

## Letters

**Scrotal pain with testicular jerking: an unusual manifestation of epilepsy**

Sir: Many types of partial seizures are known to occur, depending on the site of origin of the epileptic activity in the cerebral cortex. Scrotal pain associated with testicular jerking is an unusual form of seizure.

A 10 year old boy was brought with the complaint of episodic lancinating pain in the scrotum associated with repetitive up and down jerking of the right testis for the past 7 months. The attack usually lasted 2-3 min. It was abrupt in onset and equally abrupt in remission, and occurred approximately ten times every month, sometimes during sleep. The attack which occurred a day prior to hospitalisation was dramatic in that the "scrotal attack" lead on to convulsions involving the right lower limb followed by weakness.

On examination, the boy had normal mentation and cranial nerve function. The upper limbs were normal. A right crural monoparesis with blunting of superficial sensation and joint-position sense in the same limb was present. The boy could not cooperate well for detailed cortical sensory testing. The right plantar response was extensor; the abdominal reflexes and the cremasteric reflex on the right side were absent; and the right ankle and knee jerks were pathologically brisk. Two days later, the right lower limb power returned to near normal though the plantar response remained extensor and the cremasteric and abdominal reflexes could still not be elicited. The ocular fundi were normal. On the 3rd and 4th days after admission attacks were witnessed. The boy who was playing cheerfully, suddenly started screaming and clutched his scrotum with his hands. His right testis was seen to exhibit an arrhythmic jerky up and down movement (at about 2 to 3 times per second) lasting for approximately 1½ minutes, and then it remained in an elevated position for nearly a minute. The entire attack lasted about 2½ minutes and this was followed by slow return of the right testis to its normal position within the scrotum. No obvious contraction or twitching of the lower abdominal muscles could be seen or felt during the attack.

Routine investigations including blood WBC counts, plain radiographs of skull, urinalysis and stool examination were normal. The Mantoux test was strongly positive and the erythrocyte sedimentation rate (ESR) 55 mm during the first hour. EEG on the

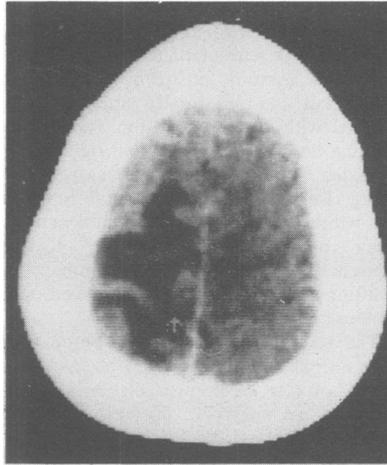


Fig Superficial cortical cut in plain CT scan showing a lesion (arrow mark) in the left paracentral region surrounded by a fairly extensive low dense zone suggestive of white matter oedema. (Hounsefield values: lesion: 48.5 before contrast and 50.6 after contrast, low dense zone, 14).

10th post-admission day showed random epileptic activity over the left central area with phase reversal at C3. CT on the 2nd post-admission day revealed a paracentral superficial cortical lesion (fig). In the subsequent CT scan after 6 months of treatment with streptomycin, rifampicin and isoniazid, the lesion had disappeared totally.

Localised pain as an epileptic manifestation is rare and is usually an outcome of parietal lesions<sup>1</sup> or occasionally due to a temporal lobe focus.<sup>2</sup> Unpleasant episodic clitoral warmth and orgasmic epilepsy have also been reported in parietal lesions.<sup>3</sup> A closely similar case with episodic scrotal pain as an ictal manifestation, sometimes associated with clonic contraction of the left thigh, arm or eyelid was described by York *et al*<sup>4</sup> who found epileptic activity originating at the right central area on the EEG. Cremasteric contraction with testicular jerking, as seen in the present case, has not been reported previously.

The cremaster (a striated muscle), on contraction, elevates the ipsilateral testis from its suspended position within the scrotum. From embryological studies, the cremaster is considered as the lowermost extension of the obliquus internus abdominis.<sup>5</sup> Bearing in mind the possibility that contraction of the larger muscle (oblique internus) could elicit simultaneous contractions in the appendage (cremaster), the lower abdominal muscle were carefully observed during an

epileptic attack. No contraction was noticed. Although developmentally the obliquus internus abdominis and the cremaster are closely related, they do not generally act in unison. Moreover, unlike the obliquus internus, the cremaster cannot usually be contracted by voluntary effort. Again, elicitation of the superficial lower abdominal reflex does not cause cremasteric contraction, nor does elicitation of the cremasteric reflex cause abdominal muscle contraction. Though apparently small and insignificant, the cremaster appears to have an independent role to play.

The lesion (a tuberculoma) in this case lay at the left paracentral area where the motor and sensory representations on the cortex for the right half of trunk, hip, and the right lower limb are represented. Epileptic phenomena, as a result of lesions in this location produce Jacksonian fits or sensory attacks usually commencing in the big toe.<sup>6</sup> Even Penfield and Rasmussen<sup>7</sup> who had conducted a systematic study of stimulation of the sensory-motor cortex at the paracentral level had not reported cremasteric contractions with testicular movement. With this clinical presentation and the CT appearances it is postulated that a cortical centre which activates the cremaster exists, a centre that cannot, however, be controlled by voluntary effort. Thus, the cremaster, in addition to elevating the testis by neural mechanisms for purposes of the neuro-regulation or during the sexual act<sup>8</sup> can possibly contract by supraspinal (cortical) influences as well. The neurophysiological implication of such a mechanism remains unclear.

PA BHASKAR

The Department of Neurology,  
and Clinical Neurophysiology,  
Medical College Hospital,  
Thanjavur, India 613 004.

