

Middle Cerebral Artery Dissection

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Abstract

A 12-year-old girl had minor head trauma, with resultant mild headache, one day prior to onset of an apparently generalized tonic clonic seizure, right hemiparesis and dysphasia. A cranial CT scan showed evidence of a left middle cerebral artery infarct. Despite ventilation, fluid restriction, mannitol, thiopental infusion and intracranial pressure monitoring she deteriorated and died. Autopsy demonstrated dissection of the left middle cerebral artery with an intact internal carotid artery and no evidence of vasculitis. Middle cerebral artery dissection in children is very rare. Most reported cases are diagnosed at autopsy. The pathogenesis of cerebral artery dissection may include preceding minor head trauma or exertion and this should be sought for in the history.

Key words

Childhood stroke – Cerebral artery dissection

Middle cerebral artery (MCA) dissection in children is rare. We present this case to demonstrate the difficulty in diagnosis and to increase awareness of this as a cause of ischemic stroke.

Case report

A 12-year-old girl presented with a seizure, right hemiparesis and reduced level of consciousness. The day prior to the seizure she had minor trauma, hitting her head on a coffee table, without loss of consciousness. This resulted in a small abrasion and headache but she continued to have an active day. The following day she had an apparently generalized 10-minute tonic-clonic seizure. Subsequently she was drowsy and confused with dysphasia and a right hemiparesis.

She had previously been well with no history of seizures, congenital heart defect or evidence of a coagulopathy. She was taking no medication. There was no family history of seizures or thrombotic disorder.

An initial cranial computerized tomography (CT) scan a few hours after seizure onset showed early evidence of a left MCA infarct with hypodensity in this region. On

admission she had blood pressure 160/100 mmHg and heart rate 88/min. No cardiac or carotid or cranial bruits were heard. She was intubated but was able to respond appropriately to her parents' faces and requests. She had a gaze preference to the left and a right hemiparesis involving the face, arm and leg. Examination of the cerebrospinal fluid showed protein 0.25 g/l, no WBC, $14 \times 10^6/l$ RBC and negative bacterial and viral culture. Blood culture was sterile. Serum electrolytes, glucose, calcium, urea and creatinine were normal. Blood count was normal apart from slightly reduced platelets at $133 \times 10^9/l$. Prothrombin time and activated partial thromboplastin time were normal. Sedimentation rate was 4 mm/hr and anti-nuclear antibody (ANA) and anti-cardiolipin antibody were negative. Antithrombin III (chromogenic and antigen) and protein S antigen (total and free) were normal. Protein C activity and antigen were both borderline low. Prothrombin antigen, thrombin time and plasminogen were normal. Plasma amino acids, cholesterol and triglycerides were normal and no abnormalities were detected on electrocardiogram and transthoracic echocardiogram. Electroencephalogram (EEG) showed marked slowing over the left hemisphere and normal right hemispheric activity.

She initially improved slightly but 29 hours after her seizure she deteriorated with a drop in heart rate, dilatation of the left pupil and reduced level of consciousness. On examination she did not have papilledema but venous pulsations of the left optic disc were absent. Extraocular movements and corneal reflexes were present. She had response to pain on the left side only. A repeat CT scan at that time showed a much larger area of infarction with midline shift (Fig. 1). She was treated with mannitol, fluid restriction, hyperventilation and intracranial pressure monitoring with thiopental infusion to produce a burst suppression pattern on EEG. She was not given anticoagulants. A repeat cranial CT scan 2 days later showed further increase in the area of infarction and midline shift and effacement of the basal cisterns. She continued to deteriorate, developing diabetes insipidus. Five days after her admission she was brain dead with absent cerebral perfusion on nuclear brain scan.

At autopsy sections from all the cerebral arteries were examined. These demonstrated dissection of the left MCA and no involvement of the internal carotid artery (ICA) (Fig. 2). The smooth muscle of the media was elsewhere histologically unremarkable with no evidence of vasculitis. There was infarction of most of the left cerebral hemisphere with cerebral edema, cerebellar tonsillar herniation and ischemic necrosis of the anterior lobe of the pituitary.

