Acute anaplasmosis infection presenting with hemophagocytic lymphohistiocytosis in Quebec

INTRODUCTION

Human granulocytic anaplasmosis (HGA) is a rare, tick-borne illness with various clinical presentations. Ixodes scapularis is the main vector in Northeastern parts of the US and Canada.

The incidence of anaplasmosis has increased steadily in the past few years, from around a thousand cases in 2006 to 5,055 cases in 2019 [2] in the United States. HGA is most commonly reported in the upper midwestern and northeastern United States and happens usually in June or July. Starting in 2021, an unusual cluster of cases (25 reported cases over a year) has been described in the Estrie region of Quebec [3].

Hemophagocytic lymphohistiocytosis (HLH) is a rare presentation of various diseases (mostly infectious and neoplastic) including anaplasmosis that can require critical care.

OBJECTIVES

We herein describe a case of anaplasmosis presenting as HLH, a combination never reported in Canada.

60-year-old man living in the Estrie region, the southernmost region of the province of Quebec with no significant past medical history, no known infectious contacts and no medications presented to a referring hospital with fatigue, general malaise, high fever and rapidly progressing confusion evolving over two days.

METHOD

Physical examination
- 115/62, HR 83/min, RR 33/min, sat 94% with 4 L/min of O2 and T 38.3°C
- Toxic, slightly confused, but without nuchal rigidity, bilateral dysmetria
- Hepatosplenomegaly without peripheral adenopathies

Initial blood work
- Pancreatin (WBC 2.4 x 10^9/L, RBC 126 g/L, platelets 45 x 10^9/L)
- Normal renal function and normal electrolytes apart from a mild hyponatremia
- Normal liver function except from an elevated AST (199 U/L)
- Coagulation abnormalities (PT 1.24, aPTT 30.6 sec), elevated D-Dimer levels (17 854 mcg/L) and elevated LDH level (827 U/L)

RESULTS

72 hours after his initial presentation, the patient was transferred to our university health center. Upon arrival, further tests were performed, including a peripheral blood smear revealing neutrophils with cytoplasmic inclusion bodies suggestive of morulae [1] (picture 1). No schizocytes were found. Complementary analyses revealed an extremely elevated ferritin level (74 548 mcg/L) and hyperlipidemia (4.36 mmol/L), both suggestive of hemophagocytosis.

A bone marrow aspiration confirmed active medullary hemophagocytosis (picture 2). PCR for Anaplasma phagocytophilum confirmed the diagnosis of anaplasmosis.

Following the initiation of doxycycline, the patient’s condition rapidly improved and he was discharged home within 3 days. Doxycycline was prescribed for a total of 14 days. Upon follow-up 2 weeks after discharge, the patient reported complete resolution of all symptoms. All blood work abnormalities were resolved upon follow up 6 weeks after discharge.

CONCLUSION

The association between HGA and HLH has been rarely reported in the past [4,5,6,7]. This, to our knowledge, the first case of such association and presentation in Canada. Considering that anaplasma phagocytophilum has been reported in tick populations in all Canadian provinces and considering the increased incidence of HGA over the past few years, Canadian doctors should be aware of this disease. Clinical presentation of anaplasmosis is highly variable, including severe presentations that may require intensive care, hence critical care physicians may be confronted with the disease and should be cognizant of its presentations. HLH is an emergency to be recognized and treated as mortality remains high.

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REFERENCES


Images:

- Picture 1: Neutrophil with morulae
- Picture 2: Active medullary hemophagocytosis
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Hemophagocytic lymphohistiocytosis (HLH) is a rare presentation of various diseases (mostly infectious and neoplastic) including anaplasmosis that can require critical care.
OBJECTIVES

We herein describe a case of anaplasmosis presenting as HLH, a combination never reported in Canada.

69-year-old man living in the Estrie region, the southernmost region of the province of Quebec with no significant past medical history, no known infectious contacts and no medications presented to a referring hospital with fatigue, general malaise, high fever and rapidly progressing confusion evolving over two days.
METHOD

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• Toxid, slightly confused, but without nuchal rigidity, bilateral dysmetria
• Hepatosplenomegaly without peripheral adenopathies

Initial blood work

• Pancytopenias (WBC 2.4 x 10⁹/L, RBC 126 g/L, platelets 48 x 10⁹/L)
• Normal renal function and normal electrolytes apart from a mild hyponatremia
• Normal liver function except from an elevated AST (199 U/L)
• Coagulation abnormalities (PT 1.24, aPTT 30.6 sec), elevated D-Dimer levels (17 854 mcg/L) and elevated LDH level (827 U/L)
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REFERENCES


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