Primary Follicular Lymphoma of Mesentery in an Older Man - A Case Report

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A man aged 59 years, diagnosed as a case of cholelithiasis 11 months back, was admitted at the surgery unit of Enam Medical Collage & Hospital on 11th December 2012 with a large abdominal painless mass. On examination, a huge mass was palpable occupying the epigastric and left hypochondriac regions at empty stomach. CT scan of abdomen revealed a large soft tissue mass (10.1x7.5x5.7 cm) in the left mid abdomen which crosses midline involving mesentery and the surrounding area of superior mesenteric artery. Another 8.2 x 4.0 cm lobulated retro-peritoneal soft tissue mass and a stone in the gallbladder lumen were also reported. Histopathology of the mesenteric mass and mesenteric lymphnode specimens diagnosed as Follicular non-Hodgkin lymphoma, grade 2 on the basis of H&E and reticulin stain. Later on immunohistochemistry confirmed the diagnosis.

Key words: Follicular lymphoma, Mesentery

Introduction

Follicular lymphoma (FL) is a neoplasm of follicle centre cells which has at least a partially follicular pattern.1 Also known as nodular lymphoma, it consists of a variable proportion of centroblasts and centrocytes. In USA it constitutes about 35% of adult non-Hodgkin lymphoma but the incidence is lower in Asian countries. It is predominantly a disease of the adults, mean age being 59 years and majorities are located in the head and neck region. In all age groups, FL mainly involves lymph nodes. Less frequently spleen, bone marrow, blood and Waldeyer’s ring are involved.1 The gastrointestinal tract is the most common site for extranodal lymphomas, but follicular lymphomas involving the mesentery are rare.2,3

Histologically FL are recognized by follicular pattern of tumour cell growth in which follicle formation may be greater than 75% (follicular pattern), less than 25% (minimally follicular pattern) or in between 25-75% (follicular and diffuse pattern). Two main types of cells found in FL are centrocytes or cleaved follicular centre cells and centroblasts or non-cleaved follicular centre cells. In 10% of cases marginal zone or monocytoid cells are observed at the neoplastic follicles.1 Follicular lymphomas are graded from 1 to 3 depending on the proportion of centroblasts. Histological grade correlates with clinical out come. Grades 1 and 2 are indolent and are not usually curable. Grade 3 is more aggressive but has potential of being cured by aggressive therapy.1

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The FLs resemble their lymphnode based counterparts in their tendency to express bcl-2. Enhanced expression of the bcl-2 oncogenic protein has been described in lymphoma cells in 75% of cases of intestinal FL. Small cleave or mixed cell lymphomas were more likely to show enhanced expression than were large cell cases. Immunohistochemical staining for enhanced bcl-2 expression is of potential diagnostic utility to distinguish between follicular lymphoma and follicular lymphoid hyperplasia in the gastrointestinal tract.2,4

Case Report
A 59 years old man, was admitted at the surgery unit of Enam Medical College Hospital on 11th December 2012 with a large painless abdominal mass and as a diagnosed case of cholelithiasis for operation. On examination, a huge mass was palpable which occupying epigastric and left hypochondriac regions at empty stomach. His appetite was more or less good and there was no significant weight loss during the last eleven months period of his illness. There was no history of haematemesis, melena, jaundice or features suggestive of gut-obstruction. On examination, the man was afebrile, anicteric, mildly pale and anxious. There were no sings of cyanosis, oedema, dehydration or clubbing. None of the superficial lymph nodes were visibly enlarged or palpable. Investigations revealed a normal blood count with a normal peripheral blood film. Blood urea and serum creatinine were within normal limit. After his hospital admission, CT scan of abdomen was done on November 2012 reported a large soft tissue mass (10.1x7.5x5.7 cm) in the left mid abdomen which cross midline involving mesentery and the surrounding area of superior mesenteric artery and multiple mesenteric enlarged lymphnodes were detected. A small biopsy from the soft tissue mass and resection of a lymphnode were taken. The gross examination revealed a 3x1.3x1 cm irregular piece of soft tissue and a lymphnode measuring 1.3 cm in diameter. The cut surface of the irregular piece was homogeneously gray white. Appropriate blocks of tissue were sampled for routine processing, paraffin impregnation and H &E stain. On microscopic examination, sections of the both specimen revealed a grade 2 follicular non-Hodgkin lymphoma made of a mixed population of centrocytes and centroblasts (Fig. 2). The tumour cells were arranged both in nodules (Fig. 1) and diffusely. Pattern of nodularity was obvious in H&E stained sections but for highlighting the reticulin fibres around nodules, silver stain (Gomori) was done (Fig. 3). One paraffin block was sent to a private laboratory for bcl-2 immunostain which yielded positive result. The patient was discharge from EMCH on 12th January 2013 and admitted to the Delta Medical College Hospital. There he got chemotherapy and had uneventful recovery and returned home after 15 days. Follow up of the case was not possible.

