Association of sinus histiocytosis with massive lymphadenopathy and idiopathic hypereosinophilic syndrome

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Summary. A patient was evaluated because of edema, pruritus and generalized painless lymphadenopathy. Laboratory tests showed marked eosinophilia without known etiology. CT scan of abdomen revealed multiple lymph nodes in retroperitoneal area. Lymph node biopsy was reported as sinus histiocytosis, bone marrow biopsy showed hipercellularity with marked infiltration of normal eosinophils. During his admission he developed Coombs positive hemolytic anaemia. Once he was stable, a laparotomy was performed and the patient died two days later because of septic shock. Autopsy revealed sinus histiocytosis with massive lymphadenopathy (SHML) with extranodal involvement of duodenum, spleen and prostate; septic liver and spleen, pyelonephritis, marked infiltration of eosinophils in lymph nodes, spleen, liver duodenum and lungs. To the best of our knowledge, this is the first case report of the association of SHML and idiopathic hypereosinophilic syndrome (HES).

Materials and methods

A fifty-eight year old black male patient was admitted to the Vargas Hospital (Caracas) in February 1985 because of a 6-month history of edema in the legs and generalized pruritus. There was no other significant history, except that he had urinary tract infection 9 months before and there was also evidence of marked eosinophilia since 4 months ago. On admission the patient appeared pale with generalized painless lymphadenopathy and edema in scrotum and legs. Spleen and liver were normal.

Results

Laboratory tests showed: hemoglobin 9.32 gr/dl, leukocyte count 31600 mm³, eosinophils 82%, neutrophils 6%, lymphocytes 12%, platelets 289000 mm³, LDH 389 u/L (normal, 55-138). Several fecal examinations were free of occult blood or ova. Cultures from blood, urine, lymph node and spinal fluid were negative for bacterias, mycobacterias and fungi. Chest X-ray, ECG and Echocardiogram were normal. CT scan of abdomen revealed multiple lymph nodes in the retroperitoneal area. A first inguinal node biopsy was reported as unspecific adenitis, a second axillary node biopsy showed sinus histiocytosis. (Figs. 1, 2) bone marrow biopsy was reported as hypercellular with marked infiltration of normal eosinophils. Citogenetic studies from peripheral blood and bone marrow were normal.

The patient developed fever without evidence of infection 14 days after admission. In April, he presented crisis of Coombs positive hemolytic anaemia with a hemoglobin level of 3 gr/dL; then he was treated with blood transfusions and prednisone, 50 mg/day. Once the patient was stable, a diagnostic laparotomy was performed but he died 2 days later because of septic shock.

Key words: Sinus histiocytosis - Hypereosinophilic syndrome
Sinus histiocytosis and hypereosinophilic syndrome

Fig. 1. Axillary lymph node biopsy with histiocytes showing some phagocytic activity and hemosiderin in the cytoplasm (arrow). H. E. ×400

Fig. 2. Axillary lymph node biopsy with infiltration of eosinophils (arrows). H. E. ×200
The autopsy revealed SHML (Fig. 3) with extranodal involvement of duodenum (Fig. 4), spleen and prostate, septic liver and spleen, pyelonephritis. There was also marked infiltration of eosinophils in lymph nodes, spleen, liver duodenum and lungs.

**Discussion**

Empiric threefold criteria have been established for idiopathic HES: persistent eosinophilia of 1500 eosinophils/mm³ for at least 6 months or death before 6 months with signs and symptoms of HES disease; lack of evidence for parasitic, allergic or other recognized causes of eosinophilia despite careful evaluation; and signs and symptoms of organ involvement or dysfunction either directly related to eosinophilia or unexplained in the given clinical setting (Chusid et al., 1975 Goh et al., 1985).

Our case represents a typical case of idiopathic HES with persistent eosinophilia without known etiology. Clinical features of edema, pruritus and fever are non-specific but they can be produced by HES (Harley, 1982) conversely generalized lymphadenopathy is uncommon in HES, but it is a characteristic finding of SHML. The diagnosis of SHML was firmly established in the patient not only by lymph node biopsy but also during autopsy. Coombs positive hemolytic anaemia has been associated with several cases of SHML as evidence of the spectrum
of immunologic abnormalities, but no association has been described with HES (Foucar et al., 1984).

Finally, the patient died because of septic shock which was probably influenced not only by steroids but also by the lymphomas because they have been associated with several abnormalities of immune function (Harley, 1982; Foucar et al., 1984).

In conclusion, we report as unusual patient with idiopathic HES and SHML with Coombs positive hemolytic anemia. We believe that this is the first case with this association in the literature.

References


Accepted February 9, 1987