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Moyamoya disease in a 12-year-old Caucasian boy presenting with acute transient psychosis

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Abstract This paper reports the case of a 12-year-old boy who presented with an acute transient psychosis. Investigations revealed Moyamoya disease with occlusion of the left middle cerebral artery. There was no previous history of transient ischaemic attacks, no history of psychiatric disease, no family history of psychosis and no history of illicit drug use. Although no previous

cases of Moyamoya disease with psychosis as the sole presenting feature have been described, we suggest the possibility of a causal link.

Key words Moyamoya disease – psychosis – adolescent – temporal lobe pathology

Introduction

Moyamoya (Japanese for ‘cloud of smoke’) is a cerebrovascular disease caused by segmental stenosis or occlusion of one or both internal carotid arteries. This leads to a network of small anastomotic collaterals visible on carotid angiograms. The aetiology is uncertain and it is assumed to be related to an acquired lesion of the blood vessels at the base of the brain (8). The main presentations of the disease, which has been described predominantly in Japan, are hemiparesis, localised muscular weaknesses, headaches, or seizures, first appearing during childhood and adolescence. The nature of the disorder is uncertain and there are probably several causes for the characteristic radiological appearance. Complications include mental deterioration, recurrent strokes, or intracranial haemorrhage (7). Transient ischaemic attacks are commonly seen and seem closely related to hypoxia after hyperventilation (5).

Psychotic symptoms as the sole manifestation of Moyamoya disease have, to our knowledge, not been described to date. However, McDade (6) described a 19-year-old Asian man suffering from Moyamoya who presented with a schizophrenia-like illness. This man had a strong family history of Moyamoya, was attend-

ing a school for those with moderate learning disability and had, at the ages of 6 and 7 years, suffered episodes of right hemiplegia caused by his illness.

In the present paper we report the clinical case of a 12-year-old boy with no family history of schizophrenia and no previous psychiatric or neurological history who presented with an acute psychosis. He was subsequently diagnosed as suffering from Moyamoya disease. The possibilities for a causal link will be discussed.

Case

Family History

The boy lived at home with both parents and his 16-year-old brother. Both parents were middle class professionals and were of European origin. There were no extraordinary stresses in the family or social situation prior to the onset of the symptoms and family relationships were described as good. The only family history of psychiatric illness reported was that the paternal grandfather had had a “nervous breakdown” at some point. Details of that illness were not known, except that the problems had not led to discharge from the Army.

Personal History

The boy was born after a normal pregnancy and delivery. His neonatal period was uncomplicated and developmental milestones were normal. He was raised bilingual. When he had started speaking, he had required a short period of speech therapy to help with pronunciation. He had attended mainstream schools throughout and his attainments were usually in the average range, although he had been slow to learn reading and spelling during his primary school years. He enjoyed sports and art, did not feel overly pressured at school and had good peer relationships. His pre-morbid personality, as described by his mother, was "very sweet, very sensitive, and strong-willed." He had no history of serious illnesses, allergies, or operations. In particular, he had not experienced any episodes of dizziness, of transient hemiplegia, or seizures. He denied any illicit drug use, was not smoking cigarettes, and was not receiving any medication.

Presentation

His parents reported that, nine days before his admission to hospital, their son had taken part in a 1500-metre run at school, which had considerably exerted him. From then on, his behaviour had become increasingly disturbed. He had experienced difficulty sleeping and had been staying up all night, watching television. He had become very talkative and been worrying about philosophical matters, the brain, brain chemicals, and his sexual orientation. His speech and actions had not always made sense to his family. On one occasion, the boy had defaecated in his bed, but he had been unconcerned about this, saying that he was experimenting. His mood had become increasingly changeable, ranging from periods of sadness and tearfulness to periods of elation, agitation, and anxiety.

On mental state examination, the boy appeared frightened and, at times, confused. He was preoccupied by persecutory beliefs although it was difficult, at initial presentation, to clarify these. He referred repeatedly to his sexuality and to his physical well-being. He had visual and auditory hallucinations, stating that he was hearing laughter and babies crying and seeing dolls everywhere. Later on, he said that he believed that his mother had been replaced by another person, that people were secretly observing and camcording him, that he was being hypnotised, that he was being sent messages through the television, and that he was the subject of an experiment. He also described how his senses, especially his sense of smell, had become more acute. He knew his age and year of birth, but was unable to state his full date of birth.

On physical examination, his temperature was 37.8 C, although this returned to normal within 24 hours of hospital admission. He was right-handed. No physical or neurological abnormalities were detected and, in particular, there were no signs of hemiplegia or muscular weakness. Fundi were normal.

