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JEJUNAL PERFORATION CAUSED BY ABDOMINAL ANGIOSTRONGYLIASIS

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SUMMARY

The authors describe a case of abdominal angiostrongyliasis in an adult patient presenting acute abdominal pain caused by jejunal perforation. The case was unusual, as this affliction habitually involves the terminal ileum, appendix, cecum or ascending colon. The disease is caused by the nematode *Angiostrongylus costaricensis*, whose definitive hosts are forest rodents while snails and slugs are its intermediate hosts. Infection in humans is accidental and occurs via the ingestion of snail or slug mucoid secretions found on vegetables, or by direct contact with the mucus. Abdominal angiostrongyliasis is clinically characterized by prolonged fever, anorexia, abdominal pain in the right-lower quadrant, and peripheral blood eosinophilia. Although usually of a benign nature, its course may evolve to more complicated forms such as intestinal obstruction or perforation likely to require a surgical approach. Currently, no efficient medication for the treatment of abdominal angiostrongyliasis is known to be available. In this study, the authors provide a review on the subject, considering its etiopathogeny, clinical picture, diagnosis and treatment.

KEYWORDS: *Angiostrongylus*; Intestinal diseases; Parasitic infection; Intestinal perforation.

INTRODUCTION

In 1967, MORERA[19] and CÉSPEDES *et al*.[8] reported their discovery in Costa Rica of a human parasitism named abdominal angiostrongyliasis, produced by a filiform nematode parasite that was later named *Angiostrongylus costaricensis.* It acts as a parasite in the terminal branches of the superior mesenteric artery, particularly the ileocecal area, thus leading to an inflammatory process and thrombosis[4].

The present study describes a case of abdominal angiostrongyliasis complicated by jejunal perforation in an adult patient. The authors emphasize the need to describe both the occurrence and diagnosis in order to carefully consider this zoonosis in view of the increasing number of infected individuals, especially in South America. Since no specific treatment seems to be currently available, the use of anti-helminth drugs is likely to mask or even worsen the clinical condition. The disease may evolve to intestinal obstruction or perforation, which are known to be potentially fatal complications requiring surgical procedures.

CASE REPORT

A 62- year-old white male patient, born in Portugal and living in Santo André, State of São Paulo, Brazil, for the past 60 years, was admitted to the emergency unit presenting diffuse, spasmodic and mild abdominal pain, associated with fever, myalgia and asthenia, which he had been suffering for the previous 20 days. The patient reported an increase in pain intensity in the flank and right iliac fossa during the last three days, in addition to abdominal distension. Physical examination showed the presence of fever (37.7 ºC) and pain produced by palpating the right flank, with painful abrupt decompression present in the whole region. The hemogram presented a leukocyte count of 15,000/mm³, with 4% band cells, 60% neutrophils and 6% eosinophils. Based on the diagnostic assumption of acute abdominal inflammation, the patient was submitted to an exploratory laparotomy, which demonstrated the existence of a great amount of fibrin-purulent secretion and enteric fluid that was free in the peritoneal cavity. At a location 60 cm distal from the Treitz angle, a jejunal perforation measuring 1.0 cm in diameter was found within a loop covered with fibrin. The segment involved was submitted to an enterectomy and a primary enteroenteric anastomosis was then performed. No other intestinal segments presented any involvement. Microscope examination of the resected specimens showed intense eosinophilic infiltration throughout the intestinal wall, perforating enteritis, fibrin-leukocytic acute serositis and the presence of adult worms of *Angiostrongylus costaricensis* in the small caliber arteries (Fig.1).

On the fifth postoperative day, the patient developed an abdominal wall abscess, which was afterwards drained. On the 10th postoperative day, dehiscence of an aponeurotic suture could be observed and a new abdominal wall suture was performed. The patient was discharged on the 15th postoperative day without any additional complication. After a two-year follow-up period, the patient did not present any further signs or symptoms related to the disease.

