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Cold Urticaria: An Unusual Manifestation of Histiocytic Lymphoma

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Cutaneous manifestations of internal malignancy and lymphoma are rarely encountered [1] and the association of reactive vascular phenomena, such as erythema nodosum and urticaria, is an infrequent occurrence [2]. Recently, we encountered a patient with cold urticaria as a manifestation of histiocytic lymphoma. This association is extremely rare and worthy of report. Report of a Case

A 71-year-old female was referred to the hospital because of cold urticaria and purpura of 2 months' duration. Physical examination and laboratory data revealed no abnormalities. She was told to avoid exposure to cold and was kept under close clinical observation. 2 years later she was admitted to the hospital because of pyrexia of unknown origin. Physical examination revealed a body temperature of 38 °C. A firm round mass (6 cm in diameter) was palpable in the left upper quadrant of the abdomen. There was no lymphadenopathy. Provocative exposure of the skin to ice resulted in localized urticaria. Laboratory studies showed: ESR, 35 mm in the first hour (Wester-gren); hemoglobin, 11.2 g/dl; WBC count, 3,200/mm³ with 50% > lymphocytes. The thrombocyte count was 226,000/mm³ and the coagulation status was normal. Serological tests to detect antinuclear and rheumatoid factors, cold agglutinin and cryoglobulin were all negative. Serum complement levels and the Coombs' test were negative. There was no monoclonal gammopathy. A barium meal study showed posterior pressure on the lesser curvature of the stomach by an abdominal mass and ultrasound examination disclosed a round mass adjacent to the left lobe of the liver. Intravenous pyelography was normal. An explorative laparotomy was performed and a round mass (7 cm in diameter) adjacent to the liver was excised. The liver and the spleen were normal. Histologically, the mass was diagnosed as a histiocytic lymphoma. The patient was treated by abdominal irradiation followed by combination chemotherapy. Her response was initially favorable, but since then she has developed recurrent lymphoma in the mediastinum.

Comment

The association between cold urticaria and lymphoproliferative disorders is infrequent. Hauptmann et al. [3] first described such a patient who had monoclonal cryoglobulin, complement abnormalities, cold urticaria and lymphosarcoma. The complement profile was similar to that seen in patients with C1q Inhibitor (INH) deficiency. A review of the literature revealed 10 cases of C1q INH deficiency in association with lymphoproliferative disorders [3-5]. 6 of these patients had lymphocytic lymphoma, 3 had CLL and 1 had an immunoproliferative

disorder. 5 of the above patients had angioedema while only one showed cold urticaria. The sera from 6 of these patients had a monoclonal IgM.

The patient described in this report had no

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paraprotein or cryoglobulin and displayed symptoms of cold urticaria for a period of 2 years prior to the diagnosis of lymphoma. In the light of this experience, we suggest that lymphoma be excluded in all patients with unexplained cold urticaria of chronic duration.

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