CAPGRAS SYNDROME WITH DISORDER OF COPPER METABOLISM: A CASE REPORT

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A review of the descriptions of Capgras syndrome in literature shows that the syndrome may be defined as 'the delusional belief in the existence of doubles of others, or of one-self or both' (Berson, 1983). While the syndrome has initially been described only in the setting of functional illnesses, over the last two decades there have been several reports of an association between Capgras syndrome and organic pathology (Weston and Whitlock, 1971; Christodoulou, 1977).

Here, we report a case of Capgras syndrome in a patient with disordered copper metabolism and extrapyramidal symptoms.

CASE REPORT

The patient, a 24 year old female graduate, came from a well to do family, and had a well adjusted pre-morbid personality. Detailed questioning did not reveal any history of neurologic or psychiatric illness among relatives or of consanguinity in her parents.

At the time of admission, patient had a 3½ years history of tremors in the right upper limb, followed by tremors in the right lower limb for the past six months. Four months prior to admission, she stated showing psychiatric symptoms of suspiciousness, social withdrawal, occasional bouts of irritability and restlessness and neglect of personal care. Her sleep and appetite were disturbed. For the past one month, she started saying that her parents were not real but imposters coming disguised as her parents. The belief could not be shaken by rational arguments but remained confined to her parents and she was able to recognise other relatives and acquaintances normally. There was no history of Schneider’s first rank symptoms, hallucinations or a sustained change in mood. There was no history of seizures, confusion or disorientation nor history suggestive of hepatic dysfunction.

Physical examination revealed rapid coarse tremors of the right upper and lower limbs associated with jerky movements, more on the distal parts, more on intention and disappearing during sleep. Muscle tone was increased with cogwheel rigidity on the right side. Muscle mass, power, sensations and deep tendon reflexes were normal with planters extensor bilaterally. There was no hepatosplenomegaly and other systems were normal. On mental status examination, patient showed hostility, social withdrawal, persecutory ideas, capgras delusion, delusions of reference and a blunted affect with no insight into her psychiatric problem. She was initially admitted in the neurology ward. The urinary copper was found to be 190 μg per 24 hours (normal: 10-80 μg) and the patient was started on D-pencillamine 1 gram/day and L-dopa, Carbidopa (25 mg, 250 mg) twice daily. With this there was an improvement in tremor and rigidity, but the psychiatric symptoms showed no improvement and the patient was transferred to psychiatry ward for management.

After stating D-pencillamine, the urinary copper levels increased 3-fold (from 190 μg to 600 μg per day) and this high level was maintained in subsequent testing. Serum copper was low at 33 μg/dl (normal: 60-150 μg.) All the tests for liver function and

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other biochemical investigations were within normal limits. K-F ring was absent on ophthalmological examination. X-ray skull, EEG and CT scan of the head were found to be within normal limits. A biopsy of the liver showed normal architecture and special stains for copper and copper binding proteins were negative. She had an IQ of 70 on full scale of WAIS and a memory quotient of 50 on the Wechsler Memory Scale.

The patient was started on 200 mg thioridazine per day which was increased to 700 mg per day over 3 weeks. With this, there was an improvement in psychiatric symptoms and the Capgras delusion disappeared in about 2 weeks. When discharged 6 weeks after entering the psychiatry ward, patient’s psychiatric symptoms were markedly improved with a slight worsening of the extrapyramidal symptoms. When seen at follow-up one month later, patient was taking 400 mg thioridazine per day and the improvement in psychiatric symptoms was maintained.

DISCUSSION
The earlier views of Capgras syndrome as a functional illness without organic etiology and explainable on a psychodynamic basis (Enoch et al., 1967) have been challenged by the subsequent reports associating the syndrome with organic pathology. In two separate reviews of the literature, Capgras syndrome was found to be associated with organic factors in 14 out of 46 cases (Merrin and Silberfarb, 1976) and 31 out of 133 cases (Berson, 1983) respectively. In a series of 11 patients with Capgras syndrome, Christodoulou (1977) reported abnormalities on neuropsychological testing in all of them. While a significant association with organic factors has been documented in literature, the organic factors in themselves seem neither necessary nor sufficient to explain the particular and peculiar content of the delusion (Berson, 1983).

In our patient, even though EEG and brain scan showed no abnormality, the presence of extrapyramidal symptoms points to an organic lesion in the brain. Cummings (1985), in a review of literature of patients with delusions and an associated structural disorder of the CNS, found delusional beliefs to be particularly common among those with extrapyramidal disorders.

Our patient did not show hepatic or corneal involvement which are considered typical of Wilson’s disease presenting with neurological symptoms. However, there was clear evidence of disordered copper metabolism. Copper is a trace element of importance in the functioning of the CNS. The role of copper is possibly of importance in the formation of myelin (Beisel and Pekorek, 1972) and in the enzymatic activity of dopamine-B-hydroxylase which converts dopamine to norepinephrine (Prohaska and Wells, 1974). Nicolson et al. (1966) reported symptomatic improvement in schizophrenics treated with the copper chelating agent, D-pencillamine. But the role of copper in the etiology of schizophrenia has been controversial with some studies having demonstrated elevated copper levels in schizophrenics (Chug et al., 1973; Olatunbosun, 1975) while others have found ceruloplasmin levels of schizophrenics to be similar to those of controls (Horwitt et al., 1957).

In our patient, either the organic brain lesion is responsible for the extrapyramidal symptoms, or the disordered copper metabolism by itself, could have played an etiological role in the production of psychiatric symptoms. The third possibility, that the psychiatric problem is purely coincidental and unrelated to the organic factors, appears less likely in view of the good premorbid personality, absence of family history of psychiatric illness and the temporal association between the onset of psychiatric manifestations and physical symptoms.

The findings of this case strengthen the evidence that organic factors can play a
causative role in the production of Capgras syndrome. When seen in conjunction with the importance of copper in the functioning of the CNS, as well as its suggested importance in schizophrenia, our findings highlight the need for further research into the role of copper in the etiology of psychiatric disorders.

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