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Case Report

T cell/histiocyte rich large B cell lymphoma of ascending colon: a rare entity

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ABSTRACT

Colonic T cell/histiocyte rich large B cell lymphoma (THRLBCL) is a very unusual occurrence never described before. A 41-year anaemic male presented with loss of weight and appetite for 7 months and fever with Malena for 1 month. Abdominal examination revealed a 4×6 cm retroperitoneal lump in the right iliac fossa. Radiological investigations (USG and CECT whole abdomen) reported an asymmetrical ill-defined growth in ascending colon and caecum with loco-regional lymphadenopathy. Surgical exploration revealed an ascending colon mass with retroperitoneal lymphadenopathy. Right hemi-colectomy with end ileostomy was done and specimen was sent for histopathology which diagnosed it to be a case of THRLBCL of colon. Patient was followed up after 2 weeks and was planned for chemotherapy.

Keywords: Colonic epithelial malignancy, Diffuse large B-cell lymphoma, Lenalidomide, Nodular lymphocyte predominant Hodgkin lymphoma, T cell/histiocyte rich large B cell

INTRODUCTION

T cell/histiocyte rich large B cell lymphoma (THRLBCL) is classified as a subset of the DLBCL according to the revised WHO classification of hemato-lymphoid neoplasms 2017. It is a high grade, aggressive non-Hodgkin lymphoma, affecting mainly middle-aged men.[1,2] Morphologically it mimics NLPHL and classic Hodgkin lymphoma (cHL). It mainly has predilection for lymph nodes, spleen and liver but rare extra-nodal sites include thyroid, small intestine and thymus.[3-5] THRLBCL of colon is an extremely unusual presentation and very rare.

CASE REPORT

A 41 year male farmer, presented with progressive weakness, loss of weight and appetite for 7 months and low-grade fever and Malena for 1 month. No history of vomiting, abdominal pain, and constipation. He was a chronic smoker. Family history was insignificant. No history of concurrent medical illnesses and long-term drug intake. On examination he had fair general condition, pallor was present and abdominal examination revealed a 4×6 cm retroperitoneal lump in the right iliac fossa. No generalized lymphadenopathy was present. Per rectal examination was normal.

Investigations

Hemoglobin: 5.4 g%, serum CEA: 2.29 mcg/l

USG whole abdomen: Hypo-echoic mural thickening of distal ileum, ileo-caecal junction and caecum with multiple enlarged lymph nodes.

CECT whole abdomen: 11 cm long, asymmetrical, circumferential heterogeneously enhancing wall thickening in caecum and ascending colon. Multiple

enlarged necrotic peri-pancreatic, periportal and aortocaval lymph nodes were also noted (Figure 1).

Colonoscopy: Multiple ulcers with surrounding edema extending from hepatic flexure up to caecum. Intervening mucosa was normal in appearance.

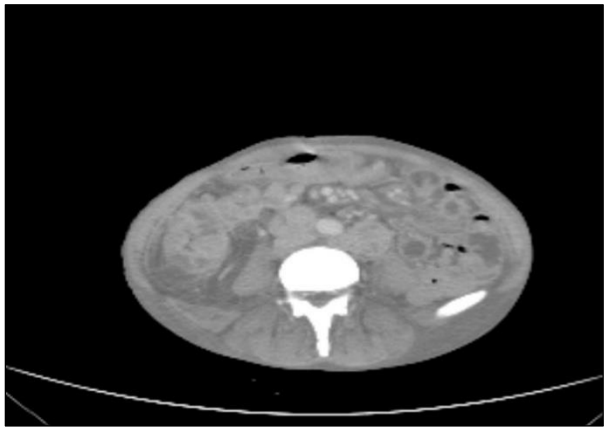

Figure 1: CECT whole abdomen depicting irregular growth in the ascending colon.

Differential diagnosis

Colonic epithelial malignancy, ileocaecal tuberculosis, Crohn's colitis.

Treatment

Patient was planned for elective laparotomy after adequate pre surgical optimization. Surgical exploration revealed an ulcero-proliferative growth in ascending extending till the hepatic flexure and fixed to a knuckle of second part of duodenum superiorly. Multiple lymph nodes, largest measuring 3 cm identified at the root of mesentery, mesocolon, and retroperitoneum. No omental, peritoneal or serosal nodules seen. Patient underwent palliative (in view of melaena) right hemicolectomy with sleeve duodenectomy with end ileostomy, distal stump closure along with duodenal rent repair. The specimen was submitted for histopathology. Hospital course of the patient was uneventful, and he was discharged after 10 days.

Outcome and follow-up

On gross examination, grey white ill-defined nodular growth measuring 6.5×4×2.7 cm in ascending colon was identified (Figure 2). Histopathology showed polymorphous population of cells with many dispersed large atypical cells dispersed in a background of lymphocytes, histiocytes, few eosinophils, and plasma cells. The large cells displayed round, lobated, or irregularly folded nuclei and distinct nucleoli and moderate amount of cytoplasm. (Figure 3) Four out of seven peri colonic lymph nodes were also involved. Immunohistochemistry showed large atypical cells positive for vimentin (Figure 4), LCA (Figure 5), CD30, Bcl2 and PAX5 (Figure 6). The background T cells and histiocytes were CD3 (Figure 7) and CD68 (Figure 8) positive.

Figure 2: Grey white ill-defined nodular growth which measures 6.5×4×2.7 cm inn ascending colon.

