Meningioma mimicking fibrous dysplasia of the skull

Dr Frankel et al1 give an interesting account of what is clearly a very unusual and instructive case. We note, however, the absence of histological proof of fibrous dysplasia. Despite very persuasive clinical and radiological evidence, we must emphasise that histological examination is crucial in establishing the validity of any association between fibrous dysplasia and the development of a meningioma. Meningiomatous infiltration of bone, without soft tissue extension, may closely simulate the typical radiological appearances of fibrous dysplasia.2 A recent case of ours, showing some similarities to that of Frankel et al, illustrates this important point.

A 46 year old woman was admitted in June 1987 with increasing proptosis and pain in the left orbit. Four years previously she had undergone a frontal craniotomy and left optic nerve decompression in another hospital. At that time her complaint had been an 18 month history of left orbital pain and progressive blindness. She also had a very long history, extending over more than 20 years, of temporomandibular joint dysfunction. On examination there was decreased sensation to light touch and pinprick over the left side of the face associated with proptosis. There was no vision in the left eye and fundal examination showed optic atrophy. She was otherwise well. Skull radiographs and CT scan of the head showed evidence of previous surgery to the postero-lateral wall of the left orbit and region of the optic foramen. There was thickening of the medial end of the left sphenoid bone and part of the postero-lateral wall of the left orbit (fig, inset). Features were regarded by both surgeons and radiologists as typical of fibrous dysplasia. Routine serum biochemistry, and in particular serum calcium and alkaline phosphatase levels, were normal.

The patient had a further frontal craniotomy and abnormal bone was removed from the left sphenoid and orbit. The side wall of the orbit was repaired using a rib graft. No soft tissue mass was identified. The bone was sent for histological examination. Tissue sections showed no evidence of fibrous dysplasia. Trabeculae were composed of dense, sclerotic but otherwise normal, lamellar bone. The intervening spaces contained infiltrating islands of transitional meningioma (fig).

Meningioma infiltrating bone without any associated soft tissue mass is a rare but recognised phenomenon.3 It is clear from our case that such a tumour associated with hyperostosis may closely mimic the clinical and radiological features of fibrous dysplasia. We suggest therefore that claims of a causal link between fibrous dysplasia and the subsequent development of meningioma cannot be proved without histological evidence of the former condition.

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Disappearing CT lesions in Indian patients with epilepsy

We read with interest the article by Ahuja et al on “disappearing” CT lesions in Indian patients with epilepsy.1 We agree with their conclusion that tuberculosis is rarely, if ever, the cause of these lesions. We are, however, convinced that in nearly all cases cysticercosis is the causative pathology. We are discussing lesions which are single, rounded, less than 10 mm in size, enhancing with contrast in patients with seizures.

In our study of 15 consecutive patients in whom lesions were excised, cysticercosis was the definite diagnosis in seven, and a parasitic granuloma was diagnosed in five patients.2 In the other three patients there was also inflammation, calcification and gliosis in the biopsy specimens which could be the result of a resistant parasitic granuloma. Since the end of this study we have stopped routine excision of these lesions as they are self-limiting and benign. In isolated cases (10) where excision was carried out because of persistence of the lesion and cysticercosis was the only pathology identified.

We presume that the authors speculated on a hypothetical “micro” lesion of unknown aetiology at the cause of some seizures, because of the inability to detect cysticercus antibodies in two-thirds of their patients. It is well known that serological tests do not identify all patients with cysticercosis and negative tests are more common in patients with benign forms of the disease.2 Negative tests have been reported even with more sensitive serological tests such as the immunoblot assay.3

We also do not agree with the authors’ contention that enhancement of the lesion is related to the seizure. In all except one patient in our study, CT scans were carried out more than two weeks after a seizure and all of them had enhancing lesions. The nature of the lesion on the CT scan and its enhancement depend on the stage of natural evolution of the parasite. The phenomenon of disappearance and reappearance of the lesion at the same site is intriguing. We have never seen such an event and one possibility to consider is that lesions can be missed if the CT scan slices in the region of interest are not thin (2–4 mm).

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1 Ahuja GK, Behari M, Prasad K. Disappearing CT lesions in epilepsy: is tuberculosis or cysticercosis the cause? J Neurol Neurosurg Psychiatry 1989;52:915–6.


Dr Ahuja et al reply:

We thank Drs Rajeshkar and Abraham for their comments. There is little doubt that we are talking of similar lesions.1 Being aware of the natural history of the CT lesions we considered excision biopsy of cerebral lesion unethical and followed a different strategy to

Island of transitional meningioma within sclerotic bone. Inset shows CT scan of skull (abnormality arrowed).
ascertain the aetiology. It is true that serology may miss some cases of cisticercosis but our results are not grossly dissimilar. They could demonstrate cisticercosis in seven out of 15 cases (46%) while in our series serology was positive in 31%. The number in both studies is relatively small and much should be done to produce consistent percentages. In our opinion persistent persistently should not be compared with disappearance lesions as the two may be entirely different.

