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CASE REPORT

CHAGASIC MEGAESOPHAGUS AND MEGACOLON DIAGNOSED IN CHILDHOOD AND PROBABLY CAUSED BY VERTICAL TRANSMISSION

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SUMMARY

Reports on children presenting symptoms compatible with the chronic phase of Chagas disease are sporadic. We report a case of a 7-year-old boy who had megaesophagus and megacolon, both of them a consequence of the trypanosomiasis. The etiology was established by means of laboratory and histological features. Based on epidemiological data, the authors concluded that vertical transmission was the most probable route of acquisition. This diagnosis should be considered in children presenting similar complaints, even those living away from endemic areas.

KEYWORDS: Chagas disease; Congenital Chagas disease; Megaesophagus; Megacolon.

INTRODUCTION

Chagas disease is caused by the parasitic protozoan, *Trypanosoma cruzi* (*T. cruzi*). It is transmitted from person to person via several hematophagous triatomid species. Other routes of infection are blood transfusion, organ transplantation and vertical transmission (congenital or rarely through breast-feeding).

For years Chagas disease has been considered an endemic health problem in Latin America rural areas. During the last decades, however, rural-urban migratory fluxes have moved thousands of *T. cruzi*-infected people to urban areas where transmission through the vector does not occur. The number of infected people living in metropolitan areas of South-East of Brazil is about half a million[17].

Chagas disease develops in three stages: acute, undeterminate and chronic. The clinical presentation varies in each phase. The acute phase is generally asymptomatic while a few cases present central nervous system or acute myocarditis manifestations. The undeterminate phase is an asymptomatic period between the acute phase and the beginning of the chronic stage. This phase may last from 10 up to 40 years. The chronic phase is marked by cardiac disorders and by digestive complaints related to megaesophagus and megacolon[15].

The prevalence of *T. cruzi* infection among fertile women ranges from 2% to 51% in Latin American urban areas and from 23% to 81% in rural regions[11]. Among chronic chagasic mothers, the frequency of fetal infection is around 1.6% of all pregnancies[13].

The infected newborns present a clinical spectrum that ranges from sepsis to the presence of a few symptoms[3,20]. The most common form of presentation includes jaundice, hepatosplenomegaly, anemia, petechiae, and convulsions[3]. Acute myocarditis, meningoencephalitis and manifestations resulting from megaesophagus or megacolon have been reported[3,5]. Prospective studies have pointed out that 25% to 67% of infected newborns can be asymptomatic, probably having acquired the infection in the last months of pregnancy[11]. In such cases it can be difficult to detect the infection only on a clinical basis.

Most of the reports on congenital Chagas disease describe the presence of the digestive symptoms related to megaesophagus and megacolon occurring at birth[1,2,6,19]. Reports on children presenting symptoms of congenital infection apart from the acute phase, and having been asymptomatic at birth, are rare. Two examples can be mentioned: a chagasic megaesophagus diagnosed in a suckling infant living in a non-endemic area and a preschool child presenting megacolon[4,9]. In these cases the precocious symptoms were considered to be an indication of congenital acquisition. Cohort surveys developed in endemic areas have diagnosed megaesophagus in children as young as 9 years old, but the possible route of infection was not investigated[10,16].

The present case report describes a seven-year-old patient who presented megaesophagus and megacolon as a consequence of Chagas disease. It is considered that vertical transmission was the most probable route of infection.

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CASE REPORT

A seven-year-old boy was admitted to the School Hospital at The State University of Campinas, Brazil. He was suffering from acute respiratory failure and denied dysphagia, vomiting or even chronic respiratory symptoms.

The child was born and used to live in Campinas, a one-million-people city in São Paulo state. The birth had been surgical, gestational age 32 weeks; weight 2,340 g, and Apgar score 2, 7 and 10. The child was breast-fed for two weeks after birth.

At 12 months of age, he was admitted to the hospital due to pneumonia and constipation, a fecal mass (fecal impaction) was removed by enema and a barium radioscopic esophagoscopy was taken. The esophagoscopy showed neither esophageal abnormalities nor gastroesophageal reflux. At 14 months, the patient had lymphocytic meningitis and received a blood transfusion. The child has been treated with lactulose and mineral oil since the age of 2, to relieve chronic constipation and at the time of admission (age 7) he was still using these medicines. The boy had also suffered recurrent pneumonias, with the first episode occurring at the age of 5.

