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Lymphography in the Assessment of Mycosis Fungoides

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Summary

Extracutaneous manifestations of mycosis fungoides imply a bad prognosis and are a major cause of death. Benign dermatopathic lymphadenopathy is associated with mycosis fungoides and often precedes lymphomatous infiltration. In this study, 10 patients in the early stages of mycosis fungoides underwent clinical and lymphographical examinations. In one the lymphoma was already present in lymph nodes. Six had signs of dermatopathic adenopathy which was verified by lymph node biopsy in 5. In one of these the disease later progressed to a malignant lymphoma. The frequent occurrence of lymph node involvement justifies the use of lymphography collaterally with staging laparotomy to determine the presence of pathologic retroperitoneal lymph nodes.

Mycosis fungoides (MF) is an uncommon malignant lymphoma originating in the skin. The clinical course is characterized by a slow progression from an erythematous lesion to infiltrative plaques and eventually to cutaneous tumours. The disease may eventually progress to involve lymph nodes as well as other internal organs. An autopsy often reveals widespread visceral lymphomatous infiltrate, regardless of whether or not this was detected before death (2).

The usefulness of careful staging by lymphography alone or combined with laparotomy to assess the extent of Hodgkin's disease and of other malignant lymphomas is well established (1, 6, 7, 8, 9). These staging procedures have contributed to our understanding of the prognosis of these diseases and the effects of therapy. With this in mind, we have used lymphography to investigate the state of the lymph nodes in early MF.

Material and Methods

Ten patients with histologically confirmed MF underwent lymphographic examinations

(Table 1). In 8 patients lymphography was performed at the time of diagnosis, and in 2 (cases 1 and 5), 9 months after the diagnosis. Lymphography was repeated 3 years later in one patient. The state of the lymph nodes was followed up through surveillance pictures in 4 patients.

The clinical skin manifestations of MF are usually classified into 3 stages: premycotic, infiltrative and tumour. At the time of lymphography 7 were in the premycotic stage and 3 in the infiltrative stage.

The lymphograms were interpreted by two radiologists with no knowledge of the clinical data. A classification of the lymphographic findings was fixed before interpretation. Class I (normal) nodes were small with even outlines and a uniform nodal structure. Class II nodes were slightly enlarged, the marginal sinus preserved and the nodal pattern coarsely granular. Class III was reserved for the alterations typical of lymphoma, i.e. greatly enlarged nodes, defective outlines and distorted, foamy, or reticular structure.

Seven patients were subjected to lymph node biopsy: 13 biopsies were taken before and 8 after lymphography.

Results

The results of lymphography and biopsy of the 10 patients are summarized in Table 1. Normal retroperitoneal lymphograms were seen in 4 patients, 3 of whom had premycotic and one infiltrative skin lesions. The physical examination had revealed no pathological inguinal lymph nodes. In one patient this was confirmed histologically both before and after lymphography. On the other hand, one of the patients had palpable lymph nodes in the axilla, shown histologically to be dermato-

Table I	Clinical Data	Lymphography and	Lymph Node Histology

Case	Age/ Sex	Duration of symptoms before diagnosis (months)	Stage of skin involvement (clinical estimat	Palpable lymph e) nodes	Lympho- graphy	Lymph node histology
1	56/F	25	premycotic	_	II	_
2	50/M	18	infiltrative	+	III	MF
3	82/M	3	infiltrative	-	I	-
4	28/M	68	premycotic	-	II	DL
5	49/M	20	premycotic	-	I	-
6	75/M	83	premycotic	+	I	DL
7	73/M	17	premycotic	+	II	DL
8	55/M	27	premycotic	- T-	I	N
9	63/M	96	infiltrative	_	H	DL
10	60/M	36	premycotic	-	II	DL

Lymphographic finding: I =

I = normal

II = non-specific nodal hyperplasi

III = lymphoma

Lymph node histology:

MF= mycosis fungoides lymphoma DL= dermatopathic lymphadenopathy

N = normal

- = no biopsy performed

pathic lymphadenopathy, in which capsules of the large nodes were preserved while the lymphoid tissue was replaced by a massive proliferation of histiocytes, whose cytoplasm contained pigments, melanin and hemosiderin.

