



Burkitt's lymphoma presenting as double intussusception in a child with human immunodeficiency virus infection

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Resumo

O diagnóstico diferencial de abdómen agudo em crianças infectadas por VIH (Vírus da Imunodeficiência Humana) inclui um amplo espectro de etiologias. Apresentamos o caso de uma criança infectada por VIH que se apresentou com uma oclusão intestinal secundária a dupla invaginação, tendo como ponto de partida um linfoma de Burkitt. A invaginação intestinal não deve ser esquecida no diagnóstico diferencial da oclusão intestinal em crianças, e é mais comum em lactentes. A ocorrência de invaginação após o primeiro ano de vida deve levantar a suspeita de um processo patológico, especialmente se houver uma doença subjacente (tal como a infeção por VIH). A invaginação pode ser dupla, como neste caso, tendo como ponto de partida dois tumores síncronos.

Palavras-chave: Linfoma de Burkitt; infeção VIH; Invaginação intestinal

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Linfoma de Burkitt manifestando-se como dupla invaginação intestinal em criança infectada por vírus de imunodeficiência humana

Abstract

The differential diagnosis of an acute abdomen in HIV (Human Immunodeficiency Virus)-infected children includes a broad spectrum of etiologies. We present the case of an HIV-infected child presenting with bowel obstruction secondary to double intussusception, with a Burkitt's lymphoma as the lead point. Intestinal intussusception should not be overlooked in the differential diagnosis of bowel obstruction in children, and is more common in infants. The occurrence of intussusception beyond infancy should raise suspicion of a pathologic lead point, especially if there is underlying pathology (such as HIV infection). Intussusception may be double, as in this case, triggered by two synchronous tumors.

Key-Words: Burkitt's lymphoma; HIV-infection; Intussusception

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Background

The HIV (Human Immunodeficiency Virus)-infected children are susceptible to diseases common to the general population; however, their state of immunodeficiency places them at increased risk for many unusual disorders, predominately infectious and neoplastic. The acute abdomen in HIV-infected children can be a diagnostic challenge and the differential diagnosis includes infectious and neoplastic diseases and anti-retroviral side-effects. The diagnosis and intervention are often delayed because of the varied and unusual presentation and their inability to mount an adequate immune response.

Common opportunistic pathogens that cause acute abdomen in the setting of severe immunodeficiency include Cytomegalovirus and *Mycobacterium avium*.

The highly active antiretroviral therapy (HAART- Highly Active Antiretroviral Therapy) has been associated with a number of drug-induced side effects, including pancreatitis.

Among neoplastic diseases in HIV-patients, lymphomas account for ten-percent¹. Burkitt's lymphoma is a mature B-cell neoplasm that mostly involves the abdomen, the head or neck region. When primary disease occurs in the abdomen, diagnosis often occurs in the setting of acute complications². Intestinal obstruction may result, from either compression of the lumen or by an intussusception triggered by a tumor mass³.

Case report

We present the case of a four-year-old boy with congenitally acquired HIV infection and currently on antiretroviral therapy (HAART), who presented to the emergency department with a one-week history of a gradually worsening crampy periumbilical pain and two-day history of vomiting (lately biliary) and constipation. The remainder of the patient's history was unremarkable. The most recent CD4 count was 687/uL.

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Physical examination revealed a well-nourished and afebrile patient. He was prostrated. Vital signs were stable. The abdomen was distended and bowel sounds were diminished. There was a tender mass in the left upper and lower quadrants.

Initial abdominal plain film demonstrated scarcity of air in the colon, and two air-fluid levels, consistent with partial small bowel obstruction. Abdominal ultrasonography revealed intestinal intussusception with the leading edge of the intussusceptum at the level of the left colon. A classic “target” sign was observed on transverse section. In the centre there was an echogenic mass that suggested a pathologic lead point such as a Meckel’s diverticulum or a polyp. Child’s age, clinical context and ultrasonographic findings precluded hydrostatic reduction.

He was submitted to laparotomy and a double ileo-ileal and ileo-cecocolic intussusception was found. Manual reduction was easily performed. The lead points were two ileal doughnut-shaped intra-luminal masses at 2,25 meters and 1 meter respectively from ileocecal valve. The former had central depression with doubtful viability (Figura, a) and the latter had central perforation (Figura, b). A double segmental resection of the ileon encompassing each mass was performed, followed by double mechanical anastomoses.



Figure – a. Ileal mass that acted as intussusception leading point, with central depression; b- Ileal mass that acted as leading point, with central perforation.

The postoperative course was uneventful, and the patient was discharged home on sixth postoperative day. Pathology studies revealed an atypical Burkitt’s lymphoma. Abdominal and

thoracic scan performed at thirteenth postoperative day, detected additional masses at the medium right lung, head and uncinate process of the pancreas and both kidneys and thoracic, mesenteric and iliac lymph nodes. The patient responded well to antiretroviral therapy and chemotherapy and evolution was favourable. The patient completed the chemotherapy treatment about an year and a half ago and has not evidence of recurrence till this moment.

Discussion

Intussusception is a frequent cause of bowel obstruction in young children and the greatest incidence occurs in infants between ages 5 and 9 months⁴.

Double intussusception is an extremely rare variant of intussusception, which is almost impossible to diagnose preoperatively. The diagnosis is usually made during laparotomy and manual reduction is usually easily performed⁵.

Pathologic lead points occur in 4-8% of intussusceptions but are more commonly found in older children⁶. More than a half of the few reported cases of double intussusception in the child had an underlying pathologic lead point⁷.

Non-Hodgkin’s lymphomas are responsible for 17% of non-ideopathic intussusception⁸. They usually occur after the age of 3, are more frequent in boys and there’s often a 8-day history of symptoms with worsening patient status^{8,9}.

Intussusception triggered by a lymphoma is unlikely to be completely reduced by enema, and surgery for manual reduction is always required³. Following reduction, in the majority of cases, the diagnosis of lymphoma can be accomplished from peripheral samples such as peritoneal fluid, pleural effusion, mesenteric lymph nodes, bone marrow aspirate or by tumor biopsy^{2,9}. However, in single and localized disease, segmentar bowel resection should be considered only if it enables complete tumor removal, in order both to confirm the diagnosis and to reduce the intensity of chemotherapy^{2,3,10}. Complicated cases such as irreducible intussusception or with perforation or necrosis must be managed with segmentar resection⁹.

The diagnosis of lymphoma should be systematically evoked in children over the age of 3, especially if clinical or ultrasonographic findings are not typical^{2,9}.

According to literature, intussusception leads to early detection of intestinal Burkitt’s lymphoma and prognosis is favourable³.

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