Physical examination: The general condition was poor, 64 breaths/minute, 108 beats/minute, BP: 100 x 60 mmHg, temperature 38.6 ºC. Signs of slight dehydration and undernutrition were present. Weight: 21.9 kg, height: 120 cm (percentiles: weight/age 10, height/age 7, weight/height 44). There were no signs of retinopathy on fundoscopy. Thorax: intercostals retraction and abundant sounds with crackling snores at the pulmonary bases. Cardiac sounds were normal. Abdomen was globe-shaped, flaccid, and painless. There was a very large fecal mass between the xiphoid process and the lower abdomen. The solid abdominal organs were not palpable.

Laboratory findings: Thorax radiograph: a granular pattern with increased lung density and prominent bronchial air shadows, was observed, suggesting lipoid pneumonia. Hemoglobin: 9.8 g%, WBC: 18,000/mm^3. Mantoux test: negative. Sweat electrolyte determination was normal. Barium radioscopic esophagoscopy: slight enlargement of the esophagus, with a delayed emptying of the barium column and no evidence of gastroesophageal reflux. Esophageal manometry: aperistalsis upon wet deglutition and a partially relaxing lower esophageal sphincter. Esophagogastroscopy: no abnormalities. The barium enema showed markedly enlarged sigmoid and rectum. Anorectal manometry identified the inhibitory reflex. Cardiac evaluation: electrocardiogram, Holter test and ultrasound did not reveal abnormalities. Lung biopsy: bronchiolitis and pneumonitis, granulomatous foreign body reaction (compatible with chronic aspiration). *T. cruzi*-antibodies were positive in patient's serum as determined by the complement fixation test: ¼ (reference value > ½) and by indirect immunofluorescence: 1/160 (reference value > 1/20). The search for circulating trypomastigotes, performed by xenodiagnosis, was positive. Patient's epidemiological antecedents for Chagas disease were absent. He had never been in an endemic area. The blood transfusion he had at 14 months of age was exhaustively investigated at the Blood Center of the Hospital to rule out infection through blood transfusion. The blood had come from only one donor who was traced and his serological tests, including those for Chagas disease, were repeated, resulting negative. Donor xenodiagnosis was done as well, resulting negative. For three subsequent years the donor was followed up and all tests remained negative.

The patient's mother was born in an endemic Chagas disease rural area and presented positive xenodiagnosis. Maternal serological tests for Chagas disease were positive (indirect immunofluorescence and complement fixation). She had normal cardiac and digestive clinical examination and laboratory tests. Therefore, the mother was diagnosed as having Chagas disease in the undeterminate form.

The patient and his mother were treated with benznidazole for 60 days. Their xenodiagnoses were negative after treatment.

A partial colectomy was necessary to treat the child's constipation. The anatomo-pathological examination revealed a markedly enlarged rectum-colon segment (7 cm diameter) with no haustration. The histological report for the intestine showed that, lympho-monocytic inflammatory cells had replaced nerve cell bodies in the myenteric ganglia.

DISCUSSION

At the present case, the absence of an epidemiological environment for Chagas disease was based on the following data: the child was born in a non endemic area where parasitic transmission has not occurred for decades, and he had never been in an endemic area. Transmission by blood transfusion was ruled out. It should be pointed out that the clinical evidence of megacolon had began at the age of 1, indicating that he had already had chronic constipation before the blood transfusion, which is a sign of previous megacolon.

Considering these facts, the authors believe that there are two possible sources of infection: intrauterine and through breast-feeding. Since transmission through breast-feeding outside the maternal acute phase is extremely rare[7], we believe that it is more likely to be a congenital infection. Therefore, we may conclude that this is certainly a case of congenital Chagas disease that was not diagnosed during the acute phase.

The present patient did not complain of dysphagia, the most common and prominent symptom of chagasic megaesophagus, but the esophageal manometry showed the typical abnormalities seen in chagasic megaesophagus: a partially relaxing lower sphincter and aperistalsis upon deglutition[14]. These findings and the endoscopic reports ruled out the most common childhood diseases associated with megaesophagus: idiopathic achalasia, esophageal strictures, vascular rings and severe esophagitis[18].

