brain weighed 1333 g and showed mild cortical atrophy. Histological examination found widespread subacute spongiform encephalopathy with vacuolation in the frontal, occipital, and both temporal lobes, basal ganglia, thalamus, and cerebellum. The brainstem and spinal cord seemed spared. In the areas of severe spongiform change in the temporal lobes there was extensive neuronal loss and reactive astrocytosis.

Immunocytochemistry for the prion protein (antibody provided courtesy of Dr J Hope, Neuropathogenesis Unit, Edinburgh) showed a positive reaction around areas of confluent vacuolation. These changes were present in both transverse temporal gyri, but the underlying white matter and medial geniculate bodies were unaffected. No amyloid plaques were identified on immunocytochemistry and there was no evidence of any other disease process in the brain. Molecular biological examination of the open reading frame of the prion protein gene showed no mutations and he was methionine-valine heterozygous at codon 129.

Disturbed hearing is unusual in the early stages of Creutzfeldt-Jakob disease. Although six out of 100 experimentally transmitted cases were reported to have "auditory sensory disturbances" in the first stage, these were not defined.[2] Will and Matthews subsequently described the clinical features in 137 pathologically or electrophysiologically confirmed cases, and none of these had auditory symptoms as presenting features or during the course of the illness.[3] Visual disturbance, by contrast, was commoner, with 9% of patients complaining of visual disturbance at the start and 13% exhibiting cortical blindness during the course of the illness. This difference may in part be due to the difficulty in assessing hearing in patients with rapidly progressing dementia. The finding of spongiform change affecting both transverse gyri supports the clinical suspicion of cortical deafness as the cause of this man's hearing loss. The normal CAEPs were unexpected, but as pointed out by Graham *et al*[4] these may be preserved in cortical deafness. It is arguable that these clinical features in the presence of normal CAEPs should be regarded as auditory agnosia, but we prefer to regard the entities of auditory agnosia and cortical deafness as part of a continuum (as do Graham *et al*[4]). It is also of note that normal flash visual evoked potentials have been recorded in cortical blindness.[5] In conclusion, this pathologically verified case of Creutzfeldt-Jakob disease presented with cortical deafness. This is, to our knowledge, the first description of such a presentation, and emphasises the variety of cortical disturbances with which this disease can present.

Speech assessment was performed by Mr Cameron Sellars, genome analysis was undertaken at the AFRC Centre for Genome Research in Edinburgh by Dr Otto Windl and Ms. Maureen Dempster, and Dr Robert Will gave encouragement and guidance.

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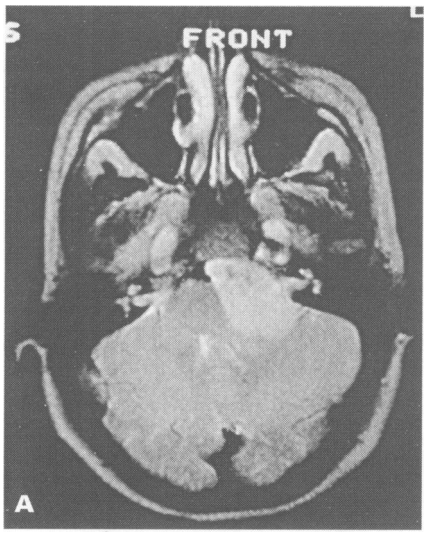
1 Tanaka Y, Kamo T, Yoshida M, Yamadori A. "So-called" cortical deafness. Clinical, neurophysiological and radiological observations. *Brain* 1991;114:2385–401.
2 Bernoulli CC, Masters CL, Gajdusek DC, *et al*. Early clinical features of Creutzfeldt-Jakob disease (subacute spongiform encephalopathy). In: Prusiner SB, Hadlow, WJ, eds. *Slow transmissible disease of the nervous system*. Volume 1. London: Academic Press, 229–41.
3 Will RG, Matthews WB (1984). A retrospective study of Creutzfeldt-Jakob disease in England and Wales 1970–79 I: Clinical features. *J Neurol Neurosurg Psychiatry* 1994; 47:134–40.
4 Graham J, Greenwood R, Lecky B. Cortical deafness. A case report and review of the literature. *J Neurol Sci* 1980;48:35–49.
5 Hess CW, Meienberg O, Ludin HP. Visual evoked potentials in acute occipital blindness. *J Neurol* 1982;227:193–200.

