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Opinion

Stroke Like Episodes in an Adolescent: What are the Metabolic Causes at this Age?

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Abstract

Adolescents are an age group that is not common to have any episode of stroke-like.

When this happen, there are three main metabolic diseases that we need to exclude.

The first disease is the classical homocystinuria. The main findings are: inferior deviation of the crystalline, cataracts, retina dislocation, high stature, marfanoid phenotype, mal occlusion of the teeth, psychomotor delay, scoliosis, kyphosis, aorta degeneration and stroke - like episodes caused by thromboembolism. The diagnosis is based on plasma and urine amino acids, that will show high levels of homocysteine e methionine.

The second disease is MELAS (mitochondrial encephalopathy, acidosis lactic and "stroke-like" episodes). They can present with stroke - like episodes and with these two or more symptoms: focal or generalized epileptic crisis, recurrent headache, vomiting and dementia. Others manifestations are variable: neurosensorial deafness, short stature, protein increase in CSF (50% of cases) and basal ganglia calcifications (30%).

The third disease is Fabry. Hands pain can be the first sign and is caused or by acroparesthesia - hands and feet with burn sensation daily or not, or by Fabry crisis - burn hands and feet in the beginning and after spread to the body that can take minutes or days. Other findings include: anhidrosis or hypohidrosis, frequent fever, intolerance to hot weather, epigastric pain, diarrhea after meal, renal failure cause by excessive proteinuria, cardiomegaly, arrhythmias, heart failure, vertigo, headache, depression and stroke - like in adolescents.

Keywords: *Stroke-Like; Adolescents; Homocystinuria; Melas; Fabry*

Background

Adolescents are an age group that is not common to have any episode of stroke-like.

When this happen, there are three main metabolic diseases that we need to exclude.

The clinical history of the patient plus the physical findings are important, because will give us very useful information about a possible cause of the stroke.

The first disease is the classical homocystinuria, an autosomal recessive disorder. These patients born with normal phenotype and until 3 years of age, they are asymptomatic. The main findings are: Inferior deviation of the crystalline, cataracts, retina dislocation, high

stature, marfanoid phenotype, mal occlusion of the teeth, psychomotor delay, scoliosis, kyphosis, aorta degeneration and stroke - like episodes at 15y cause by thromboembolism. The diagnosis is based on Plasma and urine amino acids, that will show high levels of homocysteine e methionine. The confirmation of the diagnosis is with the skin biopsy that will show cystathionine beta synthase deficiency or by the molecular study of CBS gene. For the treatment, we use low methionine diet, vitamin B6 - 200 mg, BID, oral folic acid - 10 mg, OID and oral acetylsalicylic acid – 5 mg/kg/day, oral.

The second disease is MELAS (mitochondrial encephalopathy, acidosis lactic and "stroke-like" episodes). These patients can have recurrent headache, vomiting and episodes of focal epilepsy. They common show past history of neurosensory deafness. They can present with these two or more symptoms: focal or generalized epileptic crisis, recurrent headache, vomiting and dementia. Others symptoms are variable: neurosensorial deafness, short stature, protein increase in CSF (50% of cases) and basal ganglia calcifications (30%). We can use antiepileptic's drugs, but never valproate, because can increase the level.

The third disease, is Fabry, a x-linked disorder. There is a defect in the alfa-galactosidase or alfa-GAL enzyme. This deficiency cause storage of GL-3 in the body. The patients can have symptoms in the first year of life, with anhidrosis, xerostomy and hands pain that can be confuse with juvenile rheumatoid arthritis. The macules in the body will confirm angiokeratoma after skin biopsy of the lesions. Abdomen CT scan revealed hepatic and left renal cyst. The hands pain is cause or by acroparesthesia - hands and feet with burn sensation daily or not, or by Fabry crisis - burn hands and feet in the beginning and after spread to the body that can take minutes or days. Other findings include: hipohidrosis, frequent fever, intolerance to hot weather, epigastric pain, diarrhea after meal, renal failure cause by excessive proteinuria, cardiomegaly, arrhythmias, heart failure, vertigo, headache, stroke in adolescents and depression. The diagnosis is with the determination enzyme activity of alfa-GAL and gene study. The treatment is with agalsidase beta: 1 mg/Kg, 2/2 weeks. The initial perfusion is 0,25 mg/min (15 mg/h) and increase gradually.

Conclusion

When we have an adolescent with stroke - like episode, we need to think that there are three possibilities to cause this: classical homocystinuria, MELAS and Fabry disease.

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