pulsation in both eyes. In addition he had limitation of lateral rectus function bilaterally, more pronounced on the right, and limitation of upward gaze. There were no other focal or generalised neurological signs. He was therefore diagnosed as having the Foster-Kennedy syndrome.

Computed tomography (CT) scan of the brain showed a large lobulated mass arising in the midline from near the sella turcica and extending behind the dorsum sellae into the anterior part of the posterior cranial fossa indenting the upper pons and filling the interpeduncular cisterns. It also extended upwards into the right frontal lobe distorting the ventricular cistern and obliterating the anterior horn of the lateral ventricle. There were also some areas of cystic cavitation (Fig 2).

The patient was transferred to the National Hospital for Neurology and Neurosurgery where a large right frontal tumour was excised via a frontal craniotomy. Frozen section suggested a pituitary adenoma and formal histology confirmed this and showed positive staining for prolactin. Serum prolactin was measured at 39 000 megaunits/l (normal up to 480 megaunits/l). He was subsequently started on bromocriptine and by 3 weeks postoperatively the prolactin level had fallen to 600 megaunits/l.

At 4 weeks post operation the right visual acuity remained no perception of light but the left had recovered to 6/6 unaided albeit with only a 5° central field. The lateral rectus palsies had greatly improved. Fundal examination now revealed bilateral optic atrophy.

Discussion

The syndrome described by Foster-Kennedy in 1911,¹ characterised by 'the occurrence of true retrobulbar neuritis with the formation of a central scotoma and primary optic atrophy on the side of the lesion together with concomitant papilloedema in the opposite eye', had been reported previously by Gowers² and Paton.³ It has since been associated with a diverse selection of pathologies including frontobasal tumour,^{1,4,7} abscess of the frontal lobe,¹ meningioma of the olfactory groove,^{1,2,8-11} falx¹² or sphenoidal wing,^{5,13-15} craniopharyngioma,¹⁶ plasmacytoma,¹⁷ nasopharyngeal angiofibroma,¹⁸

The Western Ophthalmic Hospital, Marylebone Road, London
S Ruben
J Elston

The National Hospital for Neurology and Neurosurgery, Queen Square, London
R Hayward

Correspondence to:
Simon Ruben, FRCS,
FCOphth, The Western
Ophthalmic Hospital,
Marylebone Road, London
NW1 5YE.

Accepted for publication
26 July 1991

Fig 1A

Figure 1 Optic discs at presentation showing (A) right optic atrophy and (B) left papilloedema.

Fig 1B

neuroblastoma,¹⁹ aneurysms,²⁰ and other diverse intracranial lesions.²¹ We believe this to be the first reported case of the Foster-Kennedy syndrome occurring in conjunction with a pituitary adenoma.

The Foster-Kennedy syndrome is not a common entity. Tonniss found only 28 cases in a series of 3033 intracranial tumours,²² Von Wovern four out of 686 frontal tumours,⁵ Huber one of 46,⁶ and Cushing seven of 29 olfactory groove meningiomas.⁷ This represents an overall incidence of less than 1%. Only 37 cases of the Foster-Kennedy syndrome have been completely documented between the years 1909–89 all but one with evidence of an intracranial mass.¹⁸ Anosmia is often included in the syndrome but its presence has not been universally reported.¹¹ In none of these cases was the mass shown to be a pituitary adenoma although

meningiomas arising from the tuberculum sellae have been described.²³ The mechanism by which the association of optic atrophy and contralateral papilloedema occurs has recently been examined and can not be attributed to a single explanation.¹⁸ The hypotheses include (1) optic nerve compression together with raised intracranial pressure, (2) bilateral optic nerve compression, and (3) chronically raised intracranial pressure without direct optic nerve compression. In this case it is difficult to exclude compression of both optic nerves from the size of the tumour. However the bulk of the tumour was on the side of the optic atrophy and there was evidence of raised intracranial pressure both clinically and on CT scan. Thus a combination of 1 and 2 seems likely and is consistent with a diagnosis of true Foster-Kennedy syndrome.^{12,24} Since it has now been established that the signs first described by Foster Kennedy cannot be used as 'an exact diagnostic sign' it has been suggested that this combination of clinical findings be termed the Foster-Kennedy sign. Sanders, in his study of six patients, concluded that fluorescein angiography should be performed on such cases in order to exclude local disorders at the optic disc.²⁵ However modern imaging techniques give a far more accurate analysis of the underlying pathology and are probably the investigations of choice in the Foster-Kennedy syndrome.

This case is of interest not only in its association with the Foster-Kennedy syndrome but also in light of the declining rate of visual loss as the presenting symptom in pituitary adenomas.^{26,27} Although pituitary tumours are common, representing about 12% of intracranial tumours,²⁶ the Foster-Kennedy syndrome has not previously been described as an association. In older series loss of visual field was considered an early diagnostic sign in cases of pituitary tumours,²⁸ but more recent studies suggest that the incidence of visual abnormalities as a presenting feature is on the decline because of earlier diagnosis.^{26,29} Hollenhorst reported a series of

Fig 2A

Fig 2B

Figure 2 CT scan at presentation showing extent of tumour.

1000 patients of whom 70% had visual loss, 34% optic atrophy, and 6% extraocular muscle abnormalities.³⁰ However in Anderson's review of 200 patients²⁶ only 9% had visual field defects, 2% optic atrophy, and 1% extraocular muscle palsy. It is also of interest that the incidence of visual symptoms in prolactinomas was even lower, the non-secreting chromophobe adenomas having the highest incidence of visual symptoms at presentation.^{26, 28}

Although the incidence of patients with pituitary tumours presenting with visual symptoms is on the decline, ophthalmologists should still be wary of patients with such complaints especially when accompanied by other symptoms such as headache.

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