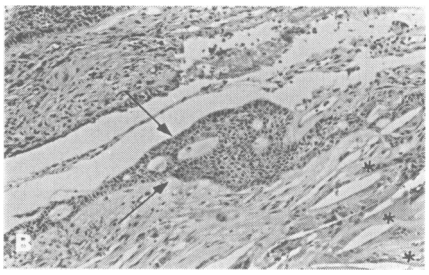
Late recurrence of craniopharyngioma in the cerebellopontine angle in a fertile woman

Suprasellar craniopharyngiomas account for about 3% of intracranial tumours. They are regarded as slowly expanding and arise from remnants of Rathke's pouch epithelium or from metaplasia of pituitary cells.[1] Their tendency to local infiltration makes total surgical removal difficult. Recurrence is to be expected in incompletely removed cases. Postoperative irradiation is generally agreed to be of benefit, but the risk of long term hypopituitarism may be increased.[2] We report a case of craniopharyngioma treated by surgery and cobalt irradiation, which recurred in the cerebellopontine angle 26 years later, in a woman with normal fertility.

A 33 year old woman presented with a 12 week history of left facial numbness and paraesthesiae. There was no visual loss, headache, or urinary symptoms. She had a suprasellar mass removed in 1963 when aged seven and postoperative radiotherapy consisting of 3000 rads of cobalt was given from both sides. The estimated field was 10 cm. There were no records available of the original surgery but the mass was regarded as entirely suprasellar, probably incompletely removed, and the histological diagnosis was craniopharyngioma. She did not receive replacement hormone treatment and had no symptoms until this presentation. She had a normal menarche and had one healthy child delivered by caesarean section. On examination she was small (148 cm), obese, and was 18 weeks pregnant. She had sensory diminution in the ophthalmic and maxillary divisions of the left trigeminal nerve, with an absent left corneal reflex. Visual fields and acuity, and the remaining cranial nerves were normal. General systemic examination, full blood count erythrocyte sedimentation rate, biochemical profile, plasma thyroxine, urinalysis, chest radiograph, left facial electromyograph, visual evoked responses, and CSF analysis were all normal. Computed tomography showed a large contrast enhancing mass in the left cerebellopontine angle extending from the clivus and continuous with an area of dense suprasellar calcification. Left verterbral and carotid arteriography showed posterior displacement of the vertebral artery. There were no feeding vessels. MRI (figure (A)) outlined the mass in the cerebellopontine angle, not involving the internal auditory meatus. The tumour was heterogeneous with areas of calcification and probable necrosis. The T1 and T2 weighted sequences suggested a fat or cholesterol containing tumour such as craniopharyngioma. Surgery was performed six months after delivery of her baby. The trigeminal nerve was stretched over a cyst containing brown fluid. The wall was excised but only limited resection could be done on the calcified anterior portions. A catheter was inserted into the residual cyst, and a Pudenz reservoir placed subcutaneously below the left ear. Postoperatively facial sensation returned to normal and she remains well two years later. The cyst contents have been aspirated on two occasions because of headache.

