

CASE REPORT

ABDOMINAL ANGIOSTRONGYLIASIS: REPORT OF TWO CASES WITH DIFFERENT CLINICAL PRESENTATIONS

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SUMMARY

Abdominal angiostrongyliasis is a sporadic infectious disease caused by the nematode *Angiostrongylus costaricensis*. It usually presents as acute abdomen, secondary to mesenteric ischemia, and pronounced eosinophilia. In some cases its course is insidious and transient, and the diagnosis is suspicious. The disease is confirmed by the detection of *A. costaricensis* elements in surgical specimen. The treatment is supportive, with avoidance of antihelminthic administration due to a possible erratic migration followed by worsening of the disease. We report two cases, both with intense eosinophilia and serum IgG-ELISA positive to *A. costaricensis*. The first case presented ileal perforation and was surgically treated. The second one showed hepatic nodules at ultrasound and was only symptomatically treated, evolving to an apparent protracted resolution. These two cases exemplify different clinical forms of the disease, one of them with liver involvement.

KEYWORDS: *Angiostrongylus costaricensis*; Abdominal angiostrongyliasis; Eosinophilia; Acute abdomen; Liver.

INTRODUCTION

Abdominal angiostrongyliasis (AA) is caused by the nematode *Angiostrongylus costaricensis*, which may affect children and adults with similar distribution between males and females¹⁰. It is a sporadic disease, occurring mainly in the south of Brazil and other Latin America countries, particularly in Costa Rica². However, the disease seems to be under diagnosed by the lack of suspicion when a patient presents with abdominal pain and eosinophilia⁴.

A wild rodent is taken as definitive host of *A. costaricensis*, whereas intermediated hosts are represented by mollusks of Veronicellidae family^{2,6}. The parasite spreads to humans by means of consumption of vegetables containing *A. costaricensis* larvae. According to serological studies, the number of subclinical cases may be even higher than cases with symptomatic disease².

When symptomatic, AA manifests as intestinal lesions characterized either by infarction, pseudotumor or acute appendicitis, associated with marked eosinophilia. Most cases present as an acute disease, requiring urgent laparotomy¹⁵. The diagnosis of AA is confirmed by the identification of eggs, larva or adult worms of *A. costaricensis* in surgical specimen. In the absence of parasitic elements, the disease can be diagnosed based on three fundamental histopathological findings: a) a massive infiltration of eosinophils in all layers of the intestinal wall; b) a granulomatous reaction; and c) eosinophil vasculitis affecting arteries,

veins, lymphatics and capillaries³. A presumptive diagnosis may be indicated when serology reveals serum globulins against the parasite^{1,2,5,14}. The serological test uses crude antigen of female worms, with sensitivity of 76% and specificity of 91%⁵.

This study reports two cases of abdominal angiostrongyliasis which illustrate different clinical and histological spectra of the disease.

Case 1. Male, 32 years old, farmer, coming from the municipality of Sarandi, in the southernmost State of Rio Grande do Sul, Brazil. The patient was assisted in the emergency room due to acute and diffuse abdominal pain, followed by fever and signs of peritoneal irritation. There was history of intermittent and weak abdominal pain in the lower right quadrant during the last year. The patient was in good nutritional state, febrile (38.5 °C), with distended abdomen and painful wall decompression. Blood analysis revealed high leukocyte count (29,000/μL) with eosinophilia (12,470/μL). Abdominal X-ray showed signs of static ileum. The patient underwent exploratory laparotomy, which revealed free enteric liquid in the peritoneal cavity, as well as a phlegmon in the ileocecal region associated with ileal perforation. Surgical approach consisted of ileocelectomy with enterocolonic anastomosis. Patient discharge occurred seven days latter.

The macroscopical analysis of the surgical specimen showed fibrinous purulent exudate covering the ileocecal segment and ileal perforation. There were coalescent and flat ulcers in the mucosal surface.