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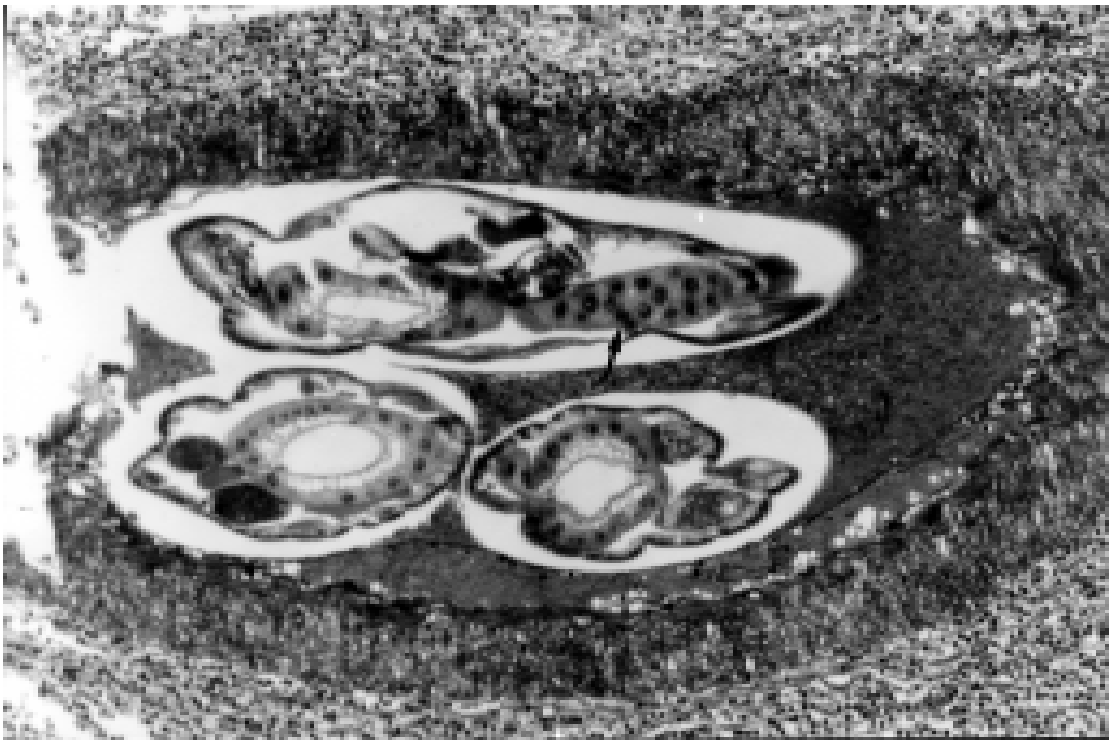

Fig. 1 - Parasitic artery branch with *Angiostrongylus costaricensis*' lumen transversal section (H&E, x 200).

DISCUSSION

Several species of the nematode *Angiostrongylus* are known to infect animals, particularly rodents and carnivorous species. *Angiostrongylus costaricensis* is the agent responsible for occurrences of abdominal angiostrongyliasis, and was initially observed in Costa Rica. It is thought likely that this disease mostly affects children[23], and it is becoming disseminated to other countries of Central and South America[3], including Brazil[1,2,5,6,10,14,17,21,25,26].

