Celiac disease in patient with Turner syndrome

Turner sendromlu hastada çölyak hastalığı

To the Editor,

Turner syndrome (TS) is a genetic disorder associated with abnormalities of the X chromosome (1). Individuals with TS are prone to develop autoimmune conditions such as Celiac disease (CD), thyroiditis and type 1 diabetes mellitus (2). The incidence of CD increases 11-fold in TS (3).

A 29-year-old female was admitted to the hospital due to growth retardation and primary amenorrhea. On physical examination, no breast development or any other secondary sexual characteristics were prominent. The karyotype analysis revealed 45X0 consistent with Turner stigmata. She was consulted to our gastroenterology department in order to determine the etiology of her anemia. She had no gastrointestinal complaints except for gastric discomfort after meals. The laboratory findings revealed hypochromic microcytic anemia, hypocalcemia, hypophosphatemia, and elevated serum alkaline phosphatase levels. Serum 25-hydroxy vitamin D level was low (6.3 ng/ml) and accompanied by an elevated parathormone level of 223.6 pg/ml (normal range, 10–65 pg/ml). Transferrin saturation was 3.42 and the serum folate level was under the normal limits (2.7 ng/ml). High levels of antigliadin immunoglobulin (Ig)A and IgG and antiendomysial IgA antibodies were also detected. Upper gastrointestinal endoscopy demonstrated the presence of atrophic gastritis and scalloping of duodenal folds, and the duodenal mucosa was pale and edematous. On pathologic examination, subtotal villous atrophy and increased number of intraepithelial lymphocytes in the duodenal mucosa were detected. The femur neck and lumbar vertebrae were osteoporotic with an increased risk of spontaneous fracture revealed by bone mineral density measurement with dual-energy X-ray absorptiometry (DEXA) (T scores of -5.4, -3.7, respectively). A gluten-free diet for CD was prescribed. Replacement treatment with iron, calcium, and vitamin D was initiated.

The prevalence of EMA positivity detected by screening in TS is 4.2% (4). Reviewing the data in the TS population, serological screening appears to be an effective method of identifying subclinical CD.

Most of the patients diagnosed with TS who also have growth retardation do not respond to growth hormone therapy if they have coexisting CD. On the other hand, some of the patients with CD who have persistent growth retardation and pubertal immaturity despite a gluten-free diet are diagnosed with TS afterwards (5). The available data and publications indicate that screening for CD should be performed in patients with TS, and intestinal biopsy should be carried out in patients with positive results (6). The early diagnosis makes it possible to initiate an appropriate hormonal treatment to achieve normal growth and to induce puberty.

In conclusion, considering that the patients with TS have a high prevalence of autoimmune disease, an early investigation for CD should be carried out in those with TS.

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Is intensive phototherapy a risk factor for pathogenesis of intussusception?

Yoğun fototerapi, intussepsiyon patogenezinde bir risk faktörü sayılabilir mi?

To the Editor,

Intussusception in the newborn is reported rarely, accounting for only 0.3% of all cases of intussusception (1). The ileocecal region is most commonly affected. The tetrad of clinical features includes sudden onset of abdominal colicky pain, bloody stools, a palpable abdominal mass, and vomiting. In full-term infants with neonatal intussusception, a pathologic lead point is found in one-third of the cases and the colon is typically involved (2). The etiological factors include duplication cyst, hamartoma or Meckel's diverticulum (3).

The 2800 g firstborn of a set of 38-week male twins was delivered by elective cesarean section to a 28-year-old gravida 2, para 1 woman. A normal pregnancy period was noted in routine visits. In the infant's early neonatal course, no problem was seen. Meconium was passed on the first day. On day 4 of life, he was admitted to the hospital for phototherapy for indirect hyperbilirubinemia (peak total bilirubin 19 mg/dl). After two days of intensive phototherapy, the baby was noted to have mild abdominal distension. There was some epigastric distension, but no masses were palpable. There was a large gush of soft, jelly-like dark-red feces after the rectal examination. Plain radiographs of the abdomen showed a small quantity of gas in the stomach and small intestines (Figure 1). Abdominal ultrasonography was normal. Exploratory laparotomy was done without further investigation because of his rapid clinical deterioration (hypoperfusion/shock-like syndrome). An ileocolic intussusception, which could not be reduced manually, was detected (Figure 2). Six centimeters of bowel appeared nonviable and was resected. The pathology of the resected segment showed mucosal hemorrhage, necrosis and congestion. There was no lead point for the intussusception. The patient was tolerating enteral feeds by day 4. He was discharged from the hospital at the age of 40 days in good general condition. His weight at discharge was 3495 g. Although intussusception is rare in the newborn, it is a relatively common surgical emergency in infants and young children, with an incidence of 1 to 2 per 1000 births (2). The etiology of intussusception in most infants remains unclear. More than 90% of the cases of ileocolic intussusception are idiopathic, without an obvious lead point (3). While abdominal ultrasound is a very useful tool for diagnosing intussusception in older infants, it has not yet been established in the investigation of newborns. In full-term as well as preterm infants, intussusception is predominantly localized in the small bowel; contrast enema is of no value and may even be harmful, because frequ-

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