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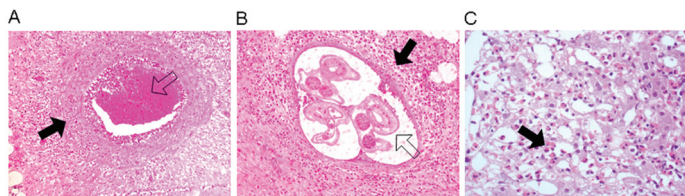


Fig. 1 - Microscopic aspects of surgical specimen from case 1 (A and B), and hepatic biopsy from case 2 (C) (HE stain). **A** (x200), Intestinal artery containing thrombus (open arrow) and showing eosinophilic arteritis (solid arrow). **B** (x200), *A. costaricensis* in the lumen of intestinal artery (open arrow), with pronounced perivascular infiltrate of eosinophils (solid arrow). **C** (400x), Hepatic parenchyma showing marked eosinophilic infiltrate (solid arrow), particularly in portal spaces.

The microscopic analysis revealed trans-mural necrosis associated with intense arterial thrombosis (Fig. 1A), as well as an exuberant perivascular eosinophilic infiltrate. Six adult worms of *A. costaricensis* were found in the lumen of intestinal arteries (Fig. 1B). The serological study (*ELISA*) was positive for G immunoglobulin against the parasite (title of 0.487). Patient evaluation eight weeks after surgery revealed uneventful clinical recovery with normal leukocyte count (6,800/ μ L) and almost resolution of eosinophilia (544/ μ L).

Case 2. Female, 34 years old, nutritionist, coming from the municipality of Água Santa, in the southernmost State of Rio Grande do Sul, Brazil. The patient was first assisted in a doctor's office complaining of vague pain in the right upper abdominal quadrant, associated with general muscle pain and malaise in the last four weeks. Physical examination revealed painful hepatomegaly. A peripheral blood analysis indicated the presence of leukocytosis (30,200/ μ L) with pronounced eosinophilia (24764/ μ L). The patient was hospitalized for 20 days, during which showed protracted clinical evolution, with periods of muscle and abdominal pain responsive to per-oral analgesia. Serum analysis showed a slight increase in serum aminotransferases and lactate dehydrogenase. Abdominal ultrasound showed hepatic nodules, intra-hepatic bile ducts ectasia, and enlargement of spleen and lymph nodes in hepatic hilum and retroperitoneum. An exhaustive search for *Strongyloides stercoralis* (stool analysis and indirect immunofluorescence assay (IIA) for G immunoglobulin in serum) and *Toxocara* (IIA for M

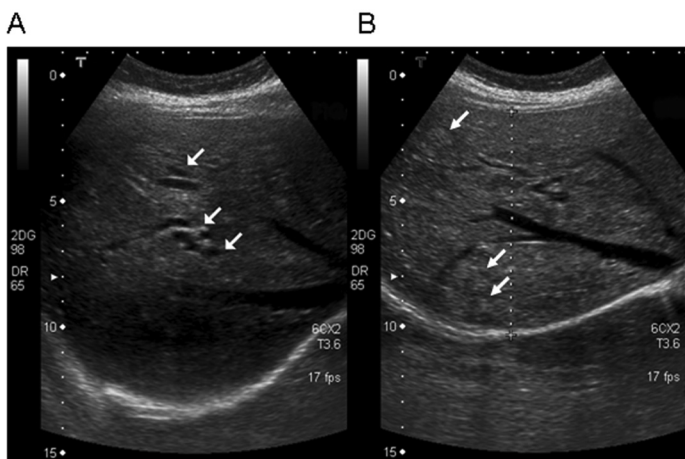


Fig. 2 - Abdominal ultrasound showing ectasia of intra-hepatic bile ducts (A) and hepatic parenchyma with heterogeneous echogenicity resembling nodules (B, arrows).

immunoglobulin in serum) was negative. Patient's husband also presented similar clinical (malaise and vague abdominal pain) and laboratorial findings (eosinophilia of 5,520/ μ L and *ELISA*-IgG positive against AA, with title of 0.235), but with lower intensity. The patient underwent ultrasound-oriented biopsy of hepatic nodules, which showed marked eosinophilia infiltrating liver parenchyma and portal spaces (Fig. 1C), with formation of sparse granuloma containing necrosis and eosinophils, without parasitic elements. The serological analysis (*ELISA*) was positive for *A. costaricensis* specific immunoglobulin (title of 0.209). Patient was oriented to keep symptomatic treatment and repeat leukocyte count. Symptoms disappeared four months after hospitalization, with gradual normalization of eosinophils count (2,403/ μ L and 45/ μ L six and 12 months latter, respectively). Abdominal ultrasound 12 months latter still showed small hepatic nodules and bile duct ectasia (Fig. 2), however without enlargement of organs or lymph nodes.

