CLINICAL EVOLUTION AND FOLLOW-UP OF A CASE OF MOYAMOYA SYNDROME

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SUMMARY

Case report: We report the follow-up of a female infant with moyamoya syndrome whose initial manifestation was an episode characterized by tonic-clonic seizures. MRI and MRA showed the presence of multiple cerebral infarctions and ischemia of the temporal-parietal-occipital left area of the brain with severe cerebral hypotrophy of the left hemisphere and asymmetry of the ventricles. Neurosurgical treatment (EMS) was performed. After 4 years the patient presented an episode characterised by loss of the muscular tone without loss of consciousness after which she had generalized hypotonia. We believe that the crisis cannot be correlated to vascular pathology, but must be seen as neurocardiogenic pre-syncope.

Key words: Moyamoya disease, pre-syncope, encephalomyosynangiosis, tonic-clonic seizures

RIASSUNTO

Caso clinico: noi riportiamo il follow-up di una bambina con sindrome di Moyamoya che si è manifestata inizialmente con un episodio convulsivo tonico-clonico. La MRI e la MRA hanno evidenziato la presenza di molteplici infarti cerebrali e d’ischemia dell’area sinistra temporoparieto-occipitale con severa ipotrofia dell’emisfero cerebrale sinistro e asimmetria dei ventricoli. La paziente è stata sottoposta a trattamento neurochirurgico (EMS). Dopo 4 anni la piccola ha presentato un episodio caratterizzato da perdita del tono muscolare senza perdita di coscienza con successivo stato d’ipotonia generalizzata. Noi crediamo che la crisi non sia correlabile alla patologia vascolare, ma che vada inquadrata come lipotimia neuro-cardiogenica.

Parole chiave: Malattia di Moyamoya, lipotimia, encefalomiosinangiosi, convulsioni tonico-cloniche

Introduction

Moyamoya syndrome is a chronic cerebrovascular occlusive disease characterized by progressive stenosis of the terminal portions of the internal carotid arteries and of the anterior and middle cerebral arteries. The appearance of this vascular network on an angiogram looks like a puff of cigarette smoke drifting in the air; thus, in 1969, Suzuki and Takaku named this novel disorder “moyamoya disease”, because “moyamoya” means “puff of smoke” in Japanese.

We reported a case of moyamoya, which has been followed up for more than 4 years.

Case report

A. D., a 7-year-old female, is the first born of non consanguineous parents. She was born by normal delivery, following an uncomplicated pregnancy, with a birth weight of 3,350 g. The neonatal period was uneventful. Neuromotor development was reported to be normal.

At the age of 2 years and 6 months she was admitted to our department because she had an episode characterized by tonic-clonic seizures localized first to the right side of body then to the left arm, with loss of consciousness. This crisis lasted approximately 10 minutes, after which she had a residual alteration of the state of consciousness with lack of awareness and responsiveness to the environment.

The neurological examination at admission revealed the alteration of the sensorium, the hypomobility of the left side of the body and of the right arm, and hypactive left patellar reflexes; during hospitalization, twelve hours after admission, the patient was transferred to intensive care for an episode of bradycardia (45b/min).

After stabilization the patient showed hypotonic trunk, hypertonic upper and lower limbs and aphasia. During hospitalization, the following tests were performed: a brain CT-scan, a brain magnetic resonance imaging (MRI), and MR Angiography (MRA) that showed the presence of multiple cerebral infarctions and ischemia of the temporal-parie-
tal-occipital left area of the brain with severe cerebral hypotrophy of the left hemisphere and asymmetry of the ventricles, compatible with Moyamoya disease.

The diagnosis was confirmed, after a month, at the Pediatric Neurosurgery Unit of Polyclinic A. Gemelli of Rome, where angiography was carried out and showed bilateral stenosis of Internal Carotid Artery (ICA) in the supraclinoid region with abnormal collateral vascular networks and hypoplasia of the Anterior Cerebral Arteries (ACA) and Middle Cerebral Arteries (MCA) with hypoplastic and irregular Posterior Cerebral Arteries (PCA).

