

A Case of Secondary Hemochromatosis



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ABSTRACT

A 28-year-old woman with a history of transfusion-dependent thalassemia major who did not receive any iron-chelating agent presented with symptoms of iron overload with marked diffuse increased liver density. Eventually, the diagnosis of secondary hemochromatosis was done based on the aforementioned history, laboratory, and imaging findings.

Case presentation

A 28-year-old woman with a history of transfusion-dependent thalassemia major who did not receive any iron-chelating agent and recently experienced an episode of diabetic ketoacidosis presented with abdominal pain, distension, and weakness. Physical examination revealed ascites, elevated jugular venous pressure, lower extremity edema, and dark skin.

Laboratory data showed normal serum glucose, pseudo-lymphocytosis (due to increased nucleated red blood cells), normocytic anemia, serum ferritin level of 22,000 ng/mL, biochemical evidence of cirrhosis, and positive serologic test for hepatitis C virus (hypoalbuminemia, high serum-ascites albumin gradient with low protein ascetic fluid, coagulation test disturbances, aspartate aminotransferase [133 U/L], and alanine aminotransferase [91 U/L]). The renal and thyroid function tests were normal.

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Figure 1. Axial Non-enhanced Computed Tomography

Shows hepatomegaly with marked diffuse increased density, the spleen is absent.

Abdominal ultrasonography was compatible with hepatomegaly and cirrhosis. Echocardiography revealed restrictive cardiomyopathy.

Non-enhanced Computed Tomography (NECT) scan showed cardiomegaly and hepatomegaly with marked diffuse increased liver density (Figures 1 and 2). The prominence of the interventricular septum in NECT is in favor of the patient's anemia [1] (Figure2). Note also the hair-on-end appearance of the skull (Figure 3).

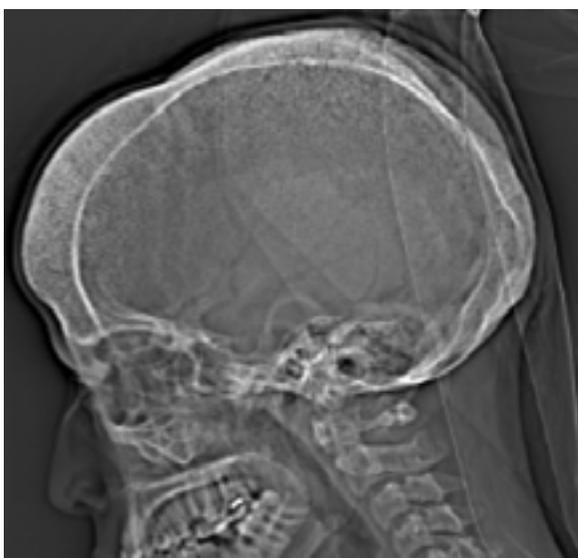


Figure 3. Computed tomography lateral scout view shows thickened calvarium and hair-on-end appearance (due to thickened trabeculae)

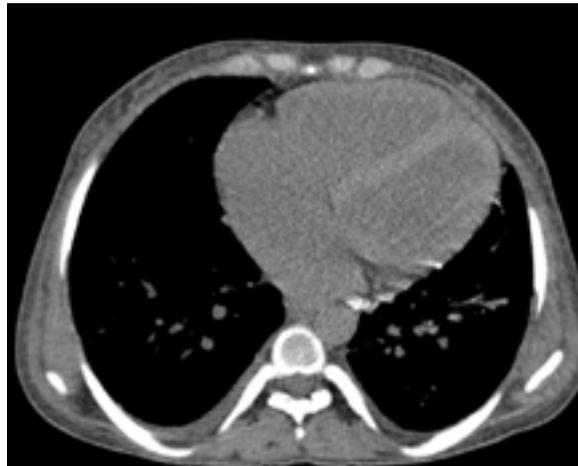


Figure 2. Cardiomegaly, note interventricular septum is visualized on unenhanced CT

Compatible with anemia (although not specific nor sensitive finding).

The diagnosis of secondary hemochromatosis was done based on the aforementioned history, laboratory, and imaging findings. The abdominal CT scan in hemochromatosis shows diffuse hyperattenuating liver [2]. Although this imaging finding is almost specific for iron overload, rare conditions such as chronic amiodarone consumption, glycogenosis, and Dubbin-Johnson syndrome could also elevate liver density [3].

Ethical Considerations

Compliance with ethical guidelines

All ethical principles were considered in this article

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Conflict of interest

The authors declared no conflict of interest.

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