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Macrophage Activation Syndrome During Diabetic Ketoacidosis in A 9-Year-Old Girl: A Case Report

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Abstract | **Case Report**

Background: Diabetic ketoacidosis (DKA) is a serious and common presentation of type 1 diabetes in children, often life-threatening without prompt management. Macrophage Activation Syndrome (MAS), a rare but severe hyperinflammatory condition, is exceptionally uncommon during DKA episodes. ***Case Presentation:*** We report the case of a 9-year-old girl with type 1 diabetes who presented with her sixth DKA episode due to poor treatment adherence. Initial assessment revealed dehydration, altered consciousness, pleural effusion, generalized edema, and abdominal distension. Laboratory tests showed bicytopenia, hepatic dysfunction, elevated triglycerides, and renal impairment—features suggestive of MAS. Despite the absence of hyperferritinemia and hypofibrinogenemia due to technical limitations, the clinical picture met the diagnostic criteria for MAS. The patient improved with intravenous insulin therapy but relapsed shortly after discharge due to treatment interruption. ***Conclusion:*** This case underlines the importance of considering MAS in children with DKA and multisystem involvement. Early recognition and multidisciplinary care are vital to prevent severe outcomes.

Keywords: Diabetic ketoacidosis, Macrophage activation syndrome, Hemophagocytic syndrome, Type 1 diabetes, Pediatric case report.

INTRODUCTION

Ketoacidosis is the most frequent and feared manifestation of type 1 diabetes in children. It is the consequence of profound insulinopenia, and can be life-threatening and neurocognitive. Its prognosis depends on how it is managed.

The diagnosis of diabetic ketoacidosis (DKA) is based on the association of at least one of the following clinical signs: Polyuro-polydipsic syndrome, vomiting, abdominal pain, Küssmaul dyspnea, signs of dehydration and disturbance of consciousness with the following biological signs: Hyperglycemia: > 2g/l (> 11 mmol/l), blood PH < 7.3 or alkaline reserves < 15mmol/l, Glucosuria (≥ ++) - Ketonuria (≥ ++) or Ketonemia (blood beta-hydroxybutyrate concentration (BOHB) ≥ 3 mmol/l).

On the other hand, macrophagic activation syndrome or hemophagocytic syndrome (HS) is characterized by a prolonged febrile state, hepatosplenomegaly, cytopenia variably affecting all three lineages, and multivisceral involvement. Pulmonary involvement is common, and focal deficits, acute confusion, convulsions and even coma have been described. When present, kidney damage is terminal in 50% of cases. Cytopenia is a hallmark of the syndrome, occurring in 80-98% of cases, depending on the series. Liver tests are disturbed in almost all patients, and hypertriglyceridemia is common (69%). A paradoxical drop in fibrinogenemia is observed in 48% of cases. Thus, hypo-fibrinogenemia associated with an inflammatory syndrome and hyperferritinemia may point the way to a diagnosis. Testing for bone marrow haemophagocytosis is part of the work-up, but is neither sensitive, specific, sufficient nor necessary for the diagnosis of HS.

DESCRIPTION

After 18 months of diagnosis, the 9-year-old child was admitted for management of her 6th decompensation due to poor compliance with therapy. The clinical picture on admission was one of disturbed consciousness GCS: 9/15, febrile at 38.5, capillary blood

glucose: 3.5g/l, US: KU = (3+) and GU = (3+), Nitrite = (-), PU = (-), red blood cells = (-), weight and height were respectively according to curves (26kg= -1DS), (126cm= -1DS), dehydration (stage C)

PP Ex = polypnoea at 56 c/min with SaO2 at 95% on room air, pulmonary auscultation was in favour of pleurisy, Cardio. Vasc Ex = blood pressure: 100/60mmgh (75%/75%), Abd Ex = no diarrhoea, no vomiting, furthermore the patient presented a generalized oedematous syndrome made of oedema of the lower limbs (OLL) taking the cup and abdominal distension with dullness on percussion, abdominal distension with dullness visualized on ultrasound and abdominal CT scan, although the thoracic X-ray confirmed the left pleurisy of small abundance, a biological workup was initiated showing on blood ionogram: N+: 128 mg/l, K+: 4.2 Ca++: 88mg/d RA: 8, bicytopenia on CBC Hb 8.6 normochromic normocytic and thrombocytopenia 58. 000, WBC: 11900, PNN: 8200, Lym.: 2200, impaired liver function with PT: 38%, hypoalbuminemia: 23.7g/l, hypoprotidemia at 46.4g/l, renal workup: normal 24h proteinuria at 26mg/kg/24h, At D7 of hospitalization renal function impaired with urea figures 1.39, Creat 20.66. The sedimentation rate was elevated to 160 in the 1st hour, and a SAM workup was completed with a TG assay that returned a high 2.15. LDH and ferritinemia, due to lack of means, returned normal in a later phase. The evolution after IV insulin therapy was marked by good clinical evolution; disappearance of signs of acidosis, signs of respiratory distress, abdominal distension and edema of the lower limbs.

Five days after discharge, the patient returned in a state of shock with DAC on discontinuation of treatment. Anti-transglutaminase antibodies (IgA and total IgA) were not detected, and the patient was transferred to intensive care.

DISCUSSION

The clinical presentation of MAS is usually acute and can sometimes be dramatic, with the rapid development of multivisceral failure requiring admission to the intensive care unit. Often, the manifestations are not very specific, and it is their association that should prompt the diagnosis. At present, the Henter criteria are accepted as the diagnostic criteria for MAS, and the diagnosis of MAS is based on the presence of five of eight criteria: fever, splenomegaly, cytopenia (hemoglobin (Hb) < 9 g/dl, platelets < 100,000/mm3, neutrophils < 1,000/mm3), hypertriglyceridemia (> 3 mmol/l) and/or hypofibrinemia (< 1.5 G/l), hemophagocytosis marrow (or other tissues: lymph node, spleen, etc.), ferritin > 500 mg/mm3.), ferritin > 500 mg/l, soluble CD25 > 2,400 U/ml and no or reduced Natural Killer (NK) activity

In this case, the child presented with the stigmata of HS, with a subfebrile to febrile state (37.9 to 38.5) and multivisceral failure: hepatocellular insufficiency without HSMG, renal insufficiency with OLL, pleurisy and intra-abdominal effusion. Biologically, there was bicytopenia, elevated TG, hypoalbuminemia, ferritinemia and fibrinemia, which were requested but not performed due to lack of resources.

MAS often complicates autoimmune pathologies such as juvenile idiopathic arthritis (JIA), lupus erythematosus, Kawasaki disease and, in the literature, one case has been described of HS complicating diabetes in a 57-year-old man on ADO. Our work reveals macrophagic activation during ketoacidosis decompensation in type 1 diabetic children.

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