Eosinophilic Ascites: a rare complication of autoimmune polyendocrinopathy

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Topic: multisystem endocrine disorders

Objectives:
Autoimmune polyendocrinopathy syndrome (APS) (orphan code ORPHA 282196) is defined as the autoimmune‐induced failure of at least two glands. APS is an inherited rare autosomal recessive disorder caused by mutations of the AIRE (autoimmune regulator) gene with organ‐specific autoimmune destruction of several, mostly endocrine. APS patients may develop autoimmune enteropathies and other intestinal dysfunctions. We describe a case of eosinophilic ascites in a nine‐year‐old girl with APS.

Methods:
The patient, an 8 year old girl, presented at the Emergency Unit with a four‐day‐history of a distended and painful abdomen, constipation and epigastric pain. She had a history of a parasitic infection due to Ascaris and giardia lamblia while living in Albania treated with antiparasitic agents and a three‐year history of autoimmune hypoparathyroidism and Addison's disease for which she was on hydrocortisone, fludrocortisone, calcium supplements, and VitD. She was apyrexial, had a distended abdomen and epigastric tenderness; no blood in stool, no vomiting. Her growth normal, Tanner stage 1.

Results:
Urea & electrolytes, liver function tests, bone profile, immunoglobulins, and urine were normal, CRP and ESR were mildly increased, D‐dimers 342 ng/ml (<500ng/ml), thyroid function was normal, PTH 0.8 (1,58‐6.03pmol/l), ACTH 75.110‐60pg/ml. Antibody(ab) screen: abs to parietal cells positive (1/40), antinuclear antibody (ANA) positive (1/80), adrenal abs positive (1/80). Immunophenotype: increased CD3+/CD4+, low CD3-/CD16 + 56+ lymphocyte counts compared to the child's age. Antibodies for parasites (Echinococcus, Toxoplasma, Toxocara) and stool cultures negative. Endoscopy and biopsy showed mild gastritis and duodenitis, colonoscopy was normal. Abdominal X‐Ray: increased amount of stool in the large bowel. Abdo CT and MRI: inflammation and fluid in the abdomen, around the stomach, liver, caecum and descending colon, mild spleen enlargement. She received a three‐week treatment with laxatives, probiotics, omeprazole, metoclopramide, erythromycin, metronidazole, amikacin, and ceftazidime with a positive response and a gradual reduction of CRP and ESR. However, no reduction of fluid was noted on ultrasound. Abdominal biopsy showed eosinophilic ascites (abundant eosinophils and their precursor forms); a 10‐day course of albendazole was given with no improvement. Diagnostic laparoscopy and drainage showed a liver cyst and multiple other cysts around the liver and abdomen area, full of fluid. One year later the patient has recovered, atrophic gastritis (figs 1 & 2) persists along with the other endocrinopathies and mild hyperhydropo货车iaemia.

Conclusions:
The eosinophilic ascites was probably caused by the original parasitic infestation. Autoimmune gastrointestinal disorders, which are relatively common among APS patients, are due to the immune reaction to the endocrine cells of the stomach and intestine (table 1). We believe that this was the case in our patient.

References:
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