Medical investigations

Investigations, including full blood count, urea and electrolytes, liver function tests and thyroid function tests were normal. Blood culture and toxicology, including tests for volatile substances, were negative. Cerebrospinal fluid investigations excluded bacterial infection, and polymerase chain reaction for Herpes simplex and enterovirus were negative. A urine drug and toxicology screen elicited no compounds except benzodiazepines, which had been given on admission. An EEG examination showed no abnormality.

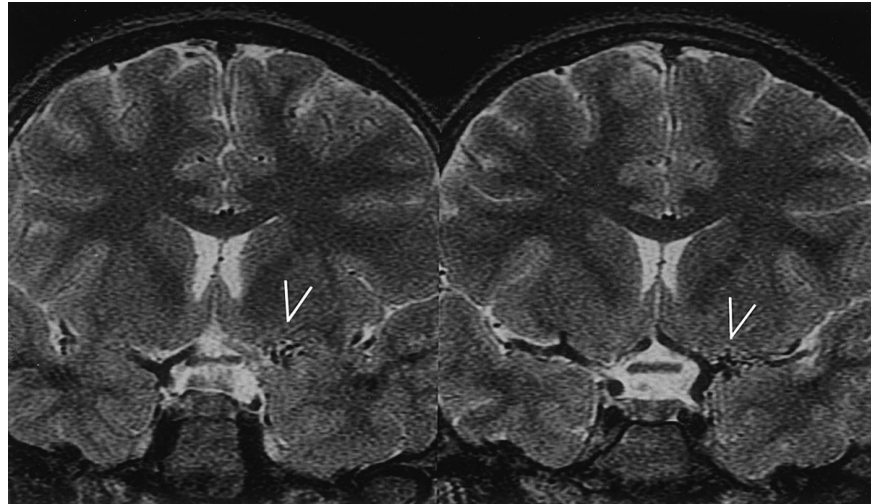
The neuroradiological investigations revealed the diagnosis of Moyamoya disease. On an initial MRI scan (Fig. 1), no abnormality was detected in the hemispheres. However, in the region of the left basal ganglia, multiple irregular signal voids indicative of small collateral vessels were seen, where the middle cerebral artery should have been. A subsequently performed MRA (Fig. 2) contrasted well the abnormal left-sided vasculature, when compared to the normal right side. The proximal middle cerebral artery is absent on the left. The left internal carotid angiogram (Fig. 3) showed that the missing vessel had been replaced by multiple small collateral vessels, filling the distal middle cerebral artery branches. These form the typical 'cloud of smoke'.

Neuropsychological assessment

The following tests were performed after the psychotic symptoms had resolved:

- Wechsler Intelligence Scale for Children (WISC-III-UK): Verbal, Performance and Full Scale Scores were all within the normal range, with no significant differences between scores;
- Wechsler Objective Reading Dimension (WORD): Reading and comprehension were within the normal range. Spelling was in the low average range;
- Wide Range Assessment of Memory (WRAML) (verbal learning and story memory subtests): Most subscales were in the normal range except for delayed recall of a list of words which was weak, falling within the borderline range;
- Parts IV and V of the Token Test for screening of language function: Each score fell within the average range;

Fig. 1 T2 weighted MRI showing multiple irregular signal voids in the region of the left basal ganglia (arrow). These are indicative of multiple collaterals in place of the left middle cerebral artery



- Executive functioning tests: The verbal fluency tests showed performance in the average range and the Trail Making Test showed impairment in both Part A and Part B. This suggests visual scanning difficulties.



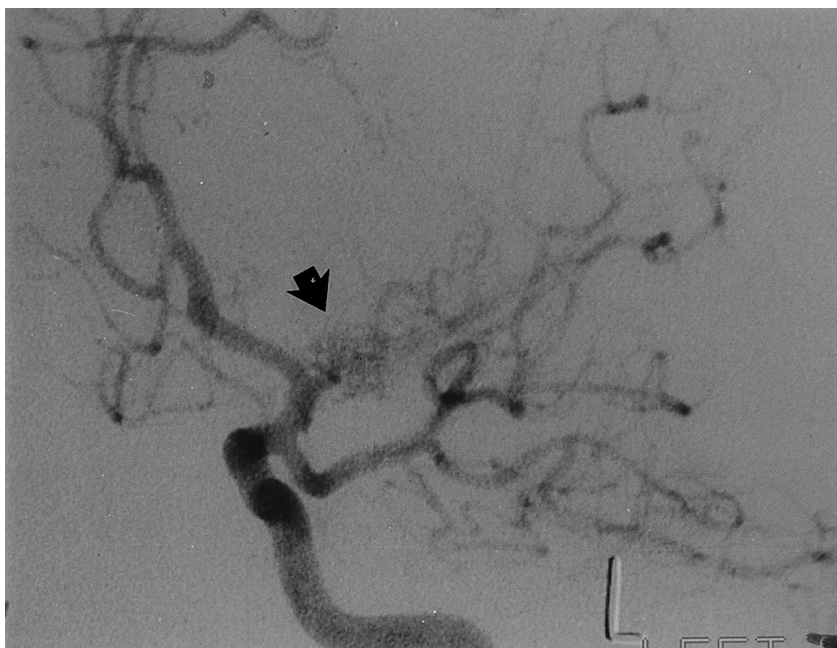
Fig. 2 MRA showing absent left proximal middle cerebral artery (arrow) compared to normal right-sided vasculature