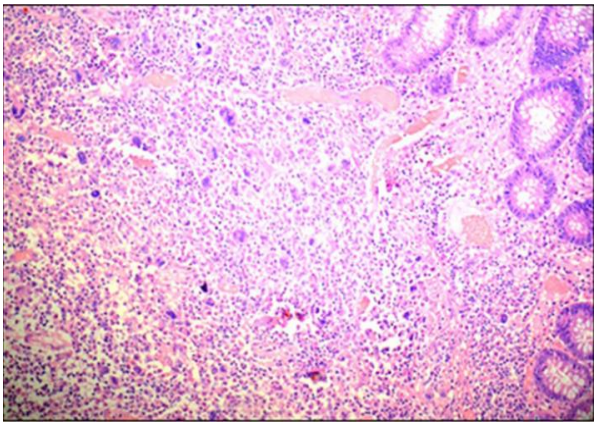

Figure 3: Section from colonic growth showing atypical cells with round to irregular folded nuclei, distant nucleoli and moderate cytoplasm. Few bizarre cells along with polymorphous population also noted (20X).

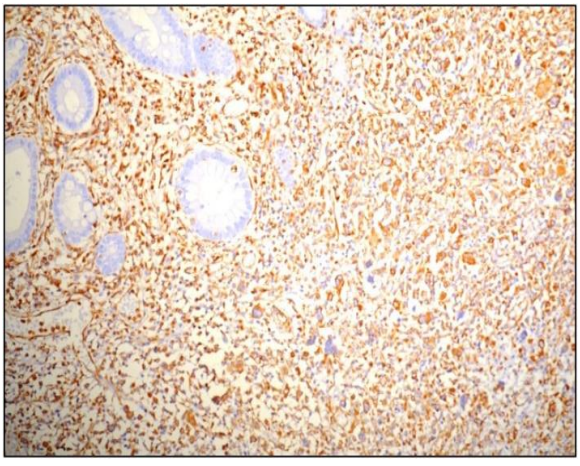

Figure 4: Photomicrograph of positive immune staining for vimentin (40X).

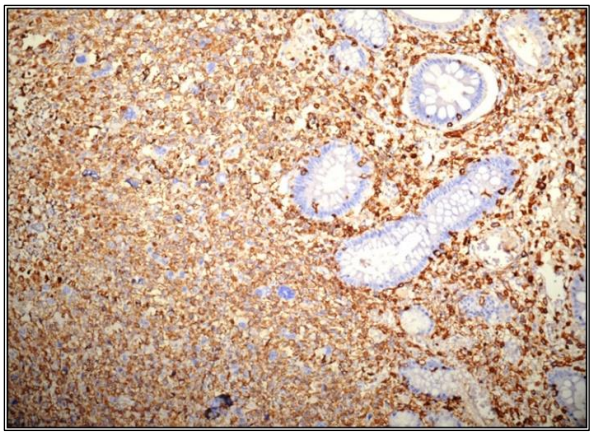

Figure 5: Photomicrograph of positive immune staining for LCA (40X).

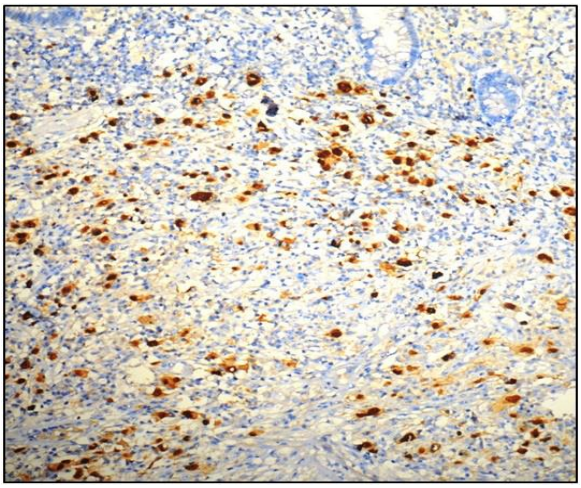

Figure 6: Photomicrograph of positive immune staining for PAX 5 (40X).

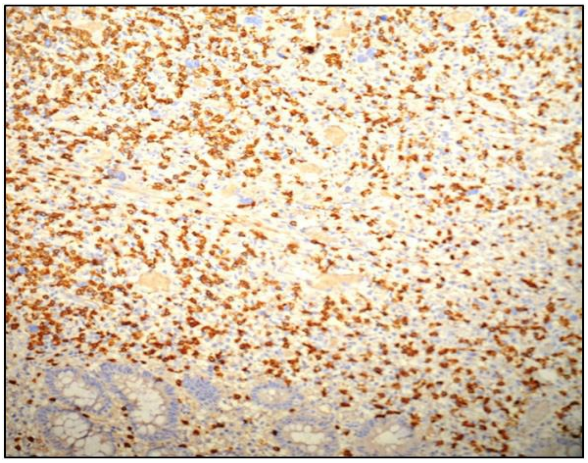

Figure 7: Photomicrograph of positive immune staining for CD 3 (40X).

CK, DOG1, CD117, SMA, CD15, Alk1, CD20 and CD56 were negative. Hence a diagnosis of colonic T/HRBCL of colon was established. Patient was started on CHOP-R (Cyclophosphamide, hydroxy-daunorubicin, oncovin, prednisolone, rituximab) chemotherapy regime and advised routine to follow up.

