Case Report

Peripheral T Cell Lymphoma of Stomach with Scattered Reed Sternberg Resembling Cells

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Introduction

A case of Peripheral T Cell Lymphomas (PTCL) of stomach presenting as a primary gastric neoplasm is presented. The morphology of this rare primary gastric malignancy is very characteristic and the response to the treatment is excellent. As the tumor contains Reed Sternberg resembling cells, it must be distinguished from primary gastric Hodgkin's disease, which is much rarer.

Summary

A case of primary Peripheral T Cell Lymphoma of the stomach has been described. The disease presented as an exophytic infiltrating gastric tumour. Microscopically many complicated multilobed nuclei containing giant cells were present. This feature suggested that the present case belonged to a generic, heterogeneous group of lymphomas designated as "Peripheral T Cell Lymphoma". The characteristic features of this group of lymphomas include extranodal presentation and excellent response to chemotherapy despite ominous looking microscopic features as seen in the present case.

Case Report

A 43-year old Syrian male was admitted with history of upper abdominal pain and nausea for last 8 months, loss of weight and bleeding per rectum for last 4 months. The pain was localized in epigastrium, dull in character and was aggravated by eating. There was no history of vomiting or haematemesis. He was having repeated episodes of malaena (dark stools) since last 4 months and these had increased during last one month. He lost 16-kg body weight over last four months. Past, family, social and personal history was non-contributory.

Physical examination revealed young well built male who showed signs of emaciation. He was pale, peripheral lymph nodes were not palpable and his vital signs were normal. Examination of the abdomen showed an ill-defined mass in the epigastrium. It was 10x3 cm in size, was mildly tender, had irregular surface and was firm in consistency. Liver and spleen were not palpable and there was no ascites.

Laboratory investigations revealed the following; CBC-Haematocrit-27.4%, WBC 5.8x10^3/cmm, Haemoglobin-9g /dl, peripheral blood smear and absolute indices were suggestive of microcytic hypochromic anaemia. Total serum protein was 5.4 g/dl with serum albumin of 2.7 g/dl. Liver enzymes and other relevant serum chemistry tests were normal.

Upper G.I. endoscopy showed a fungating mass at pre-pyloric area with extension to mucosal folds and narrowing of lumen of stomach. Multiple biopsies were taken for histopathological examination. These revealed poorly differentiated malignancy. The C.T. scan of the abdomen revealed that gastric pylorus was thickened with polypoidal projections from its wall (Fig. 1). An oval small soft tissue mass was seen just underneath the anterior abdominal wall close to the region of pylorus (? enlarged lymph node). Pancreas, spleen, liver, kidneys and adrenals were normal and there was no para-
Diagnosis of gastric malignancy was made and patient was posted for surgery. The intraoperative findings revealed a large gastric tumour in the antrum infiltrating up to serosa of the stomach with an enlarged lymph node at the greater curvature just proximal to pylorus, and a prominent pancreaticoduodenal lymph node. Liver was normal and there was no ascites. Partial gastrectomy with end to side retro-colic gastro-jejunostomy was done. Pancreaticoduodenal lymph node was excised.

Pathological Description

**Gross appearance**
Gross examination of partially resected 12x7x4-cm stomach revealed an exophytic gastric 5x4-cm tumor. The tumor was infiltrating up to the serosa. The cut surface was fish flesh gray-white. Several peri-gastric and pancreaticoduodenal lymph nodes which were submitted alongwith also showed fish flesh homogenous appearance.

**Microscopic appearance:** The gastric mucosa overlying the tumor showed focal ulceration. The resection margins were free of the tumor. Underneath the tumor was transmural extending up to the serosal layer. The tumour was comprised of peculiar large cell with complex multilobated nuclei. These multi-lobated nuclei frequently assumed mulberry or clover leaf shapes or appeared split into fragments due to marked lobulation (Figure 2). The immunohistochemical stains revealed negativity for keratin. The Lymph nodes showed large diffuse replacement with the lymphoma with occasional scanty uninvolved areas.

**Post-operative course.** The postoperative course was uneventful. Subsequently bone marrow aspiration and biopsy were done which did not show evidence of lymphomatous infiltration. Postoperatively, chemotherapy was started and he received 6 cycles of MOPP and ABVP in 6 months.