Angiostrongylus costaricensis is a helminthic filiform nematode measuring 2-3 cm in length. Its well-adapted and definitive hosts are forest rodents (*Sigmodon hispidus, Rattus rattus, Oryzomys nigripes* and *Oryzomys ratticeps)*[12] and its intermediate hosts are land mollusks, especially from the *Veronicellidae* (*Vaginulus Sarasinula plebeius*) family, commonly known as slugs[20]. Adult forms of nematodes inhabit the rodents' mesenteric arteries in the ileocecal region, which is their predominant egg-laying site. First-stage larvae emerging from embryo-filled eggs are eliminated in the rodents' feces, which will infect the slugs when ingested. The larvae reach the fibromuscular tissue of the slug, where two molts take place and they become third-stage larvae. These larvae, which are the infecting agents for vertebrate species, are eliminated through mucus secretions that are produced when the slugs move or are touched[9]. The contamination of the slugs' secretions is due to the location of the larvae, close to the excretory ducts of the slugs' mucus-producing glands[20]. Food contamination, mostly caused by inadequately cleansing vegetables, washing hands and disinfecting various instruments that are contaminated with the slugs' secretions, may lead to accidental human infection. As they are usually unseen by children, the mollusks are likely to be ingested together with the contaminated food.

In humans, the eggs fail to hatch and they degenerate, which results in formation of granulomas with intense eosinophilic infiltrate, suggesting that the infection is not transmissible[7,13]. However, within human arteries, the adult worms damage the vascular endothelium causing thrombosis and zonal necrosis when irrigated by the injured vessel. In this present case, the patient was affected by both types of event. At the same time, eggs, embryos and larvae, as well as secretory and excretory products from the parasite, may reach the intestinal wall arterioles, resulting in an inflammatory process[18].

The lesions may be classified into two macroscopic patterns: a pseudoneoplastic pattern showing predominance of intestinal wall thickening, and an ischemic-congestive pattern with congestive and necrotic segmental areas[11]. The pseudoneoplastic lesions are dependent on there being a more prolonged condition or a lower number of worms that remain in the mesentery and release antigens, which are responsible for the inflammatory reactions in the vascular walls or in different intestinal layers. The congestive ischemic lesions show a more acute course and seem to depend on larger parasitic loads or migratory stimulus from the worms as they seek out more distal segments of the arterial network[11]. In Brazil, the *Angiostrongylus costaricensis* nematode is the only helminth that is substantially localized at intra-arterial sites[22].

In decreasing order of frequency, lesions may also be found in the cecum, appendix, ascending colon and terminal ileum[16]. In our case, the intestinal segment involved was shown to be exclusively the proximal

jejunum. Histologically, the lesions are characterized by the abundant exudation of inflammatory fluid, with intense eosinophilic infiltrate in which the parasite eggs are seen, in addition to a granulomatous reaction to the foreign body[8,16].

Embryos at several developmental stages are observed to be hatching from the eggs, which are thin-walled and greater than 90µm in diameter[19]. These eggs show two peculiarities: they are not located within the small intestine but in the capillaries and may present themselves as embryo-filled whilst bearing a completely formed larva. Worms and eggs may be observed in the lymphatic nodes, while eosinophilic infiltrates, granulomas and vasculitis are also present[11].

Within medium-caliber arteries, the presence of parasitic fragments may produce thrombi likely to be responsible for the ischemic necrotic areas[8]. The *Angiostrongylus* nematode parasite could be identified in this study from intra-arterial transversal sections through cylindrical worms that showed the digestive tract and two reproductive tubes in an epithelium-impoverished cavity limited by a thinly striated cuticle[11] (Fig. 1).

In humans the parasite is located in the ileocecal-colic branches of the superior mesenteric artery causing eosinophilic vasculitis which affects arteries, veins and lymph nodes[11]. Both eosinophilic arteritis and the intra-arterial granulomatous reaction are considered peculiar to parasitosis[1]. The preferential distribution of the parasite within the mesoappendix vessels produces a clinical picture of terminal ileitis, which may mimic an acute appendicitis.

It is suggested that the interval between the onset of infection and its clinical manifestations could be similar to those observed in experiments on rats, namely, approximately 24 days[20].

Abdominal pain, fever, anorexia, nausea, vomiting and changes in intestinal habits usually make up the clinical picture[11]. In chronic cases, the patients may present low-grade fever and mild and occasional abdominal pain for several weeks[18]. When evolving to chronicity, the intestinal inflammatory reaction becomes a palpable mass in the right iliac fossa[24]. The clinical picture can be intermittent: the patient may present an alternation of remission periods followed by short symptomatic intervals; at this point, a sub-occlusive condition may occur.

