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Case Report

Castleman's Disease Associated with Volvulus on Intestinal Malrotation in a 14-Year-Old Girl: a Case Report and Literature Review

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Abstract

Introduction: Castleman's disease is a clinicopathological entity characterized by uni- or pluricentric lymph node hypertrophy histologically corresponding to lymphoid angiofollicular hyperplasia. It is rare. Its exact cause is not known, and in about 20% of cases it has an intraabdominal location. Its association with digestive pathology has been rarely described. We reported a case of Castleman's disease associated with an intestinal volvulus on malrotation in a 14-year-old girl and we did the literature review.

Keywords: Castleman's disease; Unicentric form; Volvulus on malrotation; Adolescent

Introduction

Castleman's Disease or angiofollicular lymph node hyperplasia is a lymphoproliferative syndrome that can occur in two forms: localized or unicentric and multicentric. This is a rare condition [1]. Diagnosis is made by histology. The unicentric form is the most common form with a good prognosis. Malrotation is a congenital abnormality of intestinal rotation and fixation during fetal development. About 90% of malrotations are diagnosed in the first year of life, of which 80% in the first month of life [2]. It can be revealed early in life through duodenal occlusion (internal hernia) or volvulus. It can also remain asymptomatic throughout life. The association of Castleman's disease with a volvulus on malrotation is rarely described. We report a clinical case and make a literature review.

Case Presentation

A 14-year-old teenager, student, of longiline morphotype, without a particular history, has been received in outpatient examination for intermittent abdominal pain accompanied sometimes by late postprandial vomiting, weight loss, notion of anorexia and chronic constipation. On physical examination, the temperature was normal at 36.2°C, a weight of 45 kg, a height of 1.65 m. The body mass index was 16.54 kg/m^2. The thorax was harmonious and symmetrical.

The abdomen was flat and sensitive without mass or organomegaly. The lymph nodes were free. The rest of the organs were unremarkable. An abdominal ultrasound was carried out and highlighted in the epigastric area a roundel image corresponding to a large frange of jejunal invagination and a vascular turn of spire at the Doppler color. Mesenteric and supra-centimetric lymphadenopathies were also noted (Figure 1). Due to this notion of jejunal invagination that was not palpable at the clinical examination an abdominal computed tomography was performed and objectified acute bowel malrotation with sub occlusion and mesenteric lymphadenopathy (Figure 2 and 3). The diagnosis of intestinal malrotation with duodenal stenosis was retained and a preoperative assessment performed. In Biological examination hemoglobin was 12.3 g/dl, hematocrit 37.5%, PCR=01 mg/L, fasting glucose: 4.63 mmol, ALAT: 31UI/L, ASAT: 35UI/L, Serum creatinine level: 51.1 µmol/L, Urea: 2.7 mmol/L, Na+: 132 mEq/L, K+: 3.2 mmol/L, Ca+: 92 mg/L, Cl: 98 mmol/L, Mg+: 0.80 mEq/L, Bicarbonates 25 mmol/L. The procedure was performed in a scheduled surgery. Intraoperative exploration revealed a one spire loose volvulus of the jejunum with a stenosis of the duodeno-jejunal angle by adhesions; the third and fourth duodenums were dilated relative to jejunum with large adenopathies along the mesenteric vessels. The caecum wasn't fixed. We detorsed the bowel, divided further adhesive bands and Treitz ligaments and widened the root of mesenteric vessels. We did a mesenteric lymph node curettage that we found to be abnormal. The appendectomy was done. The small intestine was placed on the right side of the abdomen and the colon on the left side. The nodes were sent for anatomopathological examination. The immediate post-operative follow-up was simple and the patient was discharged from the hospital at day 5. The anatomopathological examination showed that the lymph node tissue had follicles centered by vessels. The lymphocytes were arranged concentrically. The

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sinuses and capsule were not infiltrated. In conclusion, this was the unicentric vascular hyaline form of Castleman's disease (Figures 4 and 5). We took the opinion of the pediatric oncologist. After 3 months of follow-up, an abdominal ultrasound performed was without any particularity. The evolution was good.