The prevalence of colonic involvement among chagasic patients is not known. Dysphagia is more frequently reported than constipation. Most of the megacolon patients are asymptomatic or report minor symptoms. According to epidemiological studies conducted in regions where transmission does not occur by hematophagous vector bites, megacolon seems to occur in older age groups[12].

In the reviewed medical literature, only two well-confirmed reports of megacolon associated with congenital chagasic infection were found, one occurring in the acute phase and another at the beginning of the undeterminate phase[1,2].

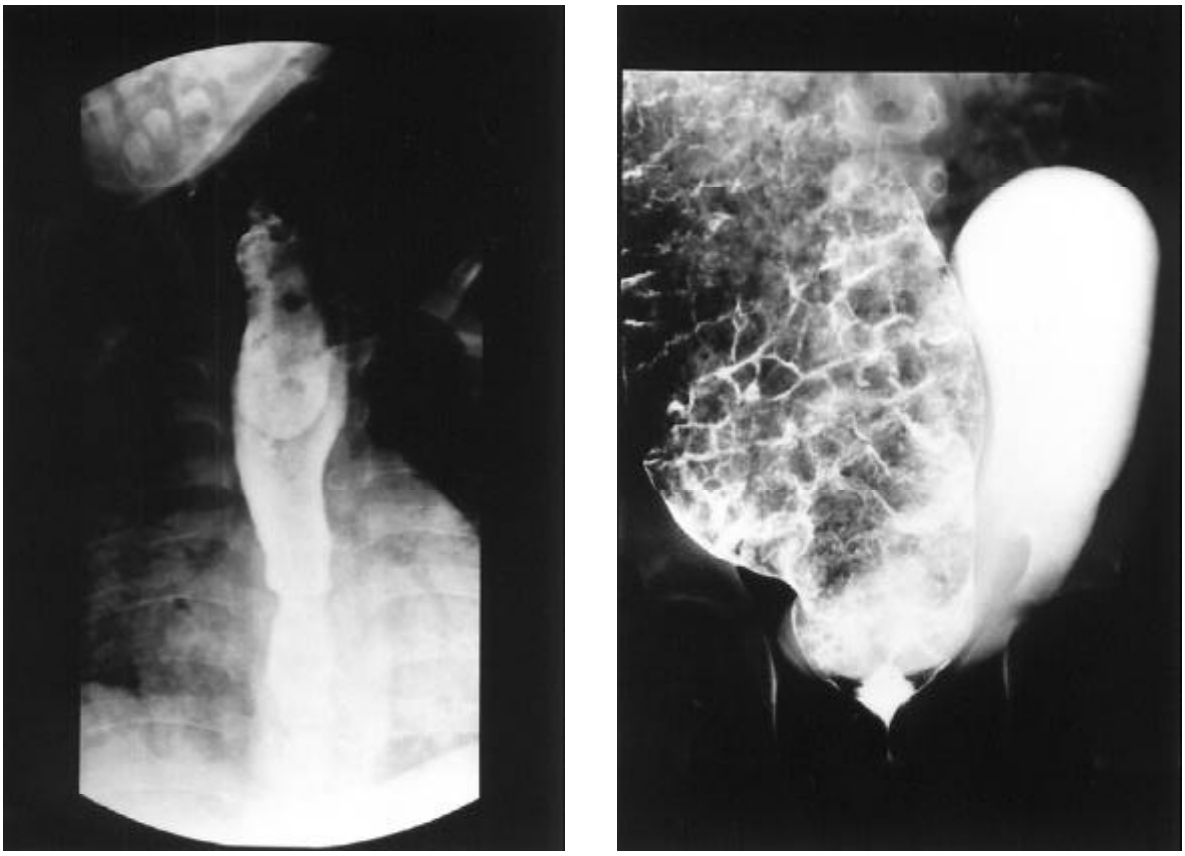

Fig. 1 - (A) Barium contrasted radiography from the esophagus showing a slightly enlarged organ, (B) barium contrasted radiography from the large intestine presenting a very large rectum-sigmoid segment filled with fecal mass.