Thus the scores generally fell within the average range. The only impairment noted appeared due to visual scanning difficulties. Such a difficulty might be expected with frontal lobe lesions, but is not normally related to left temporal difficulties. It is possible that speech, reading, and spelling problems encountered in early childhood might have been part of a language problem secondary to temporal lobe functioning abnormalities, but the test findings do not indicate any consistent impairment in the area.

Treatment and outcome

The boy was initially sedated with 2 mg diazepam, before treatment was commenced with low doses of chlorpromazine. Due to dystonic reactions, chlorpromazine was discontinued and trifluoperazine 2 mg daily commenced on the third day of admission. There was a marked decrease of psychotic symptoms over the following 48 hours. His condition improved further over the following 10 days and medication was reduced to 1 mg trifluoperazine daily. During this time the boy and his family were seen regularly to discuss this traumatic experience and to help them to cope with associated anxieties. The boy was able to return to school three weeks after admission. By then he showed no psychotic symptoms and his parents reported that he was “100% recovered.” Medication was discontinued three months after admission and the boy was advised not to exercise heavily. In view of the transient nature of symptoms and the absence of lasting functional impairment, further neurological and neurosurgical interventions were not recommended. At one-year follow up, no further neurological or psychiatric disturbances were reported and the boy was functioning well, both socially and academically.

Fig. 3 Left internal carotid angiogram showing that the missing proximal middle cerebral artery has been replaced by a network of small collaterals (arrows) which fill the distal middle cerebral artery branches



Discussion

Despite the fact that psychotic symptoms as the sole manifestation of Moyamoya have not been previously described we suggest the possibility of a causal link, on the basis of the location of the cerebral lesion, the physical exertion with associated hyperventilation that preceded the psychosis, and the improbability of the two events occurring independently.

There is a growing body of evidence in support of the view of schizophrenia as a dysfunction of the left temporal lobe (7). The association, first proposed by Flor-Henry (2), stemmed from the frequently observed association of schizophrenia-like illness and temporal lobe epilepsy. However, schizophrenia-like symptoms have also been reported in patients with temporal lobe dysfunctions without epilepsy. For example, Buckley et al. (1) found that in three patients developing schizophrenia-like symptoms after cerebral trauma, the abnormalities were confined to or included the left temporal lobe, while this was not the case in controls with schizoaffective symptoms. In our patient, the site of the lesion in the dominant left temporal lobe and the psychotic symptoms observed, complement existing findings.

The psychotic phenomena in this boy started in the summer, shortly after he had exerted himself in a 1500-metre run. A number of reasons may explain this mechanism. It might be that, while blood supply through the collaterals was sufficient at rest or during moderate exercise, this was not so during a period of heavy exertion. The possibility of slight dehydration

should also be considered. In addition, several authors have described how hyperventilation may induce a decrease in the concentration of oxy-haemoglobin and have found it closely related to ischaemic attacks in children with Moyamoya disease (3, 4, 5).

Although pre-pubertal schizophrenia and bipolar disorder are rare, by the teenage years, particularly with males, these illnesses are seen with increasing frequency. A drug-induced psychosis is also possible and, as the boy only presented nine days after the beginning of the symptoms, the negative blood and urine test cannot fully exclude all illicit substances taken at symptom onset. However, in this case, there was no previous psychiatric history and no clear family history of psychosis. Symptoms were very transient and did not reoccur at one-year follow up, even after medication was stopped. The patient's history and his own account, as well as those of his parents and his school, give no indication for any illicit drug use. Thus, whilst it is possible that this boy had a disorder on the mood spectrum associated with psychotic features or a disorder on the psychotic spectrum, we suggest that, as both Moyamoya and psychosis in pre-pubertal children are rare disorders, it would seem very unlikely that we are dealing with two independent phenomena.

We thus conclude that, although Moyamoya disease is a relatively rare disorder in Europe, it should perhaps be considered as a possible cause for psychosis in children, especially – though, as this case shows, not exclusively – in those with a history of previously unexplained transient ischaemic attacks or brief episodes of hemiplegia.

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