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## Letter to the Editor

### Idiopathic Hypereosinophilic Syndrome with Hepatic Tumoral Lesions

To the Editor,

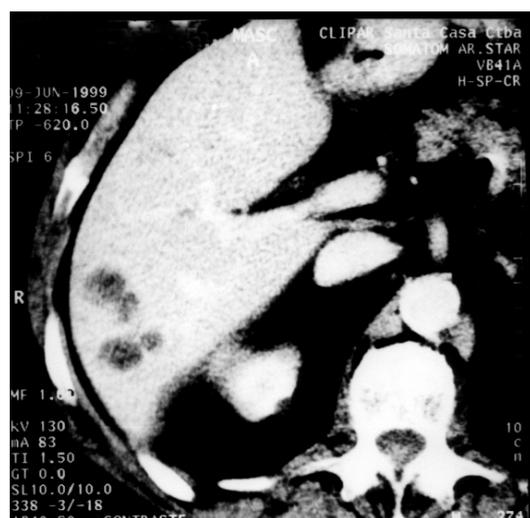
Idiopathic Hypereosinophilic Syndrome (IHS) is characterized by a sustained overproduction of eosinophils and predilection to damage specific organs (heart, lungs and liver) and systems (central and peripheral nervous and skin)<sup>[1]</sup>.

A 73-years old man was admitted on March 1997, complaining of asthenia, muscle weakness, dry cough, bronchospasm and paresthesia of arms and legs. A WBC count was  $13.5 \times 10^9/l$  with 44% of mature eosinophils. A blood test performed 7 months earlier revealed similar values. The neurological diagnosis was an axonal peripheral polyneuropathy (motor and sensitive). Stool examinations, IgE dosage, CAT scan of the brain and the thorax, echocardiogram and blood chemical values demonstrated no abnormalities. However, the CAT scan of the abdomen revealed hepatic tumors. A neoplasm of the liver with secondary blood eosinophilia was considered, and the tumor masses aspirated through a fine needle guided by CAT scan. The analysis of the aspirated material revealed infiltration by mature eosinophils. The diagnosis was IHS with hepatic tumoral lesions (HTL), and hydroxyurea (2 g per day) was started on April 1997. Six months later, the WBC count was  $4.5 \times 10^9/l$  with 5% of eosinophils. The patient had a pronounced clinical improvement, with weight gain (more than 10% over the previous weight), disappearance of the bronchospasm and recovery of neurological symptoms. A CAT scan of the abdomen, performed on June 1999, revealed reduction of the tumoral lesions (Figure 1). The patient is still receiving hydroxyurea, has normal eosinophils at WBC and re-

mains with a Karnofsky score of 100%.

IHS is defined according to three criteria: (I) presence of blood eosinophilia greater than  $1.5 \times 10^3/l$  for more than 6 months, (II) evidence of end-organ dysfunction, (II) and exclusion of known causes of eosinophilia (parasitic infections, allergic reactions, adrenal insufficiency, neoplastic and collagen vascular diseases)<sup>[2]</sup>. The overexpression of cytokines like IL-3, G-CSF, M-CSF and IL-5 is related to the IHS pathogenesis. The eosinophils activated mainly by IL-5 express additional adhesion molecules (like leukocyte function associate antigen-1, MAC-1 and VLA 4) components of the integrins family, which well determine eosinophil deposition at specific end-organs. The eosinophil deposition in the tissues is followed by the release of its granules content (major basic protein, eosinophilic peroxidase, cationic eosinophilic protein and eosinophils derived-neurotoxin), that mediate the tissue damage<sup>[3]</sup>.

Hepatic involvement in IHS occurs in 35% of the patients. Chronic active hepatitis, Budd-Chiari syndrome secondary to hepatic vein obstruction<sup>[4]</sup>, intrahepatic cholestasis<sup>[4]</sup> and sclerosing cholangitis<sup>[5]</sup> have been reported as common forms of hepatic involvement. Damage to hepatocytes and/or to the biliar system, due to a diffuse infiltration of the hepatic parenchyma by eosinophils, is the histological picture. Hepatic nodules, constituted by eosinophils, have been previously reported secondary to parasitic infetions<sup>[6]</sup>. Hepatic tu-



**Figure 1.** Abdominal CT scan (post-treatment) with remaining hypodense hepatic tumoral lesions.

moral lesions of the liver due to hepatocellular carcinoma may sometimes be accompanied by blood eosinophilia<sup>[7]</sup>.

As far as we know, there is no previous report of IHS manifesting with HTL, constituted exclusively by mature eosinophils. So, here we describe a case of IHS presenting imaging studies suggestive of a neoplasm, but secondary to the accumulation of mature eosinophils in a tumoral fashion.

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