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PEDIATRIC ONCALL
CHILD HEALTH CARE

TEACHING FILES (GRAND ROUNDS)

ACUTE DONATH-LANDSTEINER HEMOLYTIC ANAEMIA (DL-HA) IN A 5-YEAR OLD MALE CHILD - HOW TO TREAT?

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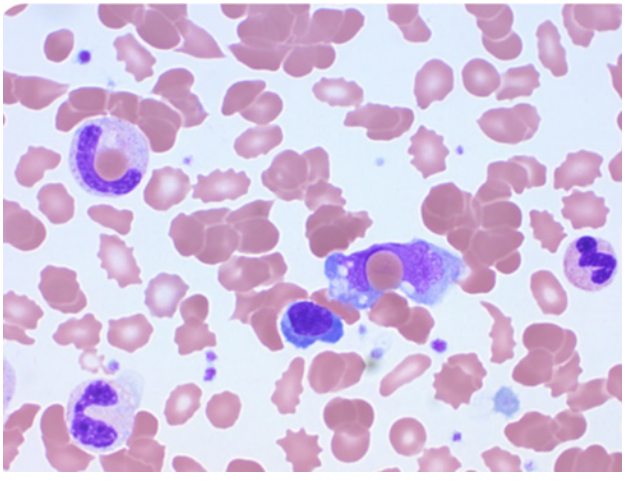
Clinical Problem:
A 5-year-old male present to the emergency department with fever for 8 days and acute onset of dark coloured urine. There was no bleeding from any other site. Five days ago, he was prescribed amoxicillin-clavulanate for infection of the ear and suspected tonsillitis. On examination, his temperature was 103.6°F, heart rate was 90/min, respiratory rate was 18/min, oxygen saturation was 98% at room air. He had jaundice and pallor. Systemic examination was normal. Investigations showed initial hemoglobin 10.4 g/dL which decreased to 5.8 g/dL after 37 hours. Haptoglobin was 12 mg/dl and serum indirect bilirubin was 1.8 mg/dl. Peripheral blood smear showed erythrophagocytosis (Figure 1). He was suspected to have acute Donath-Landsteiner hemolytic anaemia (DL-HA). A prompt Donath-Landsteiner (DL) test was performed, which confirmed the presence of a DL antibody and thus our diagnosis.

Figure 1. Peripheral blood smear test showing erythrophagocytosis



How to treat this patient?

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Discussion:
DL-HA represents 30-40% of autoimmune hemolytic anemia in children, that is generally self-limiting.[1] Autoantibody responsible for DL-HA is a cold-reacting immunoglobulin known as, DL autoantibody, capable of causing severe hemolysis even when the titre detected is low.[2] The DL autoantibody hold on tightly to red blood cell (RBC) surfaces during the peripheral circulation, where temperatures are cooler than 30°C in comparison to core body temperature.[3] After attachment to RBC surface, the DL autoantibody activates the complement cascade, leading to RBC membrane perforation and intravascular hemolysis, hence the dark coloured urine.[3] Complement activation and consequential hemolysis would become reality if binding RBCs travel to the core part of the body at a warmer temperature. Results of the direct antiglobulin test (DAT) with anti-C3 are likely to be positive, while negative for anti-IgG or anti-IgM.[3] In contrast, in cold agglutinin disease (CAD) IgM is positive.[3] The most prominent difference between DL-HA and CAD is this causative agent. Hence, a distinction must be established for proper diagnosis and treatment. Given that hemoglobinuria and personal history of travel to cold areas is not always present, diagnosis relies on lab testing.[2] A peripheral smear is always essential for diagnosis of hemolytic anaemia and may reveal spherocytes, or rouleaux formation suggestive of warm autoimmune haemolysis.[4] The best initial therapy for this patient is to keep him warm with gloves and warming blanket.[4] Warm intravenous fluids and red blood cell transfusion are vital for treatment.[4] If an underlying etiology is determined, it should be treated. The most likely etiology for this patient's DL-HA is the history of a previous viral illness and initiation of amoxicillin-clavulanate (drug-induced immune hemolytic anaemia). Though corticosteroids represent the first-line treatment for patients with autoimmune hemolytic anaemia, about 30% of patients require second-line treatment.[5] Plasmapheresis should only be used in severe cases, that are refractory to initial therapy.[4]

Compliance with ethical standards
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Conflict of Interest: None

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