

Lymphography in Malignant Histiocytosis: Case Report

R. Musumeci*, R.A. Castellino**

*Clinical Assistant, Department of Radiology, Istituto Nazionale Tumori, Milano; Professor of Radiology, University of Milano

**John S. Guggenheim Fellow 1974-1975; Associate Professor of Radiology, Stanford University Medical School, Stanford, California; Visiting Professor of Radiology, Istituto Nazionale Tumori, Milano

Summary

Malignant histiocytosis is an uncommon, progressive disease with a poor prognosis which is characterized by malaise, fever, lymphadenopathy, and splenomegaly. Lymphadenopathy is commonly present at presentation as well as during the course of the disease. The lymphographic findings in this case report were very similar to those encountered in malignant lymphoma, a not unexpected situation since the clinical and pathological resemblances to histiocytic lymphoma are striking.

Malignant histiocytosis is an uncommon disease which was described by *Scott and Robb-Smith* in 1939 as histiocytic medullary reticulosis (6). Since that time other cases have been added to the literature variously called aleukemic reticulosis, histiocytic reticulosis, histiocytic leukemia, among others (1, 2). The term of malignant histiocytosis was introduced for this disease in 1966 by *Rappaport* (5), and should not be confused with the entity known as histiocytosis-X. A comprehensive recent review of 29 cases by *Warnke, Kim and Dorfman* (7) details the clinical and pathologic features of this disease.

Malignant histiocytosis affects all ages with no apparent predilection for any age group, although there may be a male predominance. Usual presenting manifestations include weakness, malaise, fever, lymphadenopathy, and hepato- and splenomegaly. Anemia, leukopenia, and thrombocytopenia are not infrequent findings. Radiographs have shown evidence of mediastinal-hilar lymphadenopathy and scattered lytic bone lesions in some patients. The histologic findings of involved lymph nodes show an invasive proliferation of atypical histiocytes and their precursors, with distortion of the normal nodal architecture, with partial maintenance of a sinusoidal pattern. Prognosis has been abysmally poor, with a median survival of six months. The etiology is unknown.

In the 29 patients studied by *Warnke et al*, 21 presented with lymphadenopathy, of which 4 were generalized, and 17 were localized, in distribution (7). It is therefore not unreasonable to assume that in some cases intra-abdominal and retroperitoneal lymph nodes may be similarly involved - and that lymphography would be capable of demonstrating such abnormalities in some cases, as noted in the following report.

Case Report

This 13 year-old white male noted an enlarging mass in the left groin and discomfort in the region of the right maxillary sinus and upper jaw. He denied symptoms of fatigue, weight loss and fever. Physical examination revealed normal vital signs and palpable, non-tender lymphadenopathy in the left groin without involvement of other lymph node groups. A firm mass was present involving the right upper gingiva and the right tonsil was enlarged and hard. Examination of the thorax and abdomen was unremarkable, without evidence of hepatosplenomegaly.

Laboratory tests were normal, including routine urinalysis, complete blood count, liver function tests and serum electrophoresis. Radiographs of the chest and axial skeleton, and an upper gastrointestinal series, were normal. Appropriate radiographic studies demonstrated lytic destructive

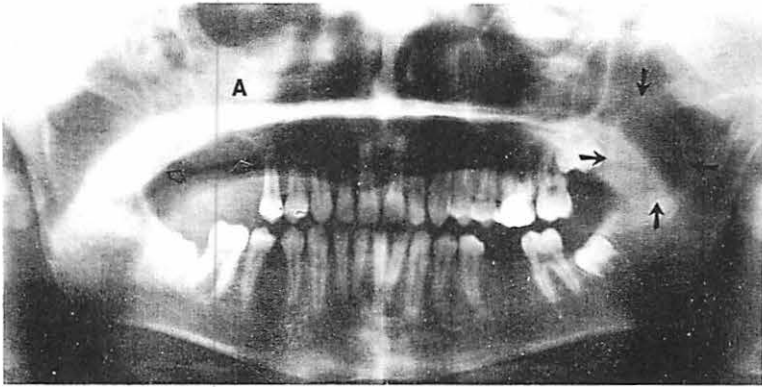


Fig. 1a Pantomogram demonstrates an opaque right maxillary antrum (A) with associated bone destruction and a soft tissue mass (open arrows) at its inferior margin. An expansile lytic lesion is present in the left mandibular coronoid process (solid arrows).

bone lesions involving the right maxilla and mandible. (Fig. 1a and b). A bipedal lymphogram revealed markedly abnormal appearing paraaortic and iliac lymph nodes which were enlarged. The internal architecture was distorted with prominent areas of non-opacification producing a "foamy" appearance (Fig. 2) such as usually identified with malignant lymphoma.