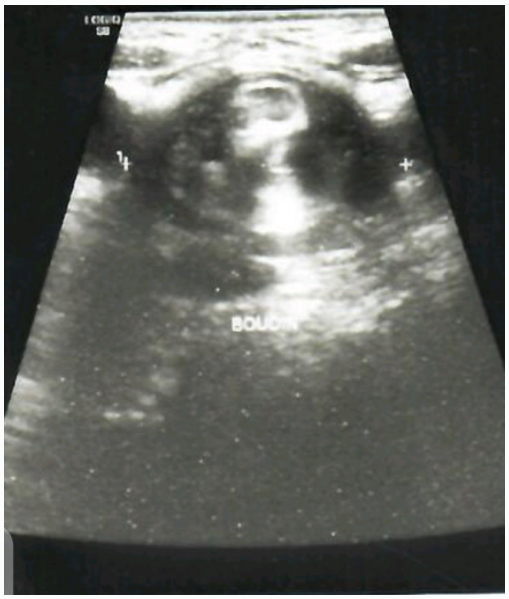

Figure 1: Ultrasound appearance of the invagination frange.

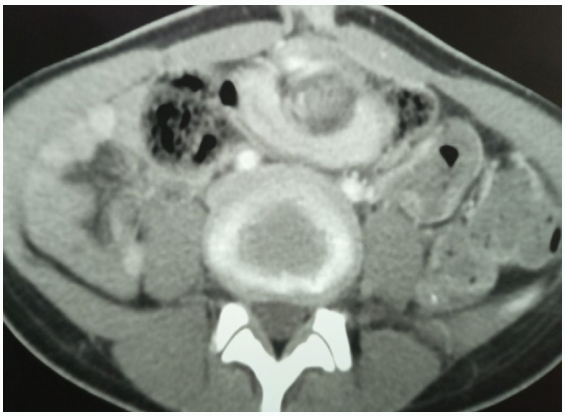

Figure 2: Abdominal CT showing interanse hypodense mass.

Discussion

Castleman's disease, initially described in 1954, is a clinicopathological entity characterized by uni or pluricentric lymph node hypertrophy histologically corresponding to lymphoid angiofollicular hyperplasia. It is also called hamartoma [3]. Its incidence is estimated at approximately 25 cases per million years/person, which represents less than 5,200 cases per year in the United States [1,4]. It can be diagnosed at any age. However, the unicentric form appears to have a higher incidence in patients in their 2nd-4th decade of life [5].

Both forms of Castleman's disease can be seen in both sexes at the same proportion. However, the unicentric form seems more common in women. Our case was about a 14-year-old girl. The exact cause of Castleman's unicentric disease is not known, and in about 20%

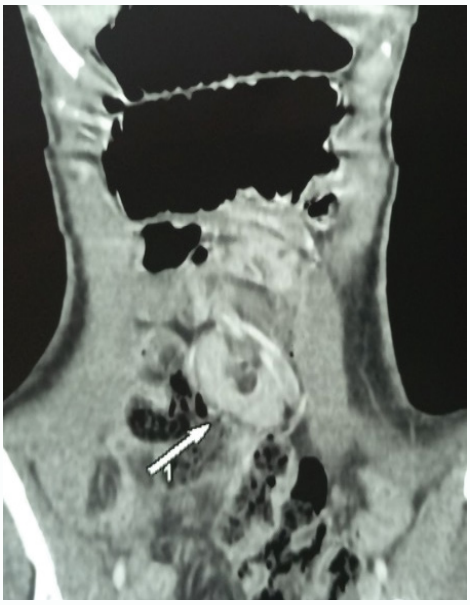

Figure 3: Abdominal CT: hypodense mass with dilated bowel suggesting subocclusion.

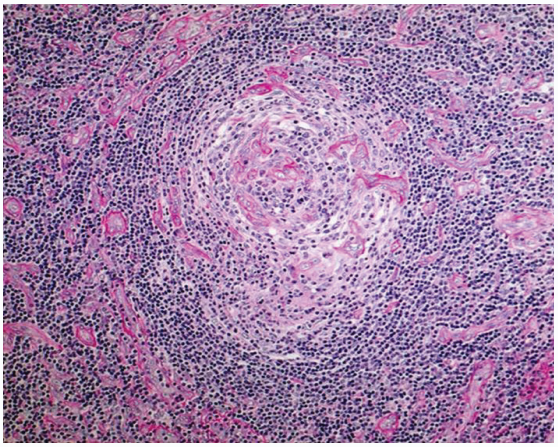

Figure 4: Histological aspect of a Castleman's disease (HE X10).