Another possible complication is the intestinal perforation caused by peritonitis and sepsis. The patients should be carefully followed up so that prompt treatment may be provided in relation to the occlusion or the intestinal perforation. These are generally considered to be acute complications, responsible for the mortality rate among patients with abdominal angiostrongyliasis[11]. Although some of these contaminated individuals may present symptoms for months or years, most patients are able to recover from the disease within weeks[15]. The presence of leukocytosis and eosinophilia provide significant data for the elaboration of a clinical diagnosis. In most cases, the leukocyte count ranges from 15,000 to 40,000/mm³ and eosinophilia from 20 to 80%[18].

The clinical diagnosis of abdominal angiostrongyliasis is obtained by means of combining the clinical picture with any history of traveling to endemic areas and a serologic validation. Although the ELISA serologic test is sufficiently sensitive, it fails in specificity as a consequence of antibody cross-reactions to other nematode parasites[13]. As no evidence of the elimination of eggs and worms is found in human feces, the definitive diagnosis is dependent upon histologic sections for parasite identification. Three major signs are then evaluated: eosinophilia, perivascular granulomatous reaction and eosinophilic vasculitis[11]. The differential diagnosis should be made on the basis of acute appendicitis, lymphoma, intestinal tuberculosis and Crohn's disease[9,11].

The pharmacological treatment of intestinal angiostrongyliasis remains controversial. Some authors are against prescribing the use of anti-helminth drugs, such as thiabendazole or diethylcarbamazine, in view of the self-limiting process that is characteristic of the disease and the possibility that the worms' erratic migration could aggravate the lesion[11]. Most patients presenting an abdominal mass in the ileocecal region must undergo laparotomy to exclude any suspicion of neoplasm formation.

The increasing number of intestinal angiostrongyliasis cases among humans could be attributed to ecological modifications resulting from untimely and inadequate use of harmful and toxic agrochemicals. Such modifications may generate imbalances in the mollusk populations that form the intermediate hosts and, as a consequence of this, generate higher levels of risk for human infection[11]. Preventive measures include careful food inspection and cleaning, especially prior to vegetable consumption[24]. Land snails such as *Megalobulimus sp*, should be adequately boiled before ingestion and their production must be undertaken in closed breeding settings so as to avoid any contact with feces and rodents; slug fishing baits must also be avoided[11]. Measures directed against domestic rodents will probably not cause impact on human transmission of *Angiostrongylus costaricensis*[11].

RESUMO

Perfuração jejunal causada por angiostrongilíase abdominal

Os autores descrevem caso de angiostrongilíase abdominal em doente adulto que se manifestou como abdômen agudo devido à perfuração de alça jejunal, evento raro, uma vez que esta afecção geralmente envolve o íleo terminal, apêndice, ceco ou cólon ascendente. A doença é causada pelo nematódeo *Angiostrongylus costaricensis* cujos hospedeiros definitivos são roedores silvestres e os hospedeiros intermediários são caracóis e caramujos. A infecção em humanos é acidental e ocorre pela ingestão de secreção mucóide destes invertebrados presentes em vegetais ou por contato direto com o muco. A angiostrongilíase abdominal é clinicamente caracterizada pela presença de febre prolongada, anorexia, dor no quadrante inferior direito do abdômen e eosinofilia periférica. Embora a doença seja de natureza benigna, seu curso pode evoluir para formas complicadas como a obstrução ou perfuração intestinais que necessitam de tratamento cirúrgico. Atualmente, não há tratamento medicamentoso eficaz para a angiostrongilíase abdominal. Neste estudo, os autores realizam uma revisão desta afecção em relação à sua etiopatogenia, quadro clínico diagnóstico e tratamento.

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