NEONATAL ADRENAL HEMORRHAGE PRESENTING AS BRYANT’S AND STABLER’S SIGNS

To the Editor:

A full-term male baby was born weighing 3020 g without fetal distress. The pregnancy was unremarkable. Apgar score was 9 at the first minute and 10 at 5 minutes. On his second day of life, scrotal bluish discoloration (Bryant’s sign) and inguinal ecchymosis (Stabler’s sign) were noted [Figure 1A]. Scrotal ultrasound showed a right scrotal hematoma with normal testes. Abdominal ultrasound revealed a well-defined heterogenous mass with a necrotic center on the upper pole of the left kidney, suggesting adrenal hemorrhage [Figure 1B]. The baby was treated conservatively. Coagulation screening tests were normal. No signs of adrenal insufficiency were found. Jaundice due to adrenal hemorrhage was noted and he was treated with phototherapy. The baby was discharged home on the fifth day of life in good condition.

Neonatal adrenal hemorrhage is associated with perinatal hypoxia, difficult delivery, shock, sepsis, and coagulation disorders [1]. Spontaneous and prenatal occurrences are well documented. The relatively large size and increased vascularity of the adrenal glands may be associated with their vulnerability to bleeding. Neuroblastoma is an important differential diagnosis of neonatal adrenal hemorrhage. Scrotal hematoma and inguinal ecchymosis are an uncommon presentation of this condition. Both signs are well-known signs of retroperitoneal hemorrhage. Extension of retroperitoneal bleeding may result in inguinoscrotal swelling. Intrapitoneal blood passes down the patent neonatal processus vaginalis, resulting in a hematocoele that may mimic neonatal testicular torsion. Right-sided adrenal hemorrhage is significantly more common than the left. A PubMed search revealed about 20 cases of scrotal ecchymosis associated with adrenal hemorrhage [2-5]. All the cases but one (contralateral) [5] were ipsilateral.

We describe the rare presentation of neonatal adrenal hemorrhage with left adrenal hemorrhage and contralateral right inguinal and scrotal ecchymosis. Scrotal hematoma often raises the suspicion of testicular torsion. However, in an appropriate clinical setting, the signs of scrotal discoloration (Bryant’s sign) and inguinal ecchymosis (Stabler’s sign) should raise the attention of the physician to retroperitoneal hemorrhage. This could help avoid unnecessary surgical intervention in the scrotum.

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References

CELIAC DISEASE IN AN ETHIOPIAN JEW

To the Editor:

Celiac disease is a permanent inflammatory disease of the small intestine triggered by the ingestion of gluten-containing cereals in genetically susceptible individuals. Until recently, gluten intolerance was thought to be a disorder that affects mainly people of European origin. However, case studies reporting clinical CD in immigrants from Africa and serological testing of various populations over the globe indicate that CD is a worldwide disorder. Additional studies revealed a high frequency of the HLA-predisposing genes like DR3-DQ2 in many parts of the world including developing countries. Despite the widespread susceptibility to CD, clinical disease may be undiagnosed in developing countries because of physician unawareness and atypical disease presentation [reviewed in (1)].

We recently diagnosed a 36 year old Ethiopian Jewish male with CD. A year earlier he complained of right lower and right upper abdominal pain that appeared each time after a 15 km run. He stopped running long distances. Several months later the abdominal pain worsened and became persistent and diffuse. There was no anemia or unintended weight loss. Colonoscopy and abdominal computed tomography were not informative. Only after one year of suffering did the patient

CD = celiac disease
undergo celiac serology tests (anti-tissue transglutaminase IgA, anti-endomysial IgA and anti-deamidated gliadin peptide antibodies), which were found to be positive. Molecular HLA testing revealed that he carries the susceptibility allele for CD: DQ81*02. Small bowel biopsy was compatible with stage 3a of the modified Marsh classification. Initiation of a gluten-free diet led to prompt clinical relief.

In the last three decades, more than 70,000 Ethiopian Jews immigrated to Israel. In addition, in the last two decades more than 36,000 refugees from Eritrea entered Israel. In Ethiopia and Eritrea, a major constituent of the diet is “injera,” a flat sour-like fermented bread [2]. In both countries injera is made from teff flour. Teff is a tiny round grain that is grown in Ethiopia and Eritrea. Teff flour is used in these countries for many purposes and substitutes for wheat flour in almost all its applications. The approximate composition of teff flour is 11% protein, 73% carbohydrates and 3% fat. Teff also contains thiamine, minerals like Fe, Mg, Ca, P and Cu, as well as essential amino acids like lysine [reviewed in (3)]. Teff has no gluten or gluten peptides and can be safely consumed by CD patients [2,4]. Teff flour is imported to Israel and can be purchased in special grocery stores, but due to its high price many of the Ethiopian and Eritrean residents prefer to consume wheat-prepared bread and other kinds of pastry and baked food. Therefore, it is expected that those Ethiopian and Eritrean residents who are genetically susceptible to gluten, and are asymptomatic when consuming teff-made products, will develop symptoms and signs of CD because of the above-described dietary shift. However, despite the hypothetical expectation of an increase in the number of CD patients among Ethiopian and Eritrean residents, no such increase has been observed. This may be related to a low level of awareness and clinical suspicion among Israeli physicians, a mild or atypical CD presentation, inaccessibility to medical services, or a combination of some or all of these factors.

The phenomenon of the “celiac iceberg” that exists worldwide may also be pertinent to adult Ethiopian and Eritrean residents in Israel. Increased awareness by Israeli physicians to the possibility of an increased risk for CD among these members of our society may help in reducing the magnitude of this iceberg.

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References

Capsule

Protective mucosal immunity mediated by epithelial CD1d and IL-10

The mechanisms by which mucosal homeostasis is maintained are of central importance to inflammatory bowel disease. Critical to these processes is the intestinal epithelial cell (IEC), which regulates immune responses at the interface between the commensal microbiota and the host. CD1d presents self and microbial lipid antigens to natural killer T (NKT) cells, which are involved in the pathogenesis of colitis in animal models and human inflammatory bowel disease. As CD1d crosslinking on model IECs results in the production of the important regulatory cytokine interleukin (IL)-10, decreased epithelial CD1d expression – as observed in inflammatory bowel disease – may contribute substantially to intestinal inflammation. Olszak et al. show in mice that whereas bone marrow-derived CD1d signals contribute to NK-cell-mediated intestinal inflammation, engagement of epithelial CD1d elicits protective effects through the activation of STAT3 and STAT3-dependent transcription of IL-10, heat shock protein 110 (HSP110, also known as HSP105), and CD1d itself. All of these epithelial elements are critically involved in controlling CD1d-mediated intestinal inflammation. This is demonstrated by severe NKT cell-mediated colitis upon IEC-specific deletion of IL-10, CD1d, and its critical regulator microsomal triglyceride transfer protein (MTP), as well as deletion of HSP110 in the radio-resistant compartment. Our studies thus uncover a novel pathway of IEC-dependent regulation of mucosal homeostasis and highlight a critical role of IL-10 in the intestinal epithelium, with broad implications for diseases such as inflammatory bowel disease.

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“Fiction gives us a second chance that life denies us”
Paul Theroux (b. 1941), American travel writer and novelist

“I don’t mind that you think slowly but I do mind that you are publishing faster than you think”
Wolfgang Pauli (1900-1958), Austrian theoretical physicist and one of the pioneers of quantum physics