**Discussion**
Malignant lymphomas of the gastrointestinal tract may be classified into primary or secondary. Primary malignant lymphoma of the gastrointestinal
tract is defined as a disorderly clonal proliferation of lymphoecticular cells originating in the gastrointestinal tract. Dawson et al proposed the following criteria for the diagnosis of primary gastrointestinal lymphomas: (1) absence of peripheral lymphadenopathy at the time of presentation, (2) lack of enlarged mediastinal lymph nodes on chest x-rays, (3) a normal total white cell and differential count, (4) the bowel lesion predominates at the time of laparotomy, and the only lymph nodes obviously affected are those in its immediate neighborhood, (5) the liver and spleen do not show lymphomatous involvement. The present case fulfills all the above criteria. It should be noted that most studies however accept those patients who initially present with gastrointestinal symptoms as having primary gastrointestinal lymphomas. This was also the case here. Extranodal Hodgkin’s lymphoma (HD) are distinctly uncommon. The most common location for extra nodal HD is liver and lung. Devaney and Jafe in their review put the incidence of gastric HD at less than 1% of all cases. About 2.5 of the primary malignancies of stomach represent lymphomas and HD represented 1.7% of these lymphomas.

The term HD has been traditionally used for a type of malignant lymphomas in which Reed-Sternberg (R-S) cells are present in a background of reactive inflammatory cells of various types i.e., mature lymphocytes, eosinophils, plasma cells and histiocytes.

HD is a heterogeneous group of lymphomas. This heterogeneity is not only manifested by their variable histology but also by their different cells of origin. The spectrum of HD merges on one hand with atypical reactive hyperplastic processes and on the other hand with non-Hodgkin’s lymphoma. In view of this heterogeneity, it is not unexpected to find occasionally composite lymphomas i.e., the lymphomas containing the features of both Hodgkin’s and non-Hodgkin’s lymphoma. It is interesting to see a case of composite lymphoma (Hodgkin’s disease together with non-diffuse large cell lymphoma) in the series of Devaney and Jafe. This reinforces the view that the boundary between Hodgkin’s disease and non-Hodgkin’s lymphoma occasionally may not be clear-cut and one neoplasm may merge into other or contain features of both. The presence of classical R-S cells with large bi-lobed mirror image nuclei; ample cytoplasm and prominent eosinophilic nucleoli

**Fig. 3:** Multilobed nuclei, a characteristic feature of peripheral T cell lymphomas, present in great numbers. (H & E x400)

**Fig. 4:** Three complicated multilobed nuclei in one field (oil immersion, (H&Ex1000)
are mandatory for initial diagnosis of HD. However at times one can see in addition other large cells which are regarded as R-S variants such as mono nuclear variants, lacunar cells, mummified cells and pop corn cells. The literature confirms different sources of origin in different cases of HD; B-lymphocytes, T lymphocytes and even macrophages (Pinkus GS et al, 1979).

The present case presented with polymorphous cell infiltrate with some cells resembling R-S cells. The lymphomatosus infiltrate in our case in addition to classical R-S cells and R-S variants also contained peculiar multi-lobed nuclei (Figures 3 & 4) hypersegmented and hyperlobed cells are classically seen in Sezary syndrome and Mycosis Fungoides. These polylobated cells have been established without any doubt as T cells. Pinkus et al have reported four cases of extra-nodal non-Hodgkin’s lymphoma with such cells. They have regarded this as ‘a distinct morphologic variant with large multilobed nuclei’. In their study these cells were positive for T cell markers. Rosai described such tumors as Peripheral T Cell Lymphoma (PTCL). He considered them a generic group that represents a family of neoplastic lymphocytes with phenotypic features of peripheral T Cells. It is considered to be a heterogeneous poorly understood, often interrelated and continuously growing group of lesions having morphologic links with several other types of lymphomas. These comprise mycosis fungoides-Sezary syndrome, lymphomatoid granulomatosis and some other types of lethal mid line granulom, T cell lymphocytic leukemia and prolymphocytic leukemia. T cell immunoblastic sarcoma, T cell lymphoma with multi lobulated nuclei, T zones lymphoma, erythrophagocytic T cell lymphoma, and plasmacytoid T cell lymphoma. Angioimmunoblastic lymphadenopathy like T cell lymphoma, pleomorphic T cell lymphoma, Ki-lymphoma and Hodgkin’s disease, the T cell nature of which has been suggested by several observers. HTLV-1 virus has been found in T cell leukemia/lymphoma in ceratin endemic region. Recently cases of T cell lymphoma containing EB viral DNA has been reported.

These peripheral T cell lymphomas (PTCL) are characterized by; extranodal location, complicated lobated nuclei and good prognosis despite ominous looking histological appearance. He urged that some cases of Hodgkin’s lymphoma may also belong to this category. In the past this entity has been reported in skin, subcutaneous tissue, bone, gonads and the central nervous system but not in the stomach. The extra nodal location complicated multilobed nuclei and excellent response to chemotherapy all favor this interpretation.

In summary, the present case represents a primary gastric Peripheral T lymphoma with multilobed nuclei. It is important to recognize that PTCL may contain Reed Sternberg type cells. The response to chemotherapy is usually excellent and prognosis is good despite bizarre and ominous looking histology, transmural extension, extra nodal location and involvement of adjacent lymph nodes.

References