DISCUSSION

This study highlights the diverse spectrum of clinical and pathological presentations of abdominal angiostrongyliasis, exemplified by two representative case reports. Case 1 illustrates the acute form of the disease, in which an acute inflammatory abdomen secondary to ileocecal damage required surgical intervention. In contrast, case 2 represents an insidious form of AA, with hepatic involvement and protracted resolution. However, the case 1 was confirmed by the detection of intraarterial adult worms of *A. costaricensis*, while the case 2 was only presumptive, based on serum analysis and eosinophilia in liver portal spaces.

The acute form seems to predominate in the clinical practice. Intestinal lesions, as illustrated in case 1, are associated with the presence of *A. costaricensis* in the mesenteric arteries, resulting in intestinal ischemia, particularly in the ileocecal segment. The final events of this injury may vary between intestinal infarct, intestinal perforation, formation of pseudotumor or acute appendicitis^{2,3}. Otherwise, liver involvement is unusual, with publications restricted to case reports^{7,11,16}. The finding of tender hepatomegaly combined with slight increase in aminotransferases and marked eosinophilia should raise suspicion for AA. MOREIRA *et al.*¹¹ considered the *A. costaricensis* adult worm migration to intrahepatic portal veins as a visceral larva-migrans-like syndrome. However, MOTA & LENZI^{12,13}, using two different animal models (Swiss Webster and *Sigmodon hispidus*) showed that the *A. costaricensis* presents, during its life cycle, an alternative migration to hepatic veins as a normal event of venous portal pathway, where the nematode matures and lays fertile eggs inside the liver. Then, according to MOTA & LENZI^{12,13}, *A. costaricensis* adult worms can reach the liver by branches of hepatic artery and portal vein. Although identification of *A. costaricensis* in tissue specimen is confirmatory of AA, in many cases this is quite difficult due to the limited number of parasites causing the disease¹⁵. In cases where *A. costaricensis* is not found, the presence of specific antibodies showed at *ELISA* test gives a presumptive diagnosis of AA, provided that other diseases such as systemic strongyloidiasis and visceral larva migrans are properly excluded.

The current treatment of AA is supportive and expectant^{2,15}. There is no proved efficacy for antihelminthic agents, with a theoretical risk of erratic migration of *A. costaricensis* after employment of these drugs⁹. Therefore, the frequent lack of knowledge about AA by the medical community may provide risk with the use of antihelminthic drugs in

patients presenting with abdominal pain and eosinophilia. MENTZ & GRAEFF-TEIXEIRA⁸ showed a good review about treatment of AA. The surgical approach is indicated for patients with acute abdomen, in order to solve ischemia related intestinal damage. General maneuvers, such as proper hydration, analgesia and nutrition are cornerstone. Since the diagnosis of AA is established, patient must be oriented regarding the unpredictable prognosis related with this disease^{2,14}.

RESUMO

Angiostrongilíase abdominal: relato de dois casos com diferentes apresentações clínicas

A angiostrongilíase abdominal é doença esporádica decorrente da infecção pelo nematódeo *Angiostrongylus costaricensis*. Costuma manifestar-se como abdome agudo secundário a isquemia mesentérica, além de marcada eosinofilia. Pode também apresentar-se de forma insidiosa e transitória, exigindo alta suspeita clínica para o diagnóstico. A doença é confirmada pela identificação de elementos do *A. costaricensis* em peças cirúrgicas. O tratamento é apenas de suporte, devendo-se evitar o uso de anti-helmínticos pela possibilidade de migração errática do verme com piora do quadro. Aqui foram apresentados dois casos, ambos com acentuada eosinofilia e ELISA-IgG sérico positivo para *A. costaricensis*. O primeiro caso cursou com perfuração ileal e foi tratado cirurgicamente. O segundo caso apresentou nódulos hepáticos ao ultrassom e foi tratado sintomaticamente, evoluindo para lenta resolução. Estes dois casos exemplificam diferentes formas de apresentação clínica da doença, uma delas com envolvimento hepático.

