a factor predisposing to the syndrome. Residual neurological sequelae have been described in some patients with NMS, mostly signs of Parkinsonism or decrease of general intellectual faculties.8-10 Our patient who had a long and severe course in relation to the syndrome was left with a hypertonia (mainly of the left hand) and dysarthria, though we cannot tell whether they are due to his Wilson’s disease or to the NMS.

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VASSILIS KONTAXAKIS
COSTAS STEFANIS
MARIOS MARKIDIS
VASSILIKI TSERPE
Department of Psychiatry, Athens University Medical School, Eginion Hospital 74, Vas Sophias Avenue, 11528 Athens, Greece

References

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Brain death and pinpoint pupils

Sir: Non-reactive pupils are one of the cardinal signs in brain death. Contrary to what was required at one stage it is no longer considered necessary for the pupils to be dilated. In fact they are most often found to be in the mid position.3 3 Pinpoint pupils are not a feature of brain death and may be the result of a bilateral pontine lesion affecting the sympathetic fibres. That pinpoint pupils may be seen in verified brain death is evident from the following case.

The patient was a 69 year old woman with coronary heart disease for 12 years, and one mild ischaemic stroke two months before carotid endarterectomy. The endarterectomy was performed successfully but twenty hours after operation the sutures of the carotid artery ruptured resulting in a profuse bleeding. The huge haematoma in the neck prevented intubation, and an emergency tracheostomy was performed. During about ten minutes the respiration was severely impaired and the situation was further complicated by cardiac arrest. After ten minutes' resuscitation the heart began to beat but the intra-arterially measured blood pressure readings stayed for 80 minutes at the level of 30–50 mm Hg systolic. The patient did not regain consciousness, and six hours later she was deeply comatous with no pupillary reactions, no response to the oculo-cerebellar test, and no grimacing during firm compression of the supraorbital nerves.

The patient was ventilated for ten minutes with 100% oxygen and then disconnected from the respirator for 10 minutes. During the disconnection 100% oxygen was insufflated into the trachea at a rate of 6 l/min. At the time of the disconnection the Paco2 level was 4±5 kPa which, according to a recent study4 results in final Paco2 levels giving a maximal stimulation of the respiratory centre. No spontaneous breathing movements occurred during the test. Because the non-reactive pupils were of pin-point size and thus not in accordance with the criteria of brain death, the patient was again connected with the respirator. Twenty-four hours later the examination gave the same result, the pupils were still of pinpoint size. The scrutiny of the case history revealed glaucoma treated with pilocarpine eye drops given twice daily in both eyes before and after surgery. The patient was disconnected from the respirator, and two days later a medicoegal necropsy showed a typical respirator brain.

Our case emphasises the importance of a thorough scrutiny of the medication used in case of suspect brain death. In his excellent articles on brain stem death Pallis mentioned as pitfalls in the diagnosis anticholinergic drugs, neuromuscular blockers, and pre-existing eye disease.3 3 In 1971, Wexler reported of two patients in irreversible coma with non-mydriatic pupils who had been using miotics for glaucoma. We would like to add pilocarpine to the list of pitfalls in the diagnosis of brain death. It acts as a cholinergic mimetic directly on cholinergic receptors, an effect which is not abolished by denervation. Because glaucoma is common especially among older people, and is often treated with pilocarpine, this possible cause of mioptic pupils in suspect brain death must be kept in mind.

RAINER FOGELHOLM
Department of Neurology
RAILI LARI-SOMPA
Department of Anaesthesiology
Central Hospital of Central-Finland
40620 Jyväskylä
Finland

References

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Acute dystonic reaction with asterixis and myoclonus following metoclopramide therapy

