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Gastroenterology

A RARE CASE OF CONCURRENT PRIMARY GASTRIC AND BONE DIFFUSE LARGE B CELL LYMPHOMA.

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ABSTRACT Primary bone lymphoma (PBL) is an uncommon clinical entity and a rare non-Hodgkin's lymphoma presentation. PBL accounts for less than 5% of malignant bone tumours. Gastric lymphoma (PGL) is also rare cancer, but it is the most common site for extranodal non-Hodgkin lymphoma. We report a rare case of concurrent occurrence of bone lymphoma and gastric lymphoma. The patient presented with a slow-growing swelling around the knee joint for two years. Positron emission tomography incidentally picked up an FDG avid lesion in the fundus of the stomach. Tissue specimens of both the lesions were obtained and conformed to have diffuse large B cell lymphoma on histology and immunochemistry. He is on follow up with the oncology department and being treated with an R-CHOP regimen (rituximab, cyclophosphamide, hydroxydaunorubicin, oncovin (vincristine), and prednisone.

KEYWORDS : Gastric lymphoma, primary bone lymphoma, non-Hodgkin's lymphoma.

INTRODUCTION:

Extranodal lymphomas are principally non-Hodgkin lymphoma (NHL) type. Gastrointestinal (GI) lymphoma comprises less than 10% of gastric malignancies but represents the most common site of extranodal lymphoma. The most common site of involvement in the GI includes the stomach, followed by the small intestine and ileocecal region (1). Primary bone lymphoma (PBL) is also a rare malignancy and accounts for less than 5% of all extranodal non-Hodgkin lymphomas (NHL) (2) (3).

Case History:

A 37-year-old male came with the chief complaint of pain and swelling of the distal part of the right thigh for two years. The pain started two years back insidiously, dull aching in character, and gradually increasing in severity. Swelling also started at the same time and was gradually increasing in size. There was a noticeable increase in the pain and swelling for the past two months prompting the current admission. The swelling caused restriction of mobility and pain while walking, causing him to use a walking aid.

There is a history of loss of appetite and 8kgs weight loss over the past six months. There is no history of trauma, fever, fistulous opening, GI symptoms, smoking or alcohol use.

Physical examination revealed a hard swelling in the distal right thigh. The swelling is very tender, warm to the touch, causing joint mobility restriction. There is no regional lymphadenopathy. The rest of the general and systemic examination was normal.

Blood investigations showed haemoglobin (HB%) 9.7gm%, with a microcytic picture on peripheral smear. WBC count 7100 cell/mm3, platelet count 3 lakhs/mm3, and normal renal and liver function. Viral markers (HBsAg, HCV, and HIV) are negative. MRI knee showed an aggressive lesion with extensive marrow infiltration, extraosseous soft tissue components and intra articular extensions.

A primary bone tumour was suspected and planned for biopsy. Right distal femur open biopsy was performed and sent for histopathological examination. A Positron Emission Tomography -Computed Tomography (PET-CT) was done to check for metastasis, which revealed an FDG avid thickening involving the fundus of the stomach with locoregional lymphadenopathy in addition to FDG avid mass in the right distal femur (FIG 1).

We performed an Upper GI endoscopy to evaluate the gastric lesion. A large ulcero-proliferative growth of size 8cm x 4 cm noted along the greater curvature of the stomach with overlying sloughed off mucosa was noted (FIG 2) and multiple biopsies were taken.

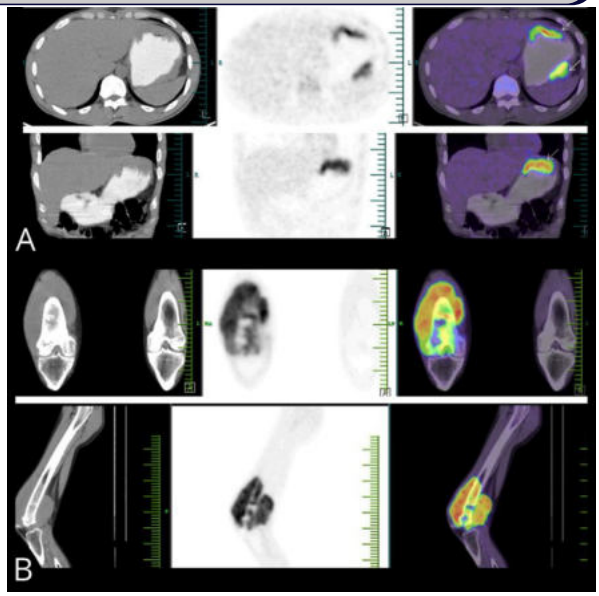

FIG 1 – Positron Emission Tomography - Computed Tomography (PET-CT) -
A - Fluorodeoxyglucose (FDG) avid wall thickening is seen involving the fundus of the stomach for a length of 7.0cm.
B- A large FDG avid soft tissue mass lesion is seen in distal 1/3rd of the right femur causing a permeative pattern of done destruction with cortical erosion of both patella and femur.

