MOYAMOYA DISEASE (PRESENTING AS SCHIZOPHRENIA)

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SUMMARY

The aetiology of Schizophrenia is still obscure. This case report is of interest because it draws attention to a hitherto neglected area in this field, i.e., the role of cerebro-vascular malformation and the resulting insufficiency in the cerebral circulation as a possible factor in the pathogenesis of Schizophrenia.

Schizophrenia is considered as a group of diseases of varied aetiology, but with similar clinical profile. Search for biological causes for the disease has produced several possible factors, ranging from variants of temporal lobe seizures to alterations in the biochemical parameters. Cerebro-vascular defects have been rarely implicated. However, this case report raises the interesting possibility that vascular defects of the brain may sometimes manifest as Schizophrenia.

CASE REPORT

A young male aged 11 years was brought to a Command Military Hospital, on 8 July '80 with the history of muscle stiffness, tremors and other involuntary movements of 8 days duration. History revealed that the boy was on phenothiazine medication, prescribed by a psychiatrist for violent and abusive behaviour. It was reported that the boy had been apparently normal till about 2 months previously. He was doing well in class and played games normally. Then he developed fever with giddiness, headache, restlessness and involuntary jerky movements of hands and legs. Although the fever subsided, the restlessness increased, and he became violent, stubborn and abusive. The psychiatrist diagnosed the case as Schizophrenia and started medication with chlorpromazine and Trifluoperazine. Within 8 days, the condition had deteriorated and he was brought to this hospital for admission.

There was no history of fits, head injury or of any serious infections in the past. Milestones were normal and he was a fairly bright student in school. He played games and mixed well. Family history was not contributory.

On examination, the sensorium was clear. Speech was slurred and there were marked E. P. signs. Other systems of the body were normal.

Psychiatric examination revealed a restless, uncooperative patient with neglected hygiene. Rapport could not be established. He spoke irrelevantly and incoherently. He laughed without apparent reason. Insight and judgement were impaired. He ate little and slept poorly.

The phenothiazines were withdrawn. The Bender-Gestalt test and Rorschach test suggested Organic brain syndrome. The EEG showed slow waves in the right frontal and temporal regions. Thereafter, bilateral carotid angiography followed by vertebral angiography was done.

The angiographic studies revealed bilateral occlusion of the Internal Carotid arteries at their superior clinoideal portions (distal anterior choroidal arteries), with basal ganglion telangiectasia. The anterior cerebral artery was not visualised. Collaterals from the anterior ophthalmic arteries and the occipital arteries of the External carotids were well seen. This radiological picture gave the diagnosis of Moyamoya.
disease.

The patient was started on Pheno- barbitone and showed improvement. At present, he is cheerful and cooperative and speaks relevantly. At times he is restless and impulsive. The E. P. symptom have disappeared. The restlessness is controlled with chlorpromazine. There has been no hemiplegia.

DISCUSSION

The term "Moyamoya" was first used by Japanese neurologists in 1965 to describe a cerebro-vascular disorder in Japanese children. Moyamoya means "a hazy screen of smoke trailing or floating", and describes the radiological picture of the bilateral, symmetrical vascular network developing directly from the cartoid syphon at the base of the brain, resulting from occlusion of both the Internal Carotid arteries at that region. In recent years a number of non-Japanese cases have also been reported. Cases from India have been reported by Acharya et al. (1974), Ahuja and Gupta (1976) and Rana et al. (1979).

The classical clinical picture in children that has been described is one of alternating and progressive hemiplegia. Suzuki and Takaku (1969) described six stages in the progress of this disease. In adults it may present as subarachnoid haemorrhage. It has been described more commonly in females. The aetiology is obscure. Yoda et al. (1965) suggested a genetic origin on the basis of detection of an abnormal long Y sex chromosome. Sogaard and Jorgensen (1975) observed a familial occurrence. Other factors that have been postulated are idiopathic arteritis, infections, radiation therapy and tumors in that region. The "moyamoya" itself has been considered to be a primary vascular malformation by Nishimoto and Takeuchi (1968). Others have considered it to be dilated collateral vessels developing in order to by-pass the arterial occlusion.

Whatever be the aetiology, the disease signifies a disturbance in the cerebral circulation, which may present as a cerebral stroke, or abnormal functioning of the brain. When one considers the site of the lesion, it is possible that it can manifest as abnormal behaviour which can resemble Schizophrenia.

For treatment of the Moyamoya disease, micro-surgical anastomosis between the superficial temporal and middle cerebral arteries appear to give promising results.

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