Sir: Extrapyramidal side effects are well recognised following medication with metoclopramide, a selective D2 dopamine antagonist.1 2 Some 95% of these effects are of the acute dystonic-dyskinetic type. They occur mostly in younger females, within 6 hours of the drug, and disappear without specific treatment. Oculogyric crises, torticolis, opisthotonos, and orofacial dyskinesias are often present. However, the occurrence of asterixis and myoclonus together with acute dystonic reactions has not been reported. We observed such a patient.
A 65 year old previously healthy farmer was brought to the emergency room 18 hours after the acute onset of symptoms. There was no family history of movement disorders, or of neuroleptic-induced extrapyramidal reactions. Four days before admission, he experienced mild epigastric distress and abdominal fullness, for which 2 days later, he took one tablet, 5 mg of oral metoclopramide, dextromethorphan, and Gasgel (simethicone, aluminum magnesium hydrate and magnesium oxide). Some 30 hours after the first dose, and 2 hours after the second dose, he developed irregular, intermittent muscle jerking, most obviously in the face, mouth, and limbs (fig. a). The jerks could be initiated when the patient began to speak or to move a limb, but were not affected by external stimuli such as pinprick, touch, vibration, light, or sound. The jerks disturbed speech and swallowing. There was no change in the level of consciousness. Obvious irregular independent flapping tremors were observed in both outstretched hands when the wrists and fingers were extended (fig. b). There was involuntary hyperextension of the neck and forceful opening of the mouth. Routine laboratory studies were all normal including liver function tests. A chest radiograph was normal. EEG (awake) revealed no abnormality. The asterixis, myoclonus, and acute dystonic reactions cleared within 12 hours. On follow-up 6 months later the patient was normal.

Myoclonus has been described in association with the acute hyperkinetic syndrome in neuroleptic agents. It has not been recorded in the extrapyramidal side effects of metoclopramide. Asterixis also is a rare sign in diffuse encephalopathies in various metabolic disorders as well as a rare manifestation of focal lesions in the parietal lobe, thalamus, midbrain andpons. Asterixis and myoclonus were the dominant features in our patient with a relatively mild acute dystonic reaction. Both sets of manifestations disappeared at about the same time. Neuroleptic-induced acute dystonic reactions may be due to enhanced dopamine release on supersensitive postsynaptic receptors. Whether the asterixis and myoclonus can also be attributed to a similar (hypothesised) transient increase of dopamine acting on postsynaptic receptors, is an open question.

Both myoclonus (using stronger stimulation) and asterixis can be produced by electrical stimulation of the human motor cortex through the intact scalp.

The possibility of a metoclopramide-induced encephalopathy responsible for both the acute manifestations of asterixis as well as myoclonus might be considered in our particular patient.

**References**


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**Bilateral spontaneous carotid-cavernous fistulas, associated with systemic hypertension and generalised arteriosclerosis: a case report**

Sir: Spontaneous carotico-cavernous fistulas have been repeatedly reported as a result of improved angiographic technique and better clinical recognition. They are usually indirect. Of the 21 cases with spontaneous carotico-cavernous fistulas observed by Nukui et al. only one was direct; all 20 cases of direct fistulas reported by Peeters and Krüger and Peeters, and all, but one, of the 54 cases reported by Debrun et al. had a history of trauma; thus direct spontaneous carotico-cavernous fistulas are very rare. Even rarer is the occurrence of bilateral direct fistulas whether of spontaneous or traumatic. With the exception of one patient in the 54 patients series reported by Debrun et al. all the direct fistulas reported above were unilateral.

We report a patient with bilateral direct spontaneous carotico-cavernous fistulas associated with hypertension and intra-cranial arteriosclerosis.

A 7 year old woman noticed one morning, on waking up, a swelling of the right eye lids and redness of the right eye, accompanied by a pulsating whizzing noise all over the head. She also experienced transient diplopia on looking to the right with horizontally shifted images, and a clumsy feeling of the left arm. Her complaints were not preceded by episodes of headache or fever. There was no history of head injury. Acuity of her vision was not altered; she had had poor vision of the left eye ever since the delivery of a child more than 30 years previously. Apart from systemic hypertension which was well controlled on betablockers and diuretics she had no other major illnesses. During the next 2 months, her clinical symptoms steadily improved; the whizzing noise got less, the eye lids were less swollen and diplopia diminished. Because of her symptoms she consulted an ophthalmologist who referred her to the neurology