CASE REPORTS

Mycosis Fungoides—Gastric Involvement with Massive GI Bleed

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Abstract

Mycosis fungoides is a primary malignant lymphoma of the skin, known to involve extra-cutaneous sites such as lymph nodes and viscera. Gastrointestinal tract involvement is uncommon. We describe the first autopsy report from India of a case with mycosis fungoides involving the stomach and complicated by ulceration and massive GI bleed resulting in death.

Key words: Mycosis fungoides, malignant gastric ulceration, GI bleed, gastric lymphomas.

Introduction

Mycosis fungoides (MF) characteristically begins as a primary cutaneous lymphoma which in course of time is known to involve regional lymph nodes and the viscera. Extra-cutaneous dissemination heralds a worse prognosis. Of the viscera involved, the gastrointestinal tract appears to be a less favoured site. Cases involving the stomach have been described in the literature, although massive GI bleed from these is not often seen.

We describe a case of mycosis fungoides in a young female. The disease was rapidly progressive and, at the time of death, the patient had lymph node involvement. Autopsy revealed malignant gastric ulceration with massive GI bleed.

Case Report

A 20 year old female was hospitalised with necrotic infected ulcers on the sides of the buttocks 10 months prior to admission. This was diagnosed as borderline lepromatous leprosy and she was treated with rifampicin, clofazimine and diocaine. A month before admission to hospital she developed increased redness of the old lesions and new lesions appeared. After a few days, the tumorous lesions ulcerated. Examination revealed two ulcers of 3 cm diameter each on the left buttck. The lesions had well defined, sloping margins, a clean floor and indurated base. There were a few small erythematous lesions in the vicinity of these ulcers. There was another deep ulcer on the left leg and a smaller ulcer on the right leg.

Investigations: Hemoglobin 6.4—5.0 g/dl, TLC 8000/c mm and ESR 40 mm/hr. Bone marrow revealed moderate hypertrabeculation but no malignant cells. The patient had normal serum bilirubin, SGOT/SGPT, creatinine, urea and alkaline phosphatase. Urine examination was normal; LE cell phenomenon and ANF were negative; VDRL test was non-reactive; chest X-ray was normal and ECG showed tachycardia.

The patient developed diarrhoea two days before death and terminally had upper GI bleed, paralytic ileus and altered sensorium.

Autopsy revealed enlargement of the mesentric and a few paraaortic lymph nodes. The stomach showed multiple ulcerated tumour nodules 1 cm-5 cm in diameter (Fig. 1) with blood encrusted at the floor. The gastric and intestinal contents were bloody. Histology revealed typical features of MF in the skin, lymph nodes and stomach (Fig. 2). Mycosis cells were seen in the gastric infiltrate. No other organs showed involvement by the tumour.

The cellular infiltrates in the skin, lymph nodes and stomach showed an absence of cytoplasmic immunoglobulins and no evidence of histiocytic differentiation (negative lysozyme and α-1-antitrypsin).

Fig. 1: Ulcerated tumour nodules and discoloured mucosa in the stomach.

Fig. 2: Microphotograph showing a pleomorphic infiltrate containing mycosis cells (W.D.E N 230).
Discussion
This patient had mycosis fungoides by both clinical and histological criteria. The presence of typical mycosis cells in the visceral infiltrate is an essential histologic criterion for the diagnosis of MF. In addition to the finding of mycosis cells in the infiltrate, the absence of cytoplasmic immunoglobulins, lysozyme and alpha-1-antitrypsin from the malignant lymphoma cells strongly supports GI involvement by mycosis fungoides.

The points of interest in our case are the relatively young age of the patient and a quick progression of the disease resulting in death within a year of the appearance of the first lesions. In a series of 144 patients, most patients developed the first lesions at age 40-60 years. The average time interval between the first lesions and diagnosis was 6.1 years (0.1-48.3 years).

Gastric involvement in MF is uncommon in some of these cases it could be the cause of significant morbidity or mortality. Massive GI hemorrhage from ulcerated tumor node(s) in the stomach appears to be a potential cause of death. In addition to hemorrhage, other GI complications seen in MF are small bowel perforation and malabsorption syndrome. In addition, gastrointestinal involvement in czerny syndrome has been noted.

To the best of our knowledge, this is the first autopsy report of a case of mycosis fungoides from India. It also is the first report from India and fifth in the English literature where massive upper GI hemorrhage from metastatic gastric nodules in a case of mycosis fungoides resulted in death.

References