Fig 1. Photomicrograph showing even distribution of follicles throughout the cortex and medulla with fading of follicles (H & E x100).
Fig 2. Photomicrograph showing centrocytes and centroblast in mesenteric follicular lymphoma (H & E x 400).

Fig 3. Photomicrograph showing reticulin fibers encircling neoplastic follicles (Silver stain x 200).

Discussion
Follicular lymphoma is predominantly a disease of adults. The mean age is 59 years. Head and neck regions are mostly affected and 50% of the tumours are of grade 3 large cell type.\(^1\) The case presented above had features different from those described in literature. It was in a man of 59 years of age, the primary tumour was in the mesentery and mesenteric lymphnode and histologically it was grade 2. Primary FL of the mesentery and mesenteric lymphnodes are rare. Most of the cases are observed in context to widespread nodal disease.\(^1,5\)

Abdominal pain was main presenting symptom in mesenteric and mesenteric lymphnodes of FL and smaller group patients may come with intestinal obstruction.\(^4\) Amazingly, it may not produce any symptoms or other peripheral lymphadenopathy other than an abdominal lump and mesenteric lymphadenopathy.\(^6\) A moderate to severe degree of anemia is encountered in any cases. The average duration of symptoms has been described 4 to 5 months.\(^7\)

In this case histological diagnosis of follicular non- Hodgkin lymphoma was only made after histological examination of the sections. Distinctive nodule formation was observed in about 50% areas of tumour. The other areas revealed diffuse pattern. The tumour was made of about equal number of centrocytes and centroblasts (Fig. 2) and thus represented WHO grade 2. Reticulin stained sections revealed nodules of tumour cells surrounded by reticulin fiber (Fig.3). Majority of the tumour cells were bcl-2 positive determined by immunohistochemical stain. Bcl-2 is considered to be a specific marker of FL\(^1,2,4,5\)

The morphological features of centroblasts and centrocytes in FL are analogous to that of cells in the normal germinal centre B cells. Analysis of the DNA shows not only clonalgene rearrangement but also evidence of ongoing somatic mutation and antigen affinity selection characteristic of germinal centres. However, contrast to the benign follicle, the cells of most cases of follicular lymphoma are bcl-2 positive. Because 85-90% of cases of follicular lymphoma have the t (14; 18) translocation, it has been assumed that this translocation causes excess stimulation of the bcl-2 gene and is therefore
responsible for the bcl-2 positivity of the neoplastic follicles. The bcl-2 protooncogene was discovered at the chromosomal breakpoint of the t (14; 18) translocation found in human follicular lymphoma; t (14; 18) juxtaposes the bcl-2 gene from chromosome 18 with the immunoglobulin heavy chain (IGH) locus on chromosome 14. This creates a bcl-2 IGH fusion gene that is markedly deregulated, resulting in the over production of bcl-2 RNA and protein. Bcl-2 has the oncogenic function of blocking programmed cell death.8

There is no unified treatment strategy for the mesenteric and mesenteric lymphnodes FL. In the 25 cases describe by Damaj (2003), seven patients did not receive any treatment; four of whom progressed after a median follow-up of 37.5 months. Treatment in rest of the patients was heterogeneous which included surgical resection and chemotherapy. Complete remission was obtained in 15 patients who lasted for a median of 31 months. Relapses were either in the GI tract (n=3) or outside the GI tract (n=3). After a median follow-up of 34 months (range 5-23), 22 patients were still alive (complete remission, 11; partial remission, three; stable disease, six; progressive disease, two.4

References