Cervical arterial dissection and ischaemic stroke in children: two cases.

UNIVERSITÀ DEGLI STUDI DI TORINO

Received Date : 01-Nov-2012
Revised Date : 04-Dec-2012
Accepted Date : 05-Dec-2012
Article type : Letter

CERVICAL ARTERIAL DISSECTION AND ISCHEMIC STROKE IN CHILDREN: TWO CASES.

Manuela Agostini*, MD; Maria Maddalena Lupica*, MD; Michael Mostert, MD*; Nicola Deluca**, MD; Luca Cordero di Montezemolo*, MD, PhD.

*Department of Pediatrics, Regina Margherita Children's Hospital, University of Turin, Italy

**Department of Neurology, Degli Infermi Hospital, Rivoli, Italy

Acknowledgements: we are grateful to the Department of Neuroradiology, S.G. Battista Hospital and Regina Margherita Children's and CTO Hospital, for performing the neuroradiological examinations.

FINANCIAL DISCLOSURE: The authors have no financial relationships relevant to this article to disclose.

Corresponding author and address reprint requests to:

Manuela Agostini, MD

Department of Pediatrics

Regina Margherita Children's Hospital

Piazza Polonia, 94

10126 Torino, Italy

Tel: +390113135840

FAX: +390113135108

E-mail: manuela.agostini@unito.it

Sir,

Cervical arterial dissection (CAD) is an important cause of childhood arterial ischemic stroke (1). Dissection occurs following a vessel wall tear or rupture of the vasa vasorum with formation of an intramural hematoma. Secondary thromboembolism in the vessel territory can cause an ischemic stroke. Dissections may be traumatic or spontaneous. Trauma ranges from severe (direct neck trauma, rapid deceleration or hyperflexion), to trivial (vomiting or coughing). Pathological conditions (inherited connective tissue disorders, fibromuscular dysplasia, infection) may underlie spontaneous or minor trauma dissections (1, 2). CAD presentation may be subtle with neck pain, headache, TIA and cerebral ischemia. In paediatric ischemic stroke, arterial magnetic resonance angiography (MRA), including neck vessels study, must always be performed as first-line investigation to exclude arterial dissection (3). MRI should include, beside diffusion weighted imaging (DWI), fluid-attenuated inversion recovery (FLAIR) and T2-T1 images of the brain, also T1 or T2 fat-saturation axial imaging through the neck (3-4). When MRI/MRA is equivocal or the child has a recurrent event and dissection is highly suspected imaging workup should be completed with a contrast-enhanced MRA, CT angiography (CTA) and/or conventional angiography (CA) on a case-by-case consideration (3). Pathognomonic arteriographic CAD features include an intimal flap or double-lumen sign, irregular narrowing and tapered, flame-like occlusions (1-4). Immediate anticoagulation is required in extracranial dissection (3, 4). We report two CAD cases:

Case 1

A 7-year-old boy presented at our Emergency Department after two days of left temporal-occipital headache and visual trouble, lasting about two hours, following repeated vigorous somersaults. He was afebrile and physical and neurological examination were negative. He had never had headaches before. The first laboratory exams were negative. Head CT showed a right hypodense temporo-occipital lesion compatible with white matter lesion or ischemia. Two days after admission the child complained of headache, neck pain, dizziness and vomiting. Neurological examination revealed ataxia, left dysmetria, nystagmus, left homonymous hemianopsia and lethargy. Encephalitis/ADEM, cerebral masses, stroke or stroke-like episodes were suspected. MRI showed hyperdense lesions in both cerebellar lobes (Fig. 1A), the right temporal-occipital hemisphere and the right thalamus, with T2 and FLAIR sequences. DWI showed restricted diffusion in the corresponding areas of T2 and FLAIR abnormalities. Clinical and neuroradiological picture suggested ischemic strokes in the vertebrobasilar territory. Acute onset of neurological symptoms after repetitive mild cervical trauma and worsening after a symptom-free period (96 hours), suggested recurrent posterior circulation thromboembolism possibly secondary to arterial vertebral dissection. Immediate head and neck vessels MRA did not visualize the left vertebral artery (Fig. 1B) strengthening our hypothesis of dissection and showed cervical internal carotid arteries asymmetry. Trans-thoracic heart echography excluded cardioembolism. CTA showed left vertebral artery occlusion at C2, occlusions of the left posterior inferior cerebellar artery and right posterior cerebral artery, compatible with vertebral artery dissection and secondary thromboembolism in the posterior cerebral circulation. Homocysteine, thrombophilic, virologic and batteriologic screening, ammonium, lactate and antinuclear antibodies, were normal. Guarded rachicentesis was negative. Immediate heparin treatment and neurosurgical monitoring were begun. On day fifteen of hospitalization, transfemoral angiography showed

left vertebral artery dissection at C2 (Fig 1C), located the intracerebral vessel occlusions and revealed that the right vertebral artery and bilateral carotid arteries were normal.C2 is an area of arterial vulnerability to trauma due to rotation at the atlantoaxial junction and lack of bone protection to lower segments. Possibly, repeated forceful neck motion caused stretching and compression of the left vertebral artery at the atlantoaxial junction promoting dissection. Cervical spine X-ray and CT showed no bone abnormalities. Skin or joint laxity was not observed but genetic and bioptic collagen studies were not performed. Oral anticoagulation for six months was performed and at present the patient is still on anti-platelet treatment. This case is interesting for its subtle presentation with headache and long-lasting visual impairment which excluded migraine, in a child with history of repetitive mild cervical trauma.

