cytological change in the bone marrow. Multiple extramedullary plasmacytoma is a rare tumour with an incidence of about 1% of all plasma cell tumours. Cranial and intracranial plasma cell tumours are also extremely rare. Cushing found that among 2000 intracranial tumours, only four were intracranial myelomas but he did not record the details.

Clarke reviewed 24 cases of cranial and intracranial plasma cell tumours from the literature and added four of his own. He distinguished three separate syndromes; multiple cranial nerve palsies, intracranial tumour formation, and a constellation of signs due to invasion of the orbit by plasma cell tumour. Intracranial abnormalities are thought to be due to plasmacytomases arising in the base of the skull. Alternatively it has been suggested that increased intracranial pressure could be due to abnormal globulin production inducing a hyperviscosity syndrome with protein deposition in the central nervous system. In our patient, CT scan showed that there was no significant mass lesion and plasma viscosity was normal. Dural involvement around the left mastoid may have lead to sinus thrombosis and consequent raised intracranial pressure.

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Fig High resolution agarose gel electrophoresis showing an abnormal protein band in patients serum and urine. (PAP 3 = normal control pool serum sample).

Abduent palsy after rapid shrinkage of a prolactinoma

Sir.—The sixth cranial or abduent nerve, by virtue of its anatomical course, is particularly susceptible to damage in pathological conditions affecting the cavernous sinus. An expanding pituitary tumour may produce a sixth nerve palsy by lateral displacement and stretching of the cavernous portion of the nerve, often with accompanying third and fourth nerve damage. We report the occurrence of a transient sixth nerve palsy in association with the rapid reduction in size of a pituitary prolactinoma induced by bromocriptine, a relationship not previously documented.

A previously well 31 year old man presented with a two-month history of blurring of vision of the left eye, and diminished libido and potency. He had no associated headache, diplopia or other symptoms. Examination of the left eye revealed a visual acuity of 6/12, an upper temporal quadrantic field loss and pallor of the optic disc. The right eye was normal, as was the remainder of neurological and the general physical examination.

Skull radiographs showed massive expansion of the pituitary fossa, with destruction of the dorsum sellae and almost complete replacement of the adjacent sphenoid sinus. A large, partly cystic pituitary tumour, with suprasellar, cavernous sinus and sphenoidal sinus extension was delineated by axial and coronal CT scanning (fig 1).

Visual evoked potentials showed bilateral prolongation of latencies with attenuation of the major positive peaks, these abnormalities being more severe on the left. The serum prolactin, as measured by radio-immunoassay, was 11,300 ng/ml (normal less than 25). Other parameters of endocrine function were normal, including serum TSH, thyroxine, FSH, LH, growth hormone, cortisol and testosterone levels. Bilateral carotid and left vertebral angiograms confirmed significant suprasellar extension, with elevation of the A 1 segment of the anterior cerebral artery, and lateral displacement of the cavernous portions of the internal carotid arteries. No evidence of intracranial aneurysm formation or of arterial encasement was present.

Oral bromocriptine was commenced at a dosage of 2.5 mg nocte, gradually increasing to 15 mg per day. The patient’s condition remained unchanged until 4 weeks after the institution of treatment, at which time he developed increasing horizontal diplopia over 4 days. There was no associated headache or changes in visual acuity. Exam-
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Fig (a) Pretreatment axial (a) and coronal (b) post-contrast CT scans showing a large, partly cystic sellar tumour with suprasellar and lateral extension. (b) Post-contrast CT scan performed after 4 weeks of treatment with bromocriptine showing a dramatic reduction in tumour size, including extrasellar component.

A combination of pituitary apoplexy due to rapid tumour expansion was suspected, and an urgent CT scan was performed. This revealed a dramatic reduction in the tumour size, including its extra-sellar component (fig. b). There was no evidence of haemorrhage within the remaining tumour. A serum prolactin was 1030 ng/ml, and this fell to 180 ng/ml after an increase of the bromocriptine dosage to 30 mg per day. The sixth nerve palsy gradually resolved over six weeks. He has remained well since, with serial cranial CT scans showing only a small persistent tumour nodule in the sphenoid sinus. Serum prolactin has remained within the normal range.