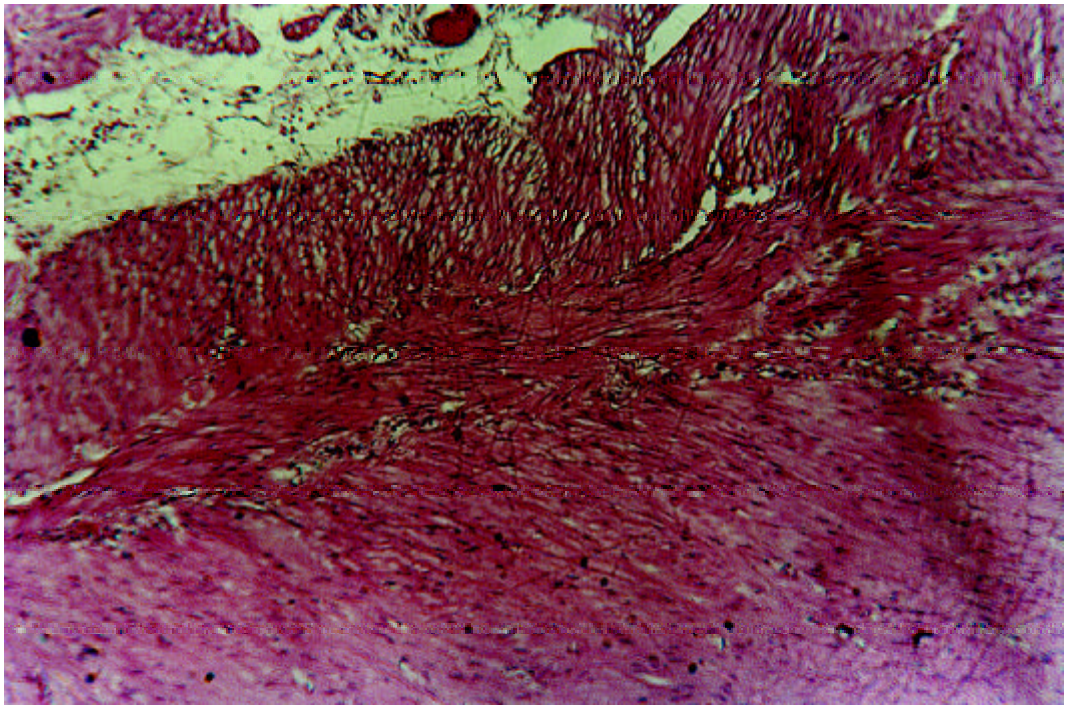

Fig. 2 - Histological section from large intestine wall. The lympho-monocytic cells have replaced ganglia neurons of myenteric plexus.

Megaesophagus and megacolon are seen separately during the acute phase in most congenital acquisition patients[1,2,5,6,19]. It is noteworthy that the present patient presented simultaneously megaesophagus and megacolon at the beginning of the undeterminate phase, an association that has not been reported previously. This clinical picture could demonstrate the difference in pathogenesis between the cardiac and digestive involvement in the acute phase of Chagas disease. In the cardiac disease pathogenesis, besides the direct effect of the parasitism on myocardial cells, an autoimmune phenomenon is considered to be involved, whereas the digestive disease would be a consequence of a destructive inflammatory process in the myenteric plexuses[8]. Therefore, in the congenital infection, the effect of the parasite on the digestive system of the immunologically immature host would result in lesions whose manifestations could occur in the neonatal period during the acute phase or just beyond it, at the beginning of the undeterminate phase[8].

Congenital Chagas disease should be considered more frequently as a differential diagnosis in children living in metropolitan areas and having gastrointestinal dysmotility diseases including severe constipation, dysphagia and even recurrent aspirative pneumonia.

RESUMO

Megacolon e megaesôfago chagásicos diagnosticados na infância e com provável transmissão vertical

A doença de Chagas congênita é mais comumente identificada nos recém nascidos com sinais sugestivos de infecção intrauterina. Outras expressões clínicas são menos conhecidas e referências de manifestações compatíveis com a fase crônica da doença são raras na faixa etária pediátrica. Apresenta-se o caso de paciente em que, aos sete anos de idade, foram diagnosticados megaesôfago e megacolon secundários a tripanosomíase. A etiologia foi confirmada por meio de xenodiagnóstico, sorologias e achados anátomo-patológicos. Com base na epidemiologia, foi possível descartar outras vias de infecção que não a transmissão vertical. O diagnóstico deve figurar entre crianças com constipação intestinal e/ou sintomas de dismotilidade esofágica, mesmo fora das zonas endêmicas para a tripanosomíase.

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