Case 2

A 2-year-old boy with Down syndrome presented at our Emergency Department for sudden irritability and right-sided weakness. Surgical closure of a ventricular septal defect had been performed at 7 months of age. On admission he had mild nasal inflammation and a temperature of 37°C. He had a thoracic scar, irritability, left-side gaze, right-side facio-brachio-crural hemiparesis joint laxity and no signs of traumatism. Cardiac, respiratory and abdominal findings were normal. Meningoencephalitis or stroke were suspected. Cerebral MRI showed hyperdense lesions in the left temporo-parietal lobes with T2-weighted and FLAIR sequences corresponding to the entire left middle cerebral artery (MCA) territory. DWI showed restricted diffusion in the corresponding areas of T2 and FLAIR abnormalities compatible with ischemic stroke. MRI and sudden onset of symptoms suggested a thromboembolic infarction in the left MCA territory involving the cortex and basal ganglia (Fig. 1D). MRA imaging confirmed left MCA origin occlusion (Fig. 1E). Intracerebral

vascular dysplasia was not observed (5). Trans-thoracic and trans-oesophageal echocardiography excluded intracardiac thrombus and residual shunts associated with paradoxical embolism. A 24-hour holter excluded rhythm abnormalities. A hemogram, flogosis index and blood chemistry were normal as were immunologic, metabolic, virologic and thrombophilic screening. Supra-aortic vessels MRA and CTA showed irregular narrowing of the left cervical internal carotid artery (ICA) compatible with dissection (Fig. 1F) and secondary thromboembolism. A cervical spine X-ray excluded atlantoaxial instability. Anti-platelet treatment was performed because of the large ischemic area and risk of bleeding with anticoagulation (3) and at present the patient is still on anti-platelet treatment. We are unaware of reported Down cases with cervical ICA dissection. In spontaneous CAD environmental and intrinsic factors are involved. In this child with mental retardation violent head hyperextension-rotation episodes had occurred in the weeks before admission and during hospitalization. In children cervical ICA dissection is more commonly traumatic than intracranial ICA dissection (1). Rapid hyperextension leads to carotid artery stretching over the cervical vertebrae. The predisposition for hypotonia, ligamentous hyperlaxity and primary angiopathies along with airways flogosis of this patient increased the risk for dissection (2,5,6). Head hyperextensions that had begun some weeks before dissection may have triggered the event. Neither case 1 nor 2 relapsed after six and four years respectively. Clinicians must exclude arterial dissection in pediatric ischemic stroke and be aware of specific child-related traumatic triggers.

REFERENCES

1) Rafay MF, Armstrong D, Deveber G, Domi T, Chan A, MacGregor DL. Craniocervical arterial dissection in children: clinical and radiographic presentation and outcome. *J Child Neurol* 2006; 21:8-16.

2) Schievink WI. Spontaneous dissection of the carotid and vertebral arteries. *N Engl J Med* 2001; 344:898-906.

3) Stence NV, Fenton LZ, Goldenberg NA, Armstrong-Wells J, Bernard TJ. Craniocervical arterial dissection in children: diagnosis and treatment. Curr Treat Options Neurol. 2011 Dec; 13:636-48

4) Roach ES, Golomb MR, Adams R, Biller J, Daniels S, Deveber G, et al. Management of stroke in infants and children: a scientific statement from a Special Writing Group of the American Heart Association Stroke Council and the Council on Cardiovascular Disease in the Young. *Stroke* 2008; 39:2644-91.

5) Worley G, Shbarou R, Heffner AN, Belsito KM, Capone GT, Kishnani PS. New onset focal weakness in children with Down syndrome. *Am J Med Genet A* 2004; 1:15-8.

6) Fleisher GR, Buck BE, Cornfeld D. Primary intimal fibroplasia in a child with Down's syndrome. *Am J Dis Child*. 1978;132:700-3.

FIGURE LEGEND

Figure 1: A-C, Case 1: cranial MRI demonstrating in the left and right cerebellar lobes focal FLAIR high signal (A black arrows). Vascular imaging (B-C) demonstrating lack of visualization of the left vertebral artery in MRA (B, black arrow) and an occlusion of vertebral artery at C2 in CA (C, black arrow).

D-F, Case 2: a large left infarction within the left MCA territory is showed by sequences with DWI, demonstrating restricted diffusion (D, 24 hours). Vascular imaging (MRA) shows reduction of the flow at the origin of the left MCA (E, white arrow) and an irregular narrowing of the left ICA just after the bifurcation (F, white arrow).

Figure 1