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Castelman's Disease Revealed by Cholestatic Jaundice: About a Case with Review of the Literature

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1. Summary

Castelman's disease is a lymphoid proliferation characterized by a wide histological spectrum and a polymorphic clinical picture. It comes in two different forms, localized and multicentric. We report an observation of Castelman's disease with a review of the literature in a 31-year-old patient who had consulted for cholestatic jaundice. It was a multicentric form with favorable evolution under corticosteroid therapy. The prognosis is different depending on whether it is a localized or multicentric form of the disease. Management depends on the clinical form involving medical and/or surgical management.

2. Introduction

Castleman's Disease (CD) is a rare lymphoproliferative disease first described by Benjamin Castelmanin 1956 [1]. It is a rare pathology with various forms of clinical presentation whose worldwide annual incidence is around 15.9 to 19.1 cases per 1 million inhabitants [2]. It can be unicentric castelman desease(UCD), manifesting as a mass of isolated lymph node origin, or multicentric Castelman desease (MCD), resulting in polyadenopathy [2].CD accompanied by jaundice is rare since there are only 12 cases reported in the literature [1].

We report through this work the case of an idiopathic multicentric Castelman's disease (MCDi) in a young patient, revealed by cholestatic jaundice.

3. Patient and Observation

3.1. Patient Information

A 31-year-old man, occasional alcoholic and smoking, was admitted to the gastroenterology department for cholestatic jaundice evolving for 2 months, associated with abdominal pain in the right hypochondrium, and vomiting in a context of apyrexia and deterioration in general condition.

3.2. Clinical Result

On admission, clinically, the patient was conscious, normocardic at 71 beats/minute and eupneic at 17 beats/minute, afebrile at 37°C. His body mass index was 23kg/m2. The jaundice was frank. Abdominal examination revealed tenderness of the right and epigastric hypochondrium, with splenomegaly (splenic arrow measured at 16cm). There were no peripheral adenopathies.

3.3. Diagnostic Approach

The patient underwent a biological assessment showing a biological cholestasis syndrome with a total bilirubin level of 323mg/l (direct bilirubin = 302mg/l), the alkaline phosphatases were at 296IU/l (N< 104IU/L). The complete blood count was without abnormality (hemoglobin at 13.5 g/dl, leukocytes at 7580/mm 3, and platelets at 173000/mm 3).

C-reactive protein was 19mg/l. Other biological parameters were normal including lactate dehydrogenase (LDH), alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), carbohydrate antigen (CA19.9).

Abdominal computed tomography showed hepatic hilar lymph node

magma and coelio-mesenteric region encompassing the hepatic pedicle with dilation of the upstream intrahepatic bile ducts, associated with homogeneous hepatosplenomegaly. The chest CT scan was without abnormality.

Abdominal computed tomography had shown hepatic hilar lymph node magma and the coelio-mesenteric region encompassing the hepatic pedicle with dilation of the upstream intrahepatic bile ducts, associated with homogeneous hepatosplenomegaly (Figure 1). The chest CT scan was without abnormality.

The radioguided biopsy was impossible given the depth of the lesion sheathing the vessels. An exploratory laparotomy was decided. During intraoperative exploration, multiple adhesions were found preventing access to the hepatic hilum, with the presence of subcentimetric mesenteric adenopathies. An excisional biopsy of a mesenteric lymphadenopathy was performed. Exploration of the rest of the peritoneal cavity was without notable abnormalities. The post-operative follow-up was simple.

Toon histological examination, it was a small lymphoid node. This one is of totally disturbed architecture.

Lymphoid follicles were variable in appearance. They are made of lymphoid elements of small regular sizes. Moreover, the architecture of this ganglion is also disturbed by the presence of vascular cavitary structures of variable size, in confluent places in favor of vascular hyperplasia (Figure 2a). The examination did not note any patent epithelio-gigantocellular granuloma or caseous necrosis with no evidence for a patent lymphomatous process.

