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Autoimmune Hemolytic Anemia Complicating Castleman's Disease A Case Report

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Abstract **Case Report**

Anemia in Castelman's disease has multiple etiologies. The occurrence of autoimmune hemolytic anemia is rarely reported. We present the case of a 49-year-old patient known to have had Castelman's disease for 12 years, and who presented with acute hemolytic anemia. AHAI is believed to be secondary to a profound disorder of cellular immunity and to the production of autoantibodies by B cells under the effect of the overproduction of IL6. Treatment is usually with corticosteroid therapy.

Keywords: Autoimmune Hemolytic Anemia Complicating Castleman's.

INTRODUCTION

Castelman's disease is characterized by polyclonal B-cell proliferation of unknown etiology, associated with excessive interleukin 6 activity [6]. There are two clinical forms: the localized form, defined by involvement of a single lymph node site, and the multicenter form where several lymph node sites are affected. Castelman's disease classically presents with lymphadenopathy accompanied by clear general signs. Haematological disorders have also been described (anemia, thrombocytopenia) as well as visceral damage [3]. We present here the case of a patient followed for localized Castelman disease who presents with autoimmune hemolytic anemia.

OBSERVATION

We report the observation of a 49-year-old patient known to have been carrying unicentric Castelman disease for 12 years. A few days before hospitalization, the patient presented with a severe anemic syndrome with conjunctival jaundice, dyspnea and dark urine. The examination shows an asthenic patient with manifest skin-mucous pallor, conjunctival subicterus, left lateral cervical lymphadenopathy, 2-TDD splenomegaly without hepatomegaly. The remainder of the exam found no abnormalities. The biological assessment shows an anemia at 6g / dl normochromic normocytic without abnormalities of the other lines, with unconjugated hyper bilirubinemia, a collapsed haptoglobin and elevated LDH, a biological inflammatory syndrome with in particular an accelerated rate of sedimentation of red blood cells, a high CRP. The direct coombs test is positive. Kidney function is normal. The immunological assessment (antinuclear antibody, anti DNA) is negative. The morphological assessment, namely an abdominopelvic ultrasound, chest X-ray and thoracic-abdominal-pelvic tomodensitometry, did not find any abnormalities, in particular no deep tumor syndrome apart from homogeneous splenomegaly. An anatomopathological study of cervical lymphadenopathy confirms Castelman's disease. On corticosteroid therapy, the outcome was favorable with improvement in clinical and laboratory signs.

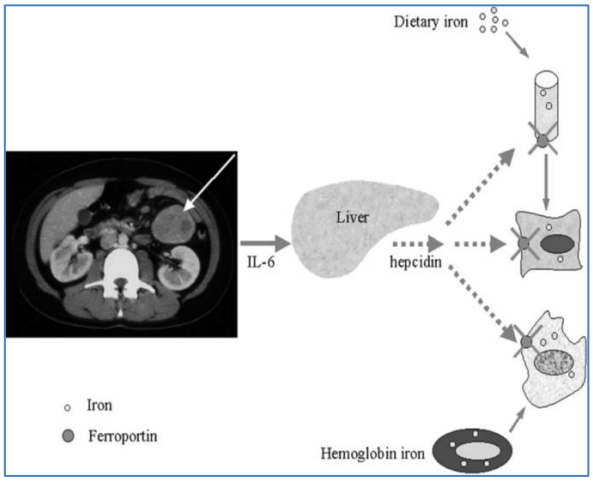

Fig-1

Fig.1 Proposed iron metabolism disturbance induced by Castleman disease. Hepcidin is an iron metabolism-regulatory hormone, which both inhibits the releaseof iron from hepatic macrophages and its intestinal absorption decreasing iron delivery to developing erythrocytes of bone marrow. Lymph node cells of Castleman disease (white arrow) produce interleukin-6 (IL-6) which in turn induces hepcidin production by hepatocytes. Secondarily, hepcidin rapidly inhibits ferroportin, a transporter that normally releases iron from hepatocytes, reticulo-endothelial macrophages and enterocytic duodenal cells, leading to hypoferremia associated with a decreased transferrine saturation. Iron is sequestrated in macrophages and intestinal iron transfer is decreased. In normal conditions (without IL-6 overproduction), low iron serum levels would have impede the hepatic transcription of hepcidin by a negative feedback loop, allowing enhanced released of iron from iron storage sites (liver, intestine).

DISCUSSION

Anemia in Castelman's disease is multifactorial (10): inhibition of erythropoiesis by TNF and IL1, alteration of iron metabolism, overproduction of IL6 which is a powerful stimulant of proliferation and differentiation B cells (9), in addition to autoimmune hemolytic anemia. Indeed, AHAI can be seen in many lymphoproliferative conditions, especially chronic lymphocytic leukemia. The presence of clinical autoimmune hemolytic anemia in CD is rare, despite a relatively frequent positive coomb test [4]. The association with autoimmune thrombocytopenia has been described leading to Evans syndrome [7]. The pathogenesis of this AHAI remains unknown. It could be a profound disruption of immune mechanisms [2] with in particular impairment of cell-mediated immunity as evidenced by the inversion of the CD4 + / CD8 + ratio found by Liberto NL et al. [5]. Another mechanism has been suggested as production of autoantibodies by B cells which proliferate and differentiate under the influence of interleukin 6 [8]. Different immunosuppressants have been tried in Castelman's disease associated with AHAI. Corticosteroids may be considered the treatment of choice, due to their effect on the immune systems and on the production of IL6 [1]. In some cases, multidrug therapy (CHOP type) has been used especially after failure of corticosteroid therapy. New therapeutic approaches have been described using monoclonal antibodies, in particular anti IL6.

CONCLUSION

The occurrence of anemia in Castelman's disease is due to multiple etiopathogenic mechanisms. The autoimmune character should be evoked in the presence, in addition to the positive coomb test, of clinical and biological signs of hemolysis. Treatment is primarily based on corticosteroid therapy. In case of failure, especially in multicenter forms, multidrug therapy should be initiated.

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