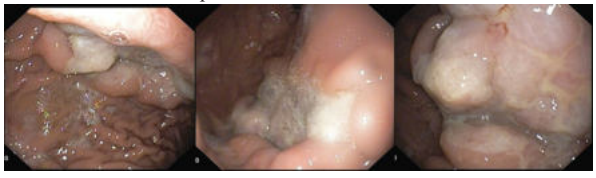

FIG 2 - Endoscopy images of stomach showing a large ulceroproliferative growth of size 8cm x 4cm noted along the greater curvature with overlying sloughed off mucosa.

The bone histopathology on Immunohistochemistry staining of tumour cells shows diffuse positivity for CD45, CD20, BCL2, BCL6, MUM 1 (FIG 3), Ki67 was 80% positive. Negative for CK7, CK20, CDX2, CD15, CD30, CD10 and Cyclin D1. Overall histopathology favoured the Diagnosis of Non-Hodgkin's Lymphoma diffuse large B cell type (DLBCL)- germinal centre B type.

Histopathology and IHC staining on the gastric biopsy was positive for CD45, CD20 and negative for CD3 and CK7 indicating High-grade B cell lymphoma. Further IHC staining for categorisation was not done.

Thus the diagnosis of a simultaneous high-grade B cell lymphoma in both bone and stomach was established. He was started on chemotherapy with an R-CHOP regimen for lymphoma and is on regular follow up.

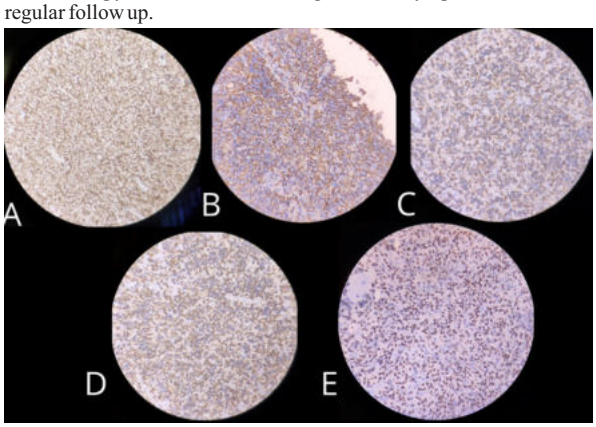

FIG 3 – histopathology of the bone lesion – showing highly cellular neoplasm composed of round polygonal cells with eosinophilic cytoplasm, vesicular nuclei, and prominent nucleoli with mitotic figures. Immunohistochemistry (IHC) showing diffuse positivity for CD 45 (A), CD 20 (B), BCL 6 (C), BCL 2 (D), Ki67(E).

DISCUSSION:

In our present case, we see a rare concomitant occurrence of both bone and gastric lymphoma. The likely primary site is bone, and the Gastric involvement may signify the spread of the disease from the primary site.

Histopathologically, most of the gastric lymphomas and 94% of the PBL are NHL, while diffused large B-cell lymphoma being the most common subtype (4) (11) (12).

Chronic H. pylori infection, immunosuppression, inflammatory bowel disease, and celiac disease can predispose to the development for the development of gastric lymphoma (6). On endoscopic examination, three patterns are recognised, ulceration, diffuse infiltration, and a polypoid mass (7). Endoscopically primary gastric lymphoma is indistinguishable from other types of gastric cancers though they play an essential role in obtaining tissue specimen and follow up. Radiological investigations are needed to establish the diagnosis and extent of the disease. Computerised Tomography (CT) chest and abdomen to exclude systemic disease, lymph node extension, and adjacent structures infiltration. Using imaging modalities thickening of the gastric wall can be identified in 85% of the cases while lymphadenopathy observed in only 50% of cases (8). Magnetic resonance imaging (MRI) shows thickened mucosal folds, irregular submucosal infiltration, annular constricting lesion, exophytic tumour growth, mesenteric masses, and mesenteric/retroperitoneal lymphadenopathy (9). Endoscopic ultrasound (EUS) can be used to assess the depth of invasion and assessing perigastric lymph nodes.

The definition of Primary bone lymphoma (PBL) has been inconsistent over the years. The World Health Organization (WHO) classification of soft tissues and bone tumours defined PBL as either single osseous lesions without regional lymph node involvement, or tumour involvement of multiple osseous sites without associated visceral or lymph node disease (10). In our patient though there is visceral involvement the development of bone swelling far precedes the GI manifestations and constitutional symptoms which made us consider bone as the primary site of involvement.

It occurs most commonly among men, and the most common age is the sixth and seventh decade of life. The typical clinical presentation is pain, surrounding soft-tissue swelling, palpable mass with or without systemic symptoms. The radiological features of a PBL are often non-specific. On MRI, the lesions often show hypointensity on T1-weighted imaging and hyperintensity on T2-weighted imaging and areas of enhancement within lesion with contrast administration (13).

The most commonly used chemotherapeutic regimen for DLBCL of the stomach and bone is a combination of cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) and Rituximab (R-CHOP).

For the treatment of PBL, radiotherapy is given in addition to chemotherapy to decrease the risk of local recurrence. But surgery is indicated for biopsy, prophylactic fixation of impending fractures, treatment of fractures before or after radiotherapy and systemic therapy, and theoretically in patients with disease unresponsive to conventional therapy (14).

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