Our hypothesis that contrast enhancement is due to a recent seizure is based on well documented evidence in the literature that seizures lead to transient breakdown in blood-brain barrier. This is supported by observation in at least five patients in whom the lesion disappeared, reappearing after a flurry of seizures to disappear again. We are unable to accept the argument that this could all be due to technical factors as suggested by Drs Rajshhekar and Abraham. Since lesions due to other causes are known to show similar CT morphology, it is not wise to state that cisticercosis is the only underlying cause of “disappearing lesions”. Larger studies using different studies and methods to answer the question and one such study has been initiated in our department.

The lacunar hypothesis

The paper by Drs Anzalone and Landi is an interesting contribution to the debate about the validity of the “lacunar hypothesis” that links a small number of clinical syndromes to occlusion of a single perforating artery by specific vasculopathies. The significant number of non-lacunar lesions in their series certainly seems to justify the early scanning of these patients, but in the context of cerebrovascular disease it is important that the result is not interpreted as a failure of the lacunar hypothesis.

Although the authors do not state the number of CT scans which showed an appropriately sized small deep infarct, our experience using a similar scanner in patients that is unlikely to have been more than about 50%. This lack of a definite clinicoradiological correlation in a significant number of cases, matched with isolated reports of patients with lacunar syndromes and more extensive (non-lacunar) areas of infarction, has meant that concern is still expressed about how often non-lacunar infarction may present in this way. This is despite recent reports which have shown that the majority of patients who present with a lacunar syndrome and a negative CT scan have an appropriate small deep infarct on MRI.

It is possible that many of the patients who we are reported to have had a lacunar syndrome from extracerebral thrombosis, and who presented with a popliteal vein thrombosis during the course of his neurological disease. The authors rightly envisaged sinus thrombosis but ruled it out on a single digitalised intravenous angiography. We think that this investigation alone is not sufficient to exclude this diagnosis in their patient. The timing of the angiography in relation to the venous thrombosis and magnetic resonance imaging (MRI) studies have shown the possibility of rapid reperfusion of the vessel. The sinus blockage is sometimes incomplete and a greater volume of contrast material may be necessary to evaluate the venous sinuses better. Collateral circulation in the sinus wall may simulate the normal opacification of the sinus by contrast material.

Though heparin has a proven efficacy in cerebral venous thrombosis the lack of improvement during anticoagulant treatment in their patient does not rule out this possibility. As for the dramatic improvement one week after starting hydroxyurea, it is compatible with both benign intracranial hypertension and dural sinus thrombosis.

The hypothesis of an intermittent sinus blockage may well be right, as sinus thrombocythaemia the cause of intracranial hypertension in their patient. However, dural sinus thrombosis has not been fully excluded. It should be looked for with appropriate techniques (four vessel arteriography or MRI) in patients with essential thrombocythaemia presenting symptoms of intracranial hypertension, be it isolated or associated with epilepsy or focal deficit.

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Plasma serotonin

Recently Anthony and Lance published their interesting results on serotonin in patients with chronic tension headache. However, the title of their paper contains a serious error which is also repeated in the text. From the methods section it can be inferred that the authors used whole blood for their study. They even state that “Plasma serotonin was expressed as ng/platelets, since they contain about 98% of serotonin in blood.” This means that in this study not plasma, but whole blood serotonin was studied and reported, as is also apparent from the results. Anderson et al recognise three compartments of whole blood serotonin, plasma,


Possibly benign intracranial hypertension and essential thrombocythaemia

We read with interest the paper by Enack et al on benign intracranial hypertension and essential thrombocythaemia.

The syndrome of isolated intracranial hypertension with normal cerebrospinal fluid and CT scan presented by their patient does indeed suggest an essential hypertension. Such a diagnosis, however, seems difficult to admit in a patient with essential thrombocythaemia, which has been reported as a possible aetiology of cerebral venous thrombosis, and who presents with a popliteal vein thrombosis during the course of his neurological disease.

The authors rightly envisaged sinus thrombosis but ruled it out on a single digitalised intravenous angiography. We think that this investigation alone is not sufficient to exclude this diagnosis in their patient. The timing of the angiography in relation to the venous thrombosis and magnetic resonance imaging (MRI) studies have shown the possibility of rapid reperfusion of the vessel. The sinus blockage is sometimes incomplete and a greater volume of contrast material may be necessary to evaluate the venous sinuses better. Collateral circulation in the sinus wall may simulate the normal opacification of the sinus by contrast material.

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MATTERS ARISING: Dr Ahuja et al reply:

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