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**Movement disorder associated with abnormal copper metabolism and decreased blood anti-oxidants**

Sir: Dystonia not due to Wilson's disease sometimes occurs in association with abnormal copper metabolism.<sup>1</sup> The copper metabolism abnormalities are similar to those found in heterozygotes for the Wilson's gene but the majority of these subjects are clearly asymptomatic. Other factors which may make patients unusually sensitive to minor disturbances of copper metabolism are not known. Increased tissue levels of transition metal ions are known to exert an increased pro-oxidant effect resulting in a greater tendency to membrane lipid damage and catecholamine oxidation.<sup>2</sup> Moreover, such mechanisms are increasingly regarded as important in extrapyramidal disorders.<sup>3</sup> The normal defence against such oxidation is provided by a range of antioxidants of which glutathione is an important intracellular one.<sup>4</sup> We describe a patient with a dystonic movement disorder who was found to have low serum copper, increased liver copper and decreased erythrocyte glutathione levels. We suggest that the combination of the copper and glutathione abnormalities may have resulted in dystonia when either on its own may have been insufficient to cause symptoms.

A 15 year old Indian schoolboy presented with a 3 year history suggestive of intellectual deterioration which improved spontaneously while undergoing investigation and a 1 year history of persistent and progressive abnormal movements and posture. During this year he had lost 6 kg in weight. He was not taking any drugs and in particular had never taken phenothiazines or other antidopaminergic medication. There were no abnormal vocalisations nor any obsessive compulsive behaviour. There was no family history of neuropsychiatric disorder or consanguinity, and perinatal and developmental history were normal. Examination showed tics of the face with frequent grimacing and to a lesser extent also

involving his neck. His knees, right elbow and spine were held in partial flexion, the right shoulder semi-abducted and there was occasional dystonic posturing of the right hand. Gait was abnormally rapid and long stepped with some propulsion and during walking his right arm was held rigid at the shoulder with resulting decreased arm swing. Formal testing of mental function was within normal limits and there were no other neurological signs. No Kayser Fleischer rings were seen on slit lamp examinations done twice by an experienced ophthalmologist.

The results of the following blood tests were normal: blood count, erythrocyte sedimentation rate, serum urea, electrolytes, bilirubin, liver enzymes, proteins including immunoglobulins, calcium and treponemal serology, serum transferrin, ascorbate, dehydroascorbate and products of lipid peroxidation in serum measured as diene conjugates and as thiobarbituric acid reactivity. CSF copper as measured by atomic absorption spectrometry and by its pro-oxidant potential<sup>5</sup> was found to be normal. In addition, chest radiograph, CT scan of the head, electroencephalograph, isotope scans of the liver and spleen and CSF microscopy and protein content were normal. Serum copper was marginally low, as was serum caeruloplasmin. Liver copper measured by atomic absorption spectrophotometry was raised but not to the levels normally accepted for Wilson's disease. Red cell reduced glutathione measured by the method of Crowley *et al*<sup>6</sup> was low. This method is in regular use in our laboratory where the normal ranges quoted have been established. These are in good agreement with normal ranges reported by other groups. These findings are summarised in the table. His parents' serum copper and caeruloplasmin were measured and were found to lie well within normal limits.

Although patients with neurological problems due to Wilson's disease who have

no Kayser Fleischer rings have very rarely been described<sup>7</sup> our patient cannot be regarded as coming within this category. The modest elevation in liver copper with normal histology, normal urine copper levels and low serum copper and caeruloplasmin might all individually occur in classical Wilson's disease but taken together they are not acceptable for that diagnosis.

Caeruloplasmin is the main anti-oxidant in human plasma<sup>8</sup> and glutathione an important one within cells<sup>4</sup> where it is known to play a role in membrane stabilisation and in providing a suitable reducing environment. When levels of one antioxidant are decreased the functional demands on other antioxidants are increased. Marginal lowering of serum caeruloplasmin along with significant lowering of intracellular glutathione may together have resulted in decreased antioxidant protection both in cells and in serum. In vitro low levels of glutathione in the presence of metal ions increase the risk of free radical mediated oxidative damage.<sup>9,10</sup> Thus, decreased levels of glutathione along with increased tissue copper may be especially harmful. It seems unlikely that the glutathione deficiency is a consequence of increased glutathione consumption as in that case levels of all antioxidants should be low, which is not true for ascorbate and also levels of serum lipid peroxidation products are within normal limits.

There are reports in the literature of abnormalities suggestive of heterozygous state for the Wilson's disease allele association with extrapyramidal dysfunction.<sup>1,11</sup> This may be the explanation in our patient although the fact that both his parents' serum caeruloplasmin concentrations are entirely normal is against this. It is equally possible that Wilson's disease is not a single entity and that patients such as we describe will prove genetically distinct from classical Wilson's disease. Penicillamine which is the drug of choice in the treatment of Wilson's

Table Results of copper and glutathione studies

Test	Result	Normal range	Method
Serum copper	0.5 mg/l	0.7-1.6	Atomic absorption spectrophotometry (AAS)
Serum caeruloplasmin	0.15 g/l 450 mg/l	0.2-0.6 480-840	Immunoassay ferroxidase activity
Basal urine copper excretion	15 µg/24 hours	< 70	
Liver copper concentration	30 µg/g wet weight	< 10 (Wilson's disease) (Range > 50)	AAS (Addenbrooke's Hospital Cambridge)
Whole blood glutathione	106 mg/l 24 mg GSH/10 <sup>12</sup> Red Cells	> 280 40-90	(Reference 6)