In 5 patients the lymphogram was abnormal, displaying moderate node enlargement and coarse granulation which put them into Class II (Fig. 1). In one patient only adenopathy had become clinically apparent before lymphography. As the lymphographic changes were non-specific and our experience of lymphography in MF poor, a biopsy was performed in 4 cases to establish the histological changes. Dermatopathic adenopathy was found in all 4 cases. Of these 5 patients only one was in the infiltrative stage, the other 4 being premycotic.

In the 10th patient, the cutaneous lesions had rapidly progressed to the infiltrative stage. Lymph node involvement was present clinically. Isotope scanning revealed hepatosplenomegaly. The lymphographic findings conformed with the clinical picture of generalized malignant lymphoma (Fig. 2). A lymph node biopsy revealed MF lymphoma.

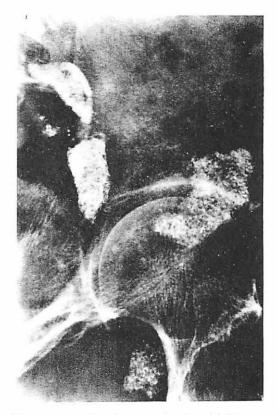


Fig. 1: Case 7. Slightly enlarged nodes with intact outlines and a coarse granulation. The microscopic diagnosis was dermatopathic lymphadenopathy.



Fig. 2: Case 2. Oblique view the day after lymphography shows multiple enlarged lymp nodes with foamy and irregular structure typical of a malignant lymphoma.

Discussion

The presence of skin tumours or palpably enlarged lymph nodes in patients with MF is of significant prognostic value (2, 5). Fuks et al. (5) observed that the eventual development of internal organ involvement was much more frequent in patients with initial lymphadenopathy, even if it mostly represented dermatopathic lymphadenitis.

In 1974 Fuks et al. (4) described 28 patients examined by lymphography, 9 had a lymphoma pattern and 6 a reactive hyperplasia. Of these 6, two subsequently developed histologically confirmed lymphoma. Escovitz et al. (3) observed a negative correlation between lymphographic assessment and clinical staging in 10 patients. These had a suspect or lymphomatous lymphographic pattern but no clinical adenopathy.

In the present 10 patients, 5 had similar abnormalities on lymphography, 4 of which were established as dermatopathic lymphadenopathy by biopsy. On physical examinations only one of the 5 had palpable nodes. This discrepancy could be due to the moderate enlargement of the nodes as well as to their soft consistency. The lymphographic finding, characterized as reactive hyperplasia, shows that whether dermatopathic lymphadenitis represents precursor abnormalities or the disease itself, the involvement may often be more widespread than can be detected clinically.

An analysis of the clinical and lymphographic findings gave no positive correlation between the localization of the skin symptoms and lymph node involvement. On the other hand, the lymphographic changes due to dermatopathic lymphadenopathy were in all cases most obvious in the inguinal and parailiac areas. The same centrifugal distribution of pathologic nodes was reported by *Escovitz* et al. (3).

Surprisingly 5 of the 6 patients with dermatopathic lymphadenopathy were in the premycotic stage at the time of lymphography and biopsy. One of these later progressed to the infiltrative stage, as the inguinal lymph nodes grew. A relymphography revealed that the node appearances had become typical of a malignant lymphoma (Figs. 3a and 3b). This suggests, as *Fuks* et al. (5) also assumed, that dermatopathic lymphadenitis and subsequent lymphoma involvement are circumstantially, and perhaps causally, associated.

Based on these results, it seems reasonable to put lymphography forward as a complementary staging procedure in patients in whom no laparotomy is performed. The detection of clinically unsuspected lymph node involvement is important in the choice of therapy and for ultimate outcome. Further studies with meticulous follow-up by lymphograms and biopsies may possibly provide valuable information about the mode of transformation of dermatopathic adenopathy into a lymphoma, as well as about the therapeutic response in lymph nodes.



Fig. 3a: Case 4. The lymph nodes are enlarged with a loosened structure corresponding to dermatopathic lymphadenopathy.

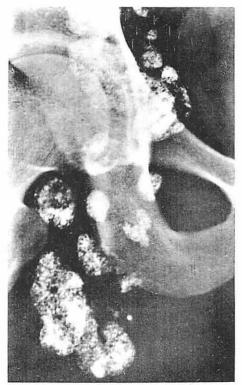


Fig. 3b: Case 4 contd. A relymphography 3 years later shows further enlargement of the nodes. In most of them there are central and peripheral defects beside the loosening of the structure. The finding indicates a malignant lymphoma.

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