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A RARE CASE OF PARANEOPLASTIC SYNDROME: VANISHING BILE DUCTS IN A KID WITH HODGKIN LYMPHOMA

Lyubov Smirnova, Lyubov Shakhgildyan

Pirogov Russian National Research Medical University, Moscow, Russiaa

BACKGROUND

The incidence of liver involvement in patients with Hodgkin lymphoma (HL) is about 50%. Cholestasis in HL can be caused by direct intrahepatic damage to the liver parenchyma or bile duct epithelium by tumor cells, as well as extrahepatic obstruction of the biliary tract by enlarged lymph nodes (LN). In addition, in a small number of patients intrahepatic cholestasis is caused by paraneoplastic syndrome.

Purpose:

This clinical case demonstrates an example of hyperbilirubinemia as a rare manifestation of paraneoplastic syndrome in children.

MATERIALS AND METHODS

In autumn 2016 a 15-year-old girl noticed an enlarged right supraclavicular LN. A dynamic monitoring was recommended. In spring 2017 she was first hospitalized with jaundice: total bilirubin (TB) was 500 μmol/L, transaminases — 300 IU/L. In April the diagnosis of liver fibrosis of unclear etiology was made with no improvement on subsequent hepatotropic therapy. In June the subclavicular LN continued to enlarge and the biopsy showed HL (histologically nodular sclerosis type 2). According to the results of sonography and PET/CT subclavicular (5×5 cm conglomerate), mediastinal (7,5×5,5 cm conglomerate) LN and lung tissue were involved. So, the diagnosis of IV EB stage of HL was made. On abdominal ultrasound the liver was mildly enlarged with hyperechoic, finely grained parenchyma; portal vein and bile ducts were normal. The patient was transferred to Institute of Pediatric Oncology. On admission she had severe intoxication and skin itch, bronze-colored skin and icteric mucosa and sclera, liver was 2 cm below the costal margin, BMI=14,7 kg/m^2. Lab results were: Hb 107 mg/L, WBC 19×10^9/L, PLT 531×10^9/L, TB 302 μmol/L, alkaline phosphatase 714 IU/L, ALT and AST — 150 IU/L, LDH 398 IU/L. It was considered that liver was affected by paraneoplastic syndrome — vanishing bile ducts syndrome.

RESULTS

The patient underwent 3 courses of chemotherapy and 2 courses of radiotherapy. Although none of the courses were completed due to developed complications, the excellent effect of the treatment was achieved: enlarged LN were not detected on imaging in any site. But the liver function tests worsened: TB was 628 μmol/L, GGT was 20 times elevated, transaminases — by 10 times.

Due to the full effect of the treatment of HL but persisting irreversible changes caused by paraneoplastic syndrome the liver transplant from mother was performed. Pathology confirmed vanishing bile duct syndrome: prolonged intrahepatic cholestasis, dystrophy and desquamation of the epithelium of small bile ducts. After surgery hyperbilirubinemia recurred that was stopped by methylprednisolone, IV immunoglobulin and a single administration of rituximab. As a result of the treatment complete normalization of biochemical parameters was achieved, complete remission of HL remained.

CONCLUSIONS

In case of prompt removal of causative factor bile ducts are capable of regeneration. However, in this case there was a severe damage to the bile ducts which led to irreversible liver damage and refractory hyperbilirubinemia, which persisted even after a complete remission of the malignant process. It must be remembered that jaundice in HL can be one of the manifestations of paraneoplastic autoimmune process that leads to damage of the epithelium of the intrahepatic bile ducts (especially when there is no tumor tissue seen on imaging).

Keywords:

chemotherapy, idiopathic cholestasis, liver transplantation