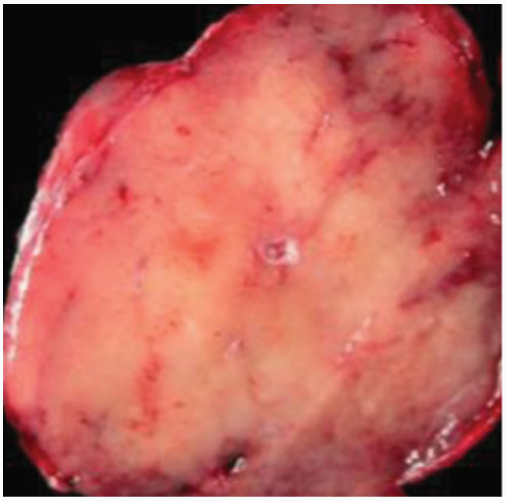

Figure 5: Macroscopic whitish appearance of Castleman's disease.

of cases its location is intra abdominal [6]. In general, the enlarged lymph node will be discovered accidentally during an examination for pathology, or in case of compression of a neighborhood organ. Often patients with the unicentric form can present various symptoms: fever, asthenia, sweating, weight loss, rash, hemolytic anemia and/or abnormally high amounts of certain immune factors in the blood (hypergammaglobulinemia). Our patient had episodes of abdominal pain sometimes accompanied by late postprandial vomiting, weight loss and notion of anorexia and chronic constipation. The association of Castleman's disease with digestive pathology has rarely been described [7]. Kartal et al. [8] described one association of Castleman's disease with intestinal obstruction. The symptoms presented by our patient were those of malrotation.

Malrotation can present a wide range of symptoms, from acute ischemic presentation of a midgut volvulus to a variety of other apparently unrelated abdominal complaints. The presentation and diagnosis of intestinal malrotation may not be simple, especially in older patients, so a high suspicion index is needed.

The diagnosis of all forms of Castleman's disease should be based on a thorough clinical assessment that includes a detailed history and a variety of specialized imaging techniques. Computed tomography may identify a non-invasive solitary mass in 50% of cases, or a primary invasive mass associated with adenomegaly or multiple adenomegaly without individualized primary mass, tissue density, intralesional calcifications, and possible central hypodensity (fat) [9]. MRI shows a hypersignal in T1 and T2, an intense and homogeneous contrast taking with feeder vessels [10]. For the diagnosis of volvulus by malrotation ultrasound can be of a capital contribution. Computed tomography identifies rotation patterns [11]. In our patient, abdominal ultrasound had found a large frange of jejunal invagination with a vascular turn spire in ultrasound Doppler color and a supra centimetric mesenteric and coeliac lymphadenopathy.

Abdominal CT had objectified acute bowel malrotation with subocclusion and mesenteric lymphadenopathy. Unicentric Castleman's disease is diagnosed by histopathological examination of the resected lymph node. On this examination, Castleman's disease in the hyaline vascular form known as the classic form is characterized by an abnormal germinative center with thickened-wall vessels; a modification of follicular dendritic cells, mantle lymphocytes concentrically arranged in onion bulbs and marked inter-follicular vascularization [12]. The anatomopathology examination of our patient's sampling showed the hyaline vascular form.

Surgical treatment remains the method of choice regardless of the patient's age. Complete removal of the lesion is a rule. Neoadjuvant chemotherapy with radiotherapy, Rituximab is performed if the tumor is unresectable. In case of incomplete resection adjuvant radiotherapy can be performed. The patient should be monitored for relapse cases reported in the literature [6]. For volvulus on malrotation, the Ladd procedure is recommended. In our patient, it was performed with curettage of lymph nodes considered abnormal. The child remains under surveillance.

Conclusion

Intestinal malrotation is a congenital disease. When associated with mesenteric lymphadenopathy it may be indicative of Castleman's disease. Lymph node removal remains the gold standard for the diagnosis of this pathology.

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