Discussion

Seven children with intracranial artery dissection were reported recently (8). All patients had an ischemic

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Fig. 1 Cranial CT scan 29 hours after her seizure. This shows a large hypodense area representing infarction of the left middle cerebral artery distribution (arrow). There is midline shift to the right and effacement of the left lateral ventricle.

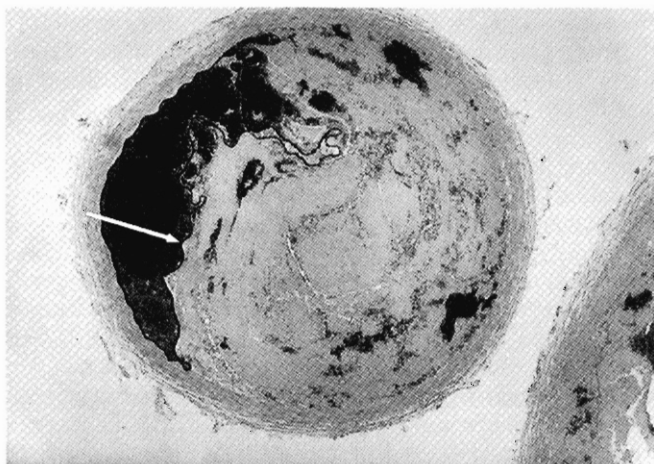


Fig. 2 Cross section of the left middle cerebral artery showing acute dissection with elevation and displacement of the internal elastic lamina (arrow) by erythrocytes, fibrin and entrapped neutrophils. The lumen is virtually obliterated by this process. This is a *van Gieson's* elastic stain with original magnification X 25.

stroke preceded by headache. Seizures occurred in 2. Physical exertion preceded onset in 3 and one had hypertension and coarctation of the aorta. Angiography in all showed an elongated area of irregular stenosis. Treatment consisted of antiplatelet therapy in 2 and supportive measures in 5. One boy died and autopsy confirmed dissection of the right distal ICA and proximal branches of the right MCA. In autopsy reports of children with dissection of the MCA or distal ICA in all cases the plane of dissection is between the internal elastic lamina and the media, rather than within the media as is seen with cervical carotid and aortic dissection (1, 3, 4, 5, 7). Minor preceding trauma, as in the present case, is often noted. It has been

suggested that trauma may predispose a minor congenital defect to dissection (9). This may add to the increased incidence of intracranial artery dissections in the younger age group (8). Also reported are gap defects in the internal elastic lamina near the site of dissection in adults and this author postulated that the gap defects underlies the spontaneous dissection in arteries (12). Primary arterial diseases such as fibromuscular dysplasia have been suspected or described although some patients have no pathological change (6).

Anticoagulation therapy is usually recommended in patients with ischemic symptoms in the absence of intracranial hemorrhage or an already completed major infarct. Intracranial artery dissections usually have diagnosis made at postmortem, reflecting the difficulty of making an antemortem diagnosis and instituting therapy. This may account for the poorer prognosis than is seen in extracranial dissections. Early angiography has been suggested in childhood stroke to assess for dissection and consider anticoagulation (10). In retrospect in this case the minor head trauma and headache were clues to the etiology and an angiogram may have given the diagnosis prior to death. However the infarct was large and already completed at the time of the initial CT scan and anticoagulant therapy would have been contraindicated. Surgical intervention and repair of the aneurysm is generally reserved for patients with a subarachnoid hemorrhage due to an intracranial dissecting aneurysm or those with embolus from an extracranial dissecting aneurysm (8).

In conclusion it is important to remember intracranial artery dissection as a cause of stroke in children especially in the context of minor head trauma or exertion accompanied by headache. Early angiography may be helpful in these circumstances to guide management.

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