REFERENCES

1. GEIGER, S.M.; LAITANO, A.C.; SIEVERS-TOSTES, C. *et al.* - Detection of the acute phase of abdominal angiostrongyliasis with a parasite-specific IgG enzyme linked immunosorbent assay. *Mem. Inst. Oswaldo Cruz*, 96: 515-518, 2001.
2. GRAEFF-TEIXEIRA, C.; AGOSTINI, A.A. & RODRIGUEZ, R. - Angiostrongilíase abdominal. In: TAVARES, W. & MARINHO, L.A.C., ed. *Rotinas de diagnóstico e tratamento das doenças infecciosas e parasitárias*. São Paulo, Atheneu, 2007. p. 98-101.
3. GRAEFF-TEIXEIRA, C.; CAMILLO-COURA, L. & LENZI, H.L. - Histopathological criteria for the diagnosis of abdominal angiostrongyliasis. *Parasit. Res.*, 77: 606-611, 1991.
4. GRAEFF-TEIXEIRA, C.; CAMILLO-COURA, L. & LENZI, H.L. - Abdominal angiostrongyliasis: an under-diagnosed disease. *Mem. Inst. Oswaldo Cruz*, 82(suppl. 4): 353-354, 1987.
5. GRAEFF-TEIXEIRA, C.; GOULART, A.H.; BRUM, C.O. *et al.* - Longitudinal clinical and serological survey of abdominal angiostrongyliasis in Guapore, southern Brazil, from 1995 to 1999. *Rev. Soc. bras. Med. trop.*, 38: 310-315, 2005.
6. GRAEFF-TEIXEIRA, C.; THOME, J.W.; PINTO, S.C.; CAMILLO-COURA, L. & LENZI, H.L. - *Phyllocaulis variegatus*: an intermediate host of *Angiostrongylus costaricensis* in south Brazil. *Mem. Inst. Oswaldo Cruz*, 84: 65-68, 1989.
7. MENTZ, J.P.; DALVESCO, J.A.; AGOSTINI, A.A. & BONADEO, N.M. - Manifestações de comprometimento hepático na angiostrongilíase abdominal e diagnóstico pelo encontro dos ovos do parasita. *Rev. AMRIGS*, 37: 289-290, 1993.
8. MENTZ, M.B. & GRAEFF-TEIXEIRA, C. - Drug trials for treatment of human angiostrongyliasis. *Rev. Inst. Med. trop. S. Paulo*, 45: 179-184, 2003.
9. MORERA, P. & BONTEMPO, I. - Acción de algunos antihelmínticos sobre *Angiostrongylus costaricensis*. *Rev. méd. Hosp. nac. Niños (Costa Rica)*, 20: 165-174, 1985.
10. MORERA, P. & CESPEDES, R. - *Angiostrongylus costaricensis* n. sp. (Nematoda: Metastrongyloidea), a new lungworm occurring in man in Costa Rica. *Rev. Biol. trop.*, 18: 173-185, 1970.
11. MORERA, P.; PEREZ, F.; MORA, F. & CASTRO, L. - Visceral larva migrans-like syndrome caused by *Angiostrongylus costaricensis*. *Amer. J. trop. Med. Hyg.*, 31: 67-70, 1982.
12. MOTA, E.M. & LENZI, H.L. - *Angiostrongylus costaricensis*: complete redescription of the migratory pathways based on experimental *Sigmodon hispidus* infection. *Mem. Inst. Oswaldo Cruz*, 100: 407-420, 2005.
13. MOTA, E.M. & LENZI, H.L. - *Angiostrongylus costaricensis* life cycle: a new proposal. *Mem. Inst. Oswaldo Cruz*, 90: 707-709, 1995.
14. PALOMINOS, P.E.; GASNIER, R.; RODRIGUEZ, R.; AGOSTINI, A.A. & GRAEFF-TEIXEIRA, C. - Individual serological follow-up of patients with suspected or confirmed abdominal angiostrongyliasis. *Mem. Inst. Oswaldo Cruz*, 103: 93-97, 2008.
15. RODRIGUEZ, R. - *Anatomia patológica da angiostrongilose abdominal*. Rio de Janeiro, 1997. (Dissertação de Mestrado - Universidade Federal Fluminense).
16. VÁZQUEZ, J.J.; SOLA, J.J. & BOILS, P.L. - Hepatic lesions induced by *Angiostrongylus costaricensis*. *Histopathology*, 25: 489-491, 1994.

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