Endocrine function 14 days after craniotomy was normal as assessed by a Synacthen test, an LHRH test, and plasma thyroxine, and prolactin concentrations. A growth hormone secretion test and TSH measurement were not done. Histological examination of the tissue showed adamantinomatous type epithelium with foci of cystic degeneration, squamous epithelium, calcification, and ossification typical of an adamantinomatous type of craniopharyngioma (figure (B)). The fluid contained polarisable crystals. This case has several





(A) Mass of the tumour outlined by MRI. (B) Histological section of the tumour.

interesting and unusual features. (1) There is an exceptionally long interval of 26 years between the original presentation and recurrence. We have found one reference to a late recurrence of 27 years but there are no details of that case published.[3] Ninety per cent of all incompletely removed craniopharyngiomas recur within 10 years, most in the first postoperative year.[3] In a review of 245 cases the average time between first and second operation was three years.[4] A recent series performed since the advent of microsurgery and better imaging techniques stressed the increased likelihood of recurrence in the adamantinomatous type of craniopharyngioma[5]; the longest interval between surgery and recurrence was eight years in adults and five in children (follow up period 13 years).

(2) This patient presented as a cerebellopontine angle tumour. Of five published accounts of craniopharyngioma shown to extend to the cerebellopontine angle by CT, only three actually presented with clinical symptoms of a cerebellopontine angle tumour. Our case had fifth nerve compression only, and had excellent recovery after operation. Trigeminal nerve palsy has not, to our knowledge, been reported before as the presenting sign in craniopharyngioma. The overwhelming majority are suprasellar but extension into the anterior, middle, or posterior fossa may sometimes occur. Only 4% of Petito's series extended into the posterior fossa.[4]

(3) The relatively normal endocrine and sexual function in our patient is unusual. Endocrine deficiency may be due to the tumour itself damaging the hypothalamic-pituitary pathways, or to treatment including surgery, irradiation, and chemotherapy. Endocrine deficiency is the most common of the potential hazards of radiotherapy after craniopharyngioma. The hypothalamic-growth hormone axis seems to be the most vulnerable, then the gonadotrophins and ACTH and TSH.[2] This patient is obese and of short stature so she is probably growth hormone deficient. Testing for growth hormone was not performed as replacement was not considered. Our patient has normal gonadotrophin activity, as evidenced by normal menstruation and pregnancy, dispite the large field of radiation she received. Many woman with craniopharyngioma have primary amenorrhea and require ovarian stimulation to achieve pregnancy. The pituitary gland itself is relatively protected, however, in irradiation for craniopharyngioma. We believe that in this patient, because the mass subsequently extended into the posterior fossa, the hypothalamic-pituitary axis was spared from local destructive effects.

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1 Rubinstein LJ. Tumours and tumour-like lesions of maldevelopmental origin. In: Russell DS, Rubinstein LJ, eds. *Pathology of tumours of the nervous system*. 5th ed. London: Edward Arnold, 1989:696.
2 Littley MD, Shalet SM, Beardwell CG. Radiation and hypothalamic-pituitary function. *Balliere's Clinical Endocrinology and Metabolism* 1990;4:147–75.
3 Wilkins RH, Rengachary SS. Neurooncology. In: Wilkins RH, Rengachary SS, eds. *Neurosurgery*. Vol 1. New York: McGraw-Hill, 1985:912-4.
4 Petito CK, De Girolami U, Earle KM. Craniopharyngiomas. A clinical and pathological review. *Cancer* 1976;37:1944–52.
5 Adamson TE, Wiestler OD, Kleihues P, Yasargil MG. Correlation of clinical and pathological features in surgically treated craniopharyngiomas. *J Neurosurg* 1990;73: 12–7.