Encephalomyosynangiosis (EMS) was done on our patient, first on the right side and after 3 months on the left side. The postoperative course was uneventful and, after one year, MR angiography showed the development of collateral vessels from the external carotid artery especially on the left hemisphere [2]. During follow-up she had no ischemic attacks and no seizures.

After four years of follow-up, the patient returned to our observation because she had an episode of loss of muscular tone resulting in a fall without loss of consciousness; this crisis lasted approximately 5 minutes, after which she manifested generalized hypotonia.

At the physical examination she appears in good general condition. She weighs 2,400 g (50-75 \(^\text{th}\) pc), she is 126 cm tall (90th pc), and her Cranial Circumference is 49.5 cm (25th pc); she shows a fairly good social interaction, a mild delay in speaking, hypomobility of the right arm, and uncertain walking with left clubfoot. The routine laboratory tests are normal and the ECG shows sinus bradycardia and normal ventricular repolarization; the echocardiogram reveals normal heart valves and chambers and normal heart wall movement. The EEG showed generalized aspecific signs. The MRI-brain and angio-MRI show no changes compared with previous examinations performed during the follow-up. The image was characterized by asymmetric dilatation of the lateral ventricles, with prevalence of the left one.

**T2-weighted image reveals high-density lesions and FLAIR reveals hypo-density lesions in hypotrophic left hemisphere and in right parieto-occipital region. The MRI also shows temporal-parieto-occipital left region atrophy and atrophic lesions in the right hemisphere due to chronic cerebrovascular insufficiency. MRA shows a lower vascularization of the territory sprinkled by the middle and left posterior cerebral arteries and by the right posterior one, with hypoplastic left transverse sinus.**

**Discussion**

Moyamoya disease, first described in 1957 by Takeuchi and Shimizu as all bilateral hypoplasia internal carotid artery, is an uncommon cerebrovascular disorder that is characterised by progressive occlusion of the supraclinoid internal carotid artery (ICA) and its main branches within the circle of Willis.

The incidence of Moyamoya disease is high in Japan[4]. The prevalence and incidence are 3.16 and 0.35 per 100,000 Japanese persons, respectively. The female to male ratio was shown to be 1-8, and the distribution of age at onset has been suggested to have two peaks: one at 5 years of age and one lower peak at about 40 years of age[5].

The clinical features of Moyamoya disease differ substantially between children and adults. Most children with Moyamoya disease develop transient ischemic attack (TIA) or cerebral infarction, whereas about half of adult patients develop intracranial bleeding, and half develop TIA or cerebral infarction or both[6]. The patient has come to our observation initially for seizures, hemiparesis and alteration of the sensorium. She has been submitted to MRI that has shown multiple cerebral infarctions in different stages of evolution; also MRA has confirmed the diagnosis of Moyamoya syndrome. Neurosurgical treatment (EMS) gave good results. The postoperative course was uneventful; she has been examined once a month for the first six months, and once every six months from then on, showing no more clinical manifestations.

However, recently the patient presented an episode characterised by loss of the muscular tone without loss of consciousness; the crisis lasted approximately 5 minutes, after which she had generalized hypotonia. A second MRI and another MRA were performed and no new cerebral vascular abnormalities were found. Clinical examination and EEG features were within normal limits. We believe that the crisis cannot be correlated to vascular pathology, but must be seen as neurocardiogenic pre-syncpe. Clearly the clinical evolution and the follow-up will give us a more defined case history, but we believe that the most recent symptoms presented are not due to a relapse, in fact, as reported in the literature, surgery should be entirely conclusive.
On the basis of previous studies, surgical revascularization is thought to improve cerebral hemodynamics and reduce the incidence of subsequent ischemic stroke in both pediatric and adult patients. In pediatric patients, the incidence of TIA rapidly decreases or even disappears after surgery; furthermore, pediatric patients rarely develop further ischemic strokes after surgery\(^7\)\(^{-15}\).

References


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