58th Annual ESPE Meeting, 19-21 September 2019, Vienna, Austria

Eosinophilic Ascites: a rare complication of autoimmune polyendocrinopathy

Tatiani Moudiou[1], GeorgiosTsikopoulos[2], Vasiliki Bisbinas[3], Andreas Giannopoulos[1], Zacharoula Karabouta[1]

[1]2nd Paediatric Academic Department, University General Hospital AHEPA, Thessaloniki, Greece
Paediatric Surgery Department, Ippokrateio General Hospital, Thessaloniki, Greece
[3] Medical School, Aristotle University, Thessaloniki, Greece

Disclosure:No conflict of interest

Topic: multisystem endocrine disorders

Objectives:

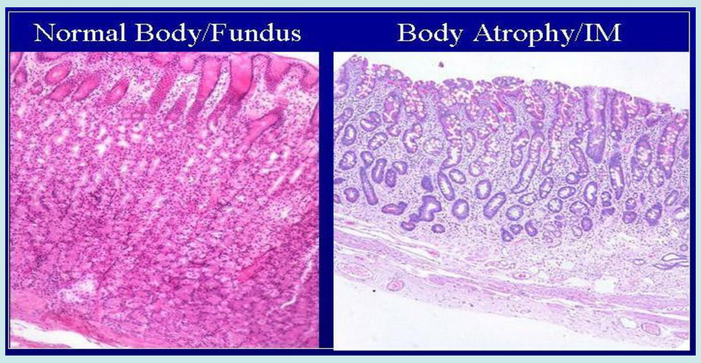
Autoimmune polyendocrinopathy syndrome (APS) (orphan code ORPHA 282196) is defined as the autoimmune-induced failure of at least two glands. APS is an inherited rare autosomal recessive disorder caused by mutations of the *AIRE* (autoimmune regulator) gene with organ-specific autoimmune destruction of 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 75.1(10-60pg/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-/CD (16 + 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 hyperthyrotropinaemia.



Figures 1 & 2.
Atrophic gastritis

APECED components	Tissue	Antigens
Addison's disease	Adrenals (cortex)	P450c21, P450c17a,P450scc
Hypoparathyroidism	Parathyroid glands	Ca^{++} sensing receptor*
Hypothyroidism	Thyroid gland	Thyroid peroxidase
		Thyroglobulin
Type 1 diabetes	Endocrine pancreas	GAD65, GAD67, ICA,IA-2 tyrosine phosphatase like protein
Autoimmune hepatitis	Liver	P450 CYP1A2, P450 CYP2A6,P450 CYP1A1, P450 CYP2B6 AADC
Vitiligo	Skin	SOX9, SOX10
Alopecia	Scalp	Tyrosine hydroxylase
Malabsorption	Gastrointestinal tract	Tryptophan hydroxylase
Autoimmune gastritis	Stomach	H^+K^+ ATPase
Pernicious anaemia	Gastric mucosa, red blood cells	Intrinsic factor

*This autoantigen in APECED has not been unequivocally proven.

Table 1.
Autoantigens in APECED

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:

-Frommer L, Kahaly GJ. Autoimmune polyendocrinopathy. J Clin Endocrinol Metab. 2019 Apr 26. pii: jc.2019-00602. doi: 10.1210/jc.2019-00602
-Arya P V A et al. Case of autoimmune polyglandular syndrome type 2: how we uncovered the diagnosis. BMJ Case Rep. 2019 Feb 26;12(2). pii: e227187. doi: 10.1136/bcr-2018-227187
-MO Bello; V V Garla. Polyglandular Autoimmune Syndrome Type I. StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2019
-Kluger N et al. Gastrointestinal symptoms in APECED syndrome. J Clin Gastroenterol. 2013 Feb;47(2):112-20
-Valensize M et al. Epidemiological and clinical peculiarities of polyglandular syndrome type 3 in pediatric age. Ital J Pediatr 2017;43(1):69. doi: 10.1186/s13052-017-0386-4
-Kahaly GJ, Frommer L. Polyglandular autoimmune syndromes. 2018 Jan;41(1):91-98. doi: 10.1007/s40618-017-0740-9
- Peterson P et al. Autoimmune polyendocrinopathy candidiasis ectodermal dystrophy (APECED): a model disease to study molecular aspects of endocrine autoimmunity. Clin exp Immunol2004 Mar; 135(3): 348–357