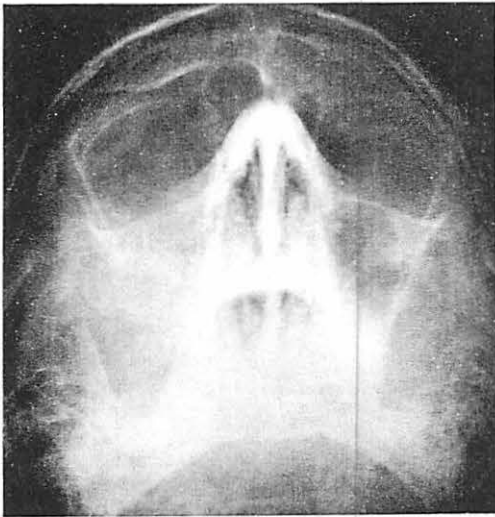


Fig. 1b Water's projection demonstrates the opaque right maxillary antrum with bony destruction of the inferior-lateral wall.



Fig. 2 Left posterior oblique the day following bipedal lymphography shows multiple enlarged and foamy opacified lymph nodes. (Appendix contains barium from prior UGI.).

Biopsies of the right gingival mass, right tonsil, and a left inguinal lymph node were interpreted as representing malignant histiocytosis. Percutaneous biopsies of bone marrow, liver, and spleen were all normal. The liver and spleen were also unremarkable at laparoscopy. He was treated with multiple drug chemotherapy (Adriamycine, vincristine, prednisone) with an initially favorable clinical response, as well as a decrease in size of the residually opacified lymph nodes. However, the disease soon recurred and progressed. He died from widespread disease with bony metastasis, bone marrow and kidney involvement, and pleural effusions 11 months following the onset of disease. No autopsy was performed. —

Discussion

The major entities in the histopathological differential diagnosis of lymph nodes containing malignant histiocytosis are histiocytic lymphoma, histiocytosis-X (especially Letterer-Siwe disease) Hodgkin's disease, sinus histiocytosis with massive lymphadenopathy, metastatic neoplasm, viral lymphadenopathy, and hairy cell leukemia (7). When these pathologic processes affect lymph nodes to such an extent as to produce gross alterations in their internal architecture, then lymphography should be able to demonstrate abnormal appearing opacified lymph nodes. Not surprisingly, all of these entities have been responsible for producing clearly abnormal lymphograms.

The clinical and radiographic presentation of the patient discussed in this report was very suggestive of lymphoma. A recent review of newly diagnosed children with non-Hodgkin's lymphoma showed a high incidence of radiographically detectable bone involvement (18%) at presentation, particularly involving the facial bones; and positive lymphograms (30%) which generally showed the characteristic findings of multiple enlarged and foamy lymph nodes (3). The lymphographic findings in this patient, although very similar to those seen in lymphoma, are non-specific and are encountered in a variety of other malignant diseases as well as in benign entities (4).

Lymphography may have a role in the evaluation of patients with malignant histiocytosis by more adequately defining the extent of disease, and by providing information regarding the biologic behavior of this entity. Monitoring the residually opacified lymph nodes by periodic surveillance abdominal films may provide an objective measure of therapeutic response and detection of disease relapse.

References

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R.A. Castellino, M.D., Department of Radiology, Stanford Medical School, Stanford, California 94305

Addendum: „Radiographic Manifestations of Malignant Histiocytosis“ by Dunnick et. al. (Amer J. Roentgenol, in press) reviews 26 cases, 8 of whom had lymphograms.