Paralysis of the sixth cranial nerve is the most frequently encountered extra ocular nerve palsy. Its susceptibility to damage is largely as a result of its long and tortuous anatomical course. After emerging from the pontomedullary sulcus it runs upwards, forwards and laterally in the pontine cistern and usually posterior to the anterior inferior cerebellar artery. Beside the dorsum sellae it turns sharply forwards and pierces the arachnoid and dura mater of the posterior cranial fossa to cross the superior border of the apex of the petrous temporal bone. Then it usually runs beneath the petrophenoidal ligament, although occasionally the nerve divides into bundles that pass above and below this. The nerve then enters the cavernous sinus and it runs forwards immediately lateral and then inferolateral to the internal carotid artery. There are many variations in the origin and course of the sixth nerve, and asymmetries in these variations may explain why damage may often be unilateral lateral. Its long vertical course and sharp angulation over the petrous temporal bone make the nerve particularly liable to damage in conditions producing increased intracranial pressure, presumably as a result of inferior displacement of the brainstem with stretching of the nerve. An increase in intracranial pressure from brain tumours, hydrocephalus or even an acute Valsalva effect may produce a sixth nerve palsy. Paradoxically, the reduction in intracranial pressure produced by both diagnostic lumbar puncture and ventricular shunting for hydrocephalus may also produce transient sixth nerve palsy, possibly as a result of sagging of the intracranial contents with stretching of the nerve. In addition, the postion of the sixth nerve in the cavernous sinus makes it susceptible to damage by pathological conditions in this region, including an expanding pituitary adenoma.
This is particularly true with rapid tumour enlargement, as may occur with haemorrhage into the tumour. Other possible causes of a sixth nerve palsy related to pituitary adenoma are hydrocephalus due to obstruction of the foramen of Monro by tumour, and a coexistent cerebral aneurysm.  

Bromocriptine, a dopamine agonist, is a potent inhibitor of the synthesis and release of prolactin. It may dramatically reduce the size of large prolactinomas and there have been several reports of improvement of visual fields and extracranial movements after its administration.  

The tumour shrinkage is most likely related to reduction in cell size. There is no evidence of widespread tumour necrosis, vascular injury, platelet aggregation or thrombosis after treatment with bromocriptine.  

Our patient developed a sixth nerve palsy in association with a rapid reduction in prolactinoma size, including its extrasellar components. There was no evidence of localised tumour expansion or haemorrhage, hydrocephalus, cerebral aneurysm, recent viral infection or other identifiable causes for the nerve palsy. Although simultaneous independent disease is a possibility, we consider that sixth nerve damage was more likely to have been related to rapid decompression or shift in position of the nerve caused by tumour shrinkage. Hence, the appearance of a sixth nerve palsy during treatment of a large prolactinoma with bromocriptine may signify either rapid tumour expansion or, more rarely, a sudden decrease in tumour size.


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No enophthalmos in Horner’s syndrome

Sir: Enophthalmos is a controversial feature in Horner’s syndrome. Some regard it as mere illusion created by narrowing of the palpebral fissure. We investigated the presence of this sign in a prospective series of patients with oculosympathetic dysfunction. Thirteen patients with unilateral oculosympathetic dysfunction were examined with a “Krahn” exophthalmometer. This instrument makes it possible to measure the exact distance between the anterior surface of the cornea and the lateral margin of the orbit. The patients were examined while sitting. The readings were “blind”, owing to an error of calibration, which caused an artificial difference between the right and left eye, and of which the examiner was unaware. We excluded all intracranial or intraorbital diseases that could affect the position of the eyeball other than by oculosympathetic denervation. Horner’s syndrome had been present in these patients for a few months up to more than ten years. The diagnosis of unilateral oculosympathetic dysfunction was based on previously published criteria involving serial photography of the pupil.  

The exophthalmometric readings in the 13 patients with oculosympathetic dysfunction gave an average of 16-2 mm on the side of oculosympathetic dysfunction and 15-8 mm at the normal side (after correction for the error of calibration). In five cases there was no difference at all, three cases showed enophthalmos (0-5 mm or 1-0 mm) and five showed exophthalmos (1 mm or 2 mm). Four patients had Horner’s syndrome for more than ten years, one for at least five years, the rest for two years or less.

Enophthalmos should no longer be regarded as a part of Horner’s syndrome. In his original description Horner mentioned only in passing that the position of the eyeball seemed slightly inward (“sehr unbedeutend zurückgesunken”). The casuistry of this remark contrasts with the completeness of his description of miosis and ptosis and with his measurements of the temperature of the face. Later writers, however, have included enophthalmos among the main features of the syndrome, and Horner’s chance remark has been perverted into the textbooks of the present day.  

Our measurements fail to show even the slightest relationship between enophthalmos and oculosympathetic dysfunction, not even in patients with miosis and ptosis of more than 10 year’s standing. Similar findings have recently been reported by Lepore and Nielsen.

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