MATTERS ARISING

Dementia and door-to-door studies in Spain

Coria *et al*[1] have recently published a very original and interesting paper on prevalence of dementia in a rural population in Spain (Turégano). The authors claim that this study is the first door-to-door survey performed in a definite community in Spain. This statement could lead to some misunderstanding because, in fact, there are more data on prevalence of dementia in Spain obtained from door-to-door surveys. In the past five years, several door-to-door surveys on prevalence of dementia have been performed in Spain. The first reported in English was carried out by Lopez-Pousa *et al*[2] in a random sample of eight towns in Gerona in 1991. The second described in English is the Zaragoza-Liverpool study (a study based on a random sample of old people in an urban city).[3] Another door-to-door survey reported in May 1993 in English was carried out by Bermejo *et al* as a random sample of old people from four districts of Madrid.[4] The preliminary data of these three studies have been reported previously in Spanish.[5] More studies have been carried out (Toledo; Pamplona; Baix de Camp, Tarragona) but as yet these studies are unpublished, apart from some preliminary data.[5]

It is noteworthy that all published studies have been performed with standardised neuropsychological protocols (CAMDEX, GMS), and international diagnostic criteria (DSM-III or DSM-III-R) and all these studies are door-to-door surveys. The most outstanding fact is that the data of prevalence of dementia (people over 60 or 65 years) are quite different and range between 14·2% in Gerona to 5·2% in Turégano (10% in Madrid, 6·7% in Zaragoza).[1–5] But there is a uniform finding: Alzheimer's disease is the most frequent cause of dementia in these Spanish surveys, as it is in others done in Western countries.

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1 Coria F, Gómez del Caso L, Minguez L, Rodriguez-Artalejo F, Claveriá LE. Prevalence of age-associated memory impairment and dementia in a rural community. *J Neurol Neurosurg Psychiatry*, 1993;56:973–6.
2 López-Pousa S, Vilalta S, Llinás J, *et al*. Prevalence of dementia in the general population: pilot study in Girona (Spain). *Neuroepidemiology*, 1991;10:99.
3 Lobo A, Dewey M, Copeland J, *et al*. The prevalence of dementia among elderly people living in Zaragoza and Liverpool. *Psychol Med* 1992;22:239–43.
4 Bermejo F, Colmenarejo C, Tejeiro J, *et al*. Prevalence of dementia in old people in four districts of Madrid: A pilot study. *Neuroepidemiology* 1993;12:135.
5 Bermejo F (ed.). *Nivel de salud y deterioro cognitivo en los ancianos*. Barcelona: SG Editores, 1993.

Coria replies:

Many different groups in Spain are conducting epidemiological studies on dementia, as claimed by Bermejo and Morales above. Nevertheless, ours is still the first to address the prevalence of age-associated memory impairment, using validated instruments.[1 2] The only available definitive data on the prevalence of dementia in Spain were included in the EURODEM study, and the results of this study were referenced (see ref. 19 in ref. 1). The high prevalence of dementia found in one study[3] is probably caused by the inclusion of cases with mild dementia and age-associated memory impairment. Subtraction of these cases gives prevalence rates of overt dementia similar to those found in Segovia (Lopez-Pousa, personal communication).

In any case, analysis and meta-analysis of the extensive data on the subject now available in Spain would provide a good opportunity to address several controversial issues related to the methodological problems associated with epidemiological studies on dementia and other related disorders. In addition, these studies should provide large numbers of well studied cases for molecular and genetic analysis of the early stages of Alzheimer's disease.[4]

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2 Gómez de Caso JA, Rodriguez-Artalejo F, Claveria LE, Coria F. The value of the Hodgkinson's test for detecting dementia and mild cognitive impairment in epidemiological surveys. *Neuroepidemiology* 1994 (in press).
3 López-Pousa S, Vilalta S, Llinás J, *et al*. Prevalence of dementia in the general population: pilot study in Girona (Spain). *Neuroepidemiology* 1991;10:99.
4 Coria F. Clinical and molecular aspects of age-associated memory impairment. *Ann Med* 1994 (in press).

NOTICE

The second joint meeting of the **British Neuropsychiatric Association** and the **American Neuropsychiatry Association** will take place in New Port, RI, USA on 21–24 July 1994. For further information, contact Professor M A Ron, Department of Neuropsychiatry, The National Hospital, Queen Square, London WC1N 3BG. Tel: + 44 71 837 3611; fax: +44 71 829 8720.