CR77

Juvenile dermatomyositis with vasculopathy

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INTRODUCTION/OBJECTIVES: Juvenile dermatomyositis (JDM) is rare, but serious disease with many possible complications. Hallmarks of this disease are heliotrope erythema, periorbital edema and Gottrone papules combined with symmetric proximal muscle weakness that usually slowly progresses over period of weeks and months. In most cases the age of onset is between 4 and 10 years. There are four subtypes of the disease with variable course, organ involvement and long-term clinical outcome.

CASE PRESENTATION: We present a case of a 7-year-old girl with characteristic skin changes and signs of proximal muscle weakness. Her disease started six months prior to hospitalization with subtle changes in her normal everyday activity and periorbital erythema. One month prior to hospitalization she had difficulty with walking up the stairs combined with progressive overall fatigue. After full laboratory and imaging work-up were done, the diagnosis of the most severe type of JDM with vasculopathy was confirmed. Initial therapy, that consisted of glucocorticoids, methotrexate, intravenous immunoglobulins, cyclophosphamide and plasmapheresis, was not sufficient and progressive deterioration of symptoms and laboratory findings were observed. Finally, the administration of infliximab (anti-TNFα) stopped the progression of the disease and led to slow regression of symptoms in period of several weeks.

CONCLUSION: It is well-known that course of the JDM depends on clinically defined subgroups, early diagnosis, and aggressive initial therapy. Besides, there are increasing number of new discoveries about immunologic and genetic aspects of the disease. Combination of well-known and newly discovered aspects of the disease open up new opportunities for taking care advancement of JDM patients.

CR78

Diabetic ketoacidosis and electrolyte disorders in patient with short bowel syndrome following acute mesenteric ischemia

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INTRODUCTION/OBJECTIVES: Acute mesenteric ischemia (AMI) is defined as a sudden occurrence of insufficient blood supply to the intestine that can lead to necrosis (gangrene) of the intestine wall or its ischemia alone. It is therefore a life-threatening condition that requires rapid diagnosis and proper treatment. In presence of irreversible ischemic lesions the only treatment option is surgery. Short bowel syndrome commonly develops as a result of such treatment and can be a precipitating factor for the emergence of acid-base and electrolyte disorders.

CASE PRESENTATION: A 62-year-old male with a history of type 2 diabetes presented to the Karlovac General Hospital Emergency department with diffuse abdominal pain and diarrhea. Following diagnostic verification of upper mesenterial artery and coeliac truncus thrombosis and rapid exacerbation of patient's condition, explorative laparotomy, resection of the gangrenous part of the small intestine and enterostomy were performed. The postoperative course was complicated by the first diabetic ketoacidosis event. The patient was then transferred to the University Hospital Center Zagreb in order to perform end-loop resection and jejunoileal anastomosis. Shortly, another explorative laparotomy was urgently needed and previously mentioned procedures were repeated. Afterwards, the patient was placed in an intensive care unit with laboratory findings indicating a second diabetic ketoacidosis event (pH=7.05) and electrolyte disorders. In the following days, the patient was contactable and had regulated glycemia due to continuous insulin therapy, but febrile and with persistent electrolyte imbalance. He is still in life-threatening condition and is undergoing treatment.

CONCLUSION: The complexity of AMI treatment is shown in its equally complex complications.