Tothe immunohistochemical study, the hyperplastic follicular contingent intensely expressed the anti-CD20 antibody (Figure 2b) and the vascular contingent represented by the vascular cavities of variable size expressed intensely the anti-CD34 antibody.All its elements were in favor of an angio-follicular hyperplasia (Figure 2).

Absence of anti-HHV8 antibody expression was observed in vascular hyperplasia endothelial cells.All the elements were in favor of Castelman's disease of the angiovascular or hyalinovascular type.

HHV8, HIV, EBV and CMV serologies were negative. The negative quantifieron and the normal chest X-ray excluded tuberculosis, as well as the IGG4 assay was normal. Normal protein electrophoresis excluded monoclonal gammopathy and Poems syndrome was excluded due to the absence of association: polyneuropathy (P), organomegaly (O), endocrinopathy (E), monoconal gammopathy (M) and skin lesions (S). A myelogram was found to be normal.

Excluding the pathologies of infectious origin, autoimmune/autoinflammatory pathologies and hematological malignancies, we were able to retain the diagnosis of idiopathic multicentric Castelman's disease of the hyalino-vascular type, because it is a diagnosis of exclusion after eliminating other causes [4].

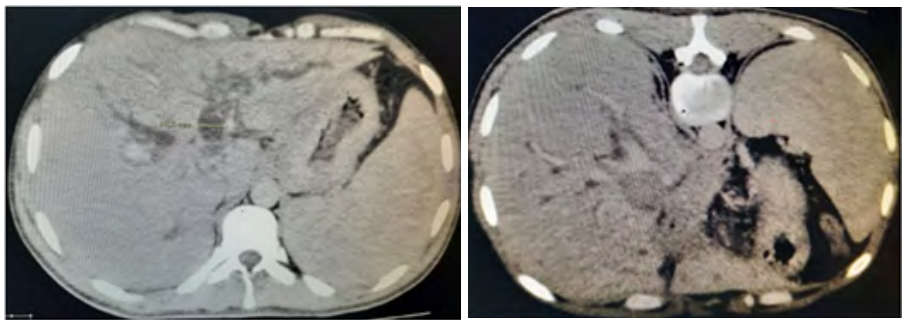

Figure 1: Abdominal CT :1: Axial view: Hilar lymph node magma on an axial view (a), dilatation of the intrahepatic bile ducts (b)

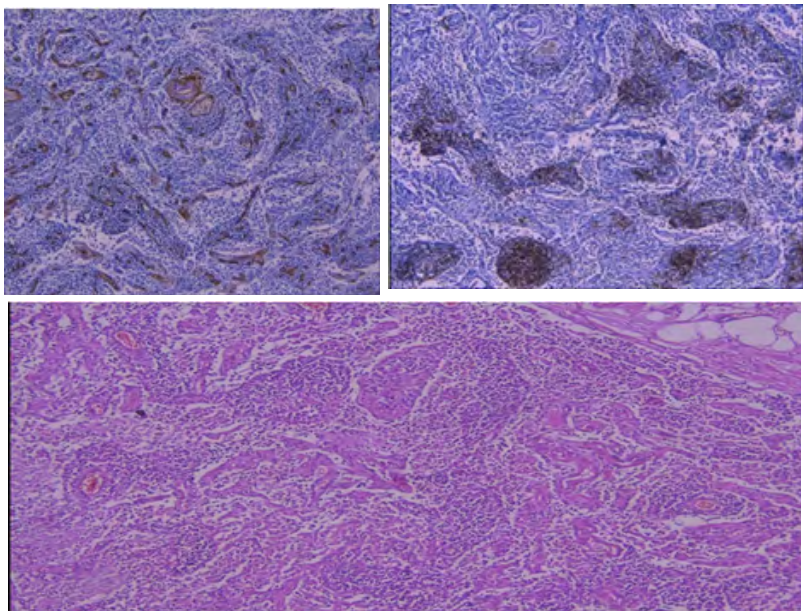

Figure2: Vascular hyperplasia (a), Follicular hyperplasia (b), Angio-Follicular Hyperplasia(C)

3.4. Therapeutic Intervention and Follow-up

The patient was treated with high-dose corticosteroid therapy with clinical and biological improvement (marked regression of jaundice, total bilirubin at 20mg/l, direct bilirubin at 15mg/l) after 1 month of treatment.

4. Discussion

In adults, cholestasis due to compression of the common bile duct by lymphadenopathy is rare, representing 3% of extrahepatic cholestasis [4]. In this context, the responsibility of a multicentric Castleman's disease is quite exceptional.

This benign atypical lymphoproliferation, considered a prelymphomatous state, is of mysterious etiology. It comes in two major clinical forms [5-6]:

- The multicentric form (MCD) is classically distinguished from UCD by the presence of systemic symptoms, such as fever, asthenia, pleural effusion, ascites, presence of lymph nodes in more than one region, hepatosplenomegaly, signs of inflammation and a poorer prognosis [7].

- The unicentric form (UCD):in about 60% of UCD cases, the presentation is asymptomatic with incidental detection of a mass. In some cases, UCD is found symptomatically due to compression of local structures by the enlarged mass [8].

Diagnosis of Castleman's disease involves lymph node biopsy on which an association of elementary histological lesions is observed. There are 3 types: the hyalinovascular or hypervascular type, the plasma cell type and the mixed type [7]. In the case of our patient, the immunohistochemical study showed vascular and follicular hyperplasia with rare plasma cell clusters.

It was a Hyaline-Vascular or Hyper-Vascular (HV) form. This histological form is marked by the importance of the regression of the germinal centers, the vascular anomalies and the anomalies of the network of the follicular dendritic cells [8]. It is the most frequent form of localized MC and very rarely in multicentric forms as seen in the case of our patient.

To date, 13 cases of CD presenting with jaundice, including the present case, have been described in 11 reports in the literature [2-7]. Of the 13 cases, the MCM and MCU were equally divided according to histological type; 7 (58.3%) were of the hyaline vascular type including our case, 4 (30.7%) of the plasma cell type and 2 (11%) of the mixed type [2].

All MCU cases were successfully resected and had complete resolution of symptoms during follow-up [8-09-10], while MCD cases were either partially controlled or relapsed [8-10].

Among the 7 cases of MCM revealed by jaundice, 2 patients benefited from surgical resection and chemotherapy with one case of relapse after 11 months of follow-up and another with unproven efficacy (patient lost to follow-up), 2 patients including our clinical case was treated with corticosteroids with good clinical evolution and regression of the mass, 3 patients benefited from surgery associated with corticosteroid therapy, of which 2 cases had partial control of the disease and 1 case of relapse with progressive multivisceral insufficiency and irreversible clinical deterioration leading to death [10].

Three forms of Multicentric Castleman's disease (MCD) are individualized: associated with an HHV-8 infection, with a "POEMS" syndrome or in the absence of these last two, we speak of a so-called idiopathic MCD (MCDi) [1]. Multicentric Castleman's disease, associated with HIV infection, is a distinct clinicopathologic entity characterized by the predominance of respiratory symptoms and by a greater association with Kaposi's sarcoma (64% of cases) [10].

HHV8 virus infection and cytokine regulation abnormalities may play a role in the pathogenesis of multicentric Castleman's disease, especially in HIV-infected patients [11]. In our case in front of the HHV-8 serologies, HIV negative and in the absence of POEMS syndrome, malignant hemopathies as well as autoimmune diseases (Lupus, IGG4 disease), inflammatory and in front of the clinico-biological improvement, we were able to retain the diagnosis of idiopathic multicentric Castelman's disease.

5. Conclusion

Castleman's disease is a lymphoid proliferation characterized by a wide histological spectrum and a polymorphic clinical picture. The prognosis is different depending on whether it is a localized or multicentric form of the disease. Management depends on the clinical form involving surgical and/or medical management.

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