Acquired Unilateral Nevoid Telangiectasia Syndrome Accompanied by Chronic Hepatitis B Virus Infection

Unilateral nevoid telangiectasia syndrome (UNTS) is a rare disease characterized by superficial telangiectasias in a dermatomal distribution. It is generally unilateral and frequently distributed in the C3-C4 or trigeminal nerve dermatomes (1). There are various opinions on the pathogenesis, which can be congenital or acquired. Here, we intended to make a new contribution to the pathogenesis of the disease by presenting a male patient with UNTS accompanied by chronic hepatitis B virus infection.

An 18-year-old male patient presented to our outpatient clinic with asymptomatic telangiectasias that appeared 3 years previously and increased gradually. He had no systemic diseases or history of alcohol consumption. Physical examination was normal. Dermatologic examination revealed a few fine, branching unilateral telangiectasias located on the left half of the upper lip, left upper and lower eyelids, and left side of the neck, which were in accordance with the C3 dermatome, ophthalmic and maxillary branches of the trigeminal nerve (Fig. 1a, b). Complete blood count, erythrocyte sedimentation rate, urine analysis, renal and hepatic parameters, thyroid function tests, bleeding and coagulation parameters, blood estrogen level, anti-nuclear antibody and thyroid autoantibodies, and HIV serology were within the normal range. Hepatitis B surface antigen (HBsAg), HBVe antibodies and HBV-DNA were positive, which led to the diagnosis of chronic hepatitis B. There was no evidence of liver cirrhosis. Punch biopsy of the lesional skin revealed fine, dilated vessels located in the middle and upper dermis (Fig. 2). Immune reactivity was negative for estrogen and progesterone receptors. Taking into consideration the distribution of the lesions, histopathology and laboratory test results, the patient was diagnosed as UNTS accompanied by chronic hepatitis B.

UNTS is thought to be associated with physiological and pathological hyperestrogenemic states such as pregnancy, puberty, hepatic disease, and hormonal therapy. However, Wenson *et al*. (1) showed that hormonal theory was not valid for all cases and there

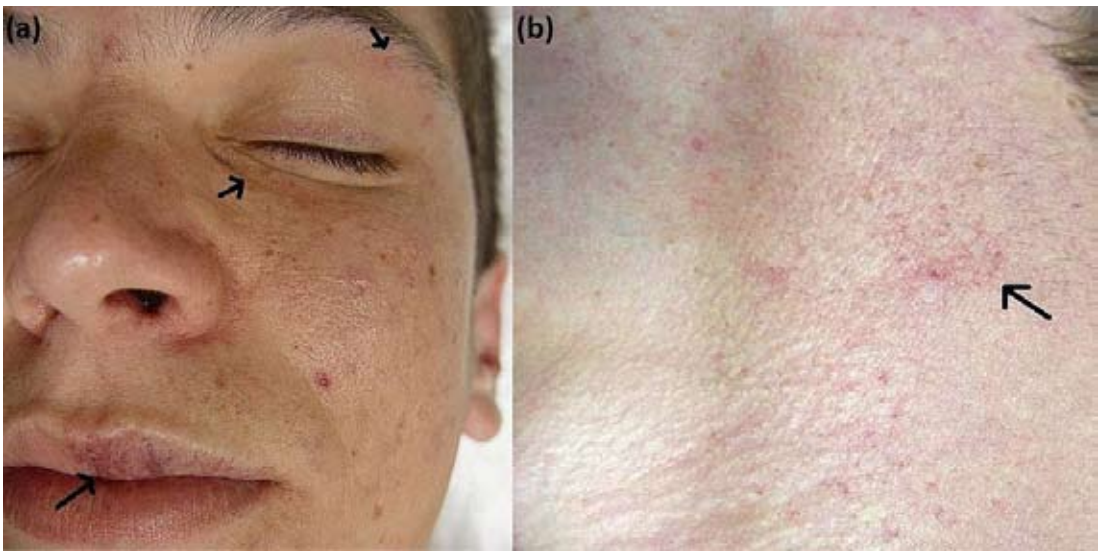

Figure 1a, b. Telangiectasias located on the upper lip, left upper and lower eyelids, and left side of the neck.

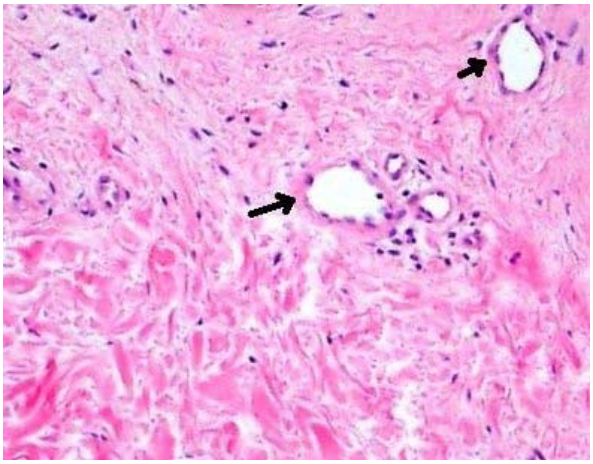

Figure 2. Dilated vessels in the dermis (hematoxylin-eosin, x40).

were cases with no underlying disease or hormonal abnormality. Even though our patient had accompanying hepatitis B infection, there was no concomitant hormonal pathology. There are a few UNTS cases associated with hepatitis B or C infection in the literature. Almazan Fernandez *et al*. (2) report on a male patient with UNTS and chronic hepatitis B in non-replicative phase without a sign of cirrhosis. Karakas *et al*. (3) report on a female hepatitis B carrier with UNTS. Hynes and Shenefelt (4) report on two male UNTS patients with hepatitis C without a sign of cirrhosis. In all three reports, serum estrogen levels and tissue estrogen/progesterone receptor levels were normal. These authors agree that the somatic mosaicism is the cause of UNTS and vague telangiectasias become visible when estrogen levels increase, either physiologically or pathologically. Different from these case reports, our case had a detectable viral load and he was a chronic hepatitis B patient, not a carrier. Even though the effect of estrogen has been referred to in those reports, neither those patients nor our patient had elevated estrogen levels. Therefore, a clear relation between UNTS and hepatitis without a sign of cirrhosis could not be established.

Vascular endothelial growth factor (VEGF) is a protein that plays a critical role in vascular development and new vessel formation. In a study on serum VEGF levels in various hepatic diseases, the authors reported that acute hepatocellular damage caused significant amounts of VEGF to be released into the circulation (5). In a similar study, Makhlouf *et al*. (6) report that VEGF levels were higher in patients with chronic hepatitis and cirrhosis than in healthy controls. Therefore, we believe that there is a somatic mosaicism in UNTS accompanied by hepatitis; however, telangiectasias may occur in response to elevated serum levels of angiogenic factors such as VEGF rather than estrogen. Although in this case we could not perform an ELISA test to detect serum VEGF levels due to inadequate laboratory conditions, we think that the hypothesis of VEGF relation with UNTS may shed light on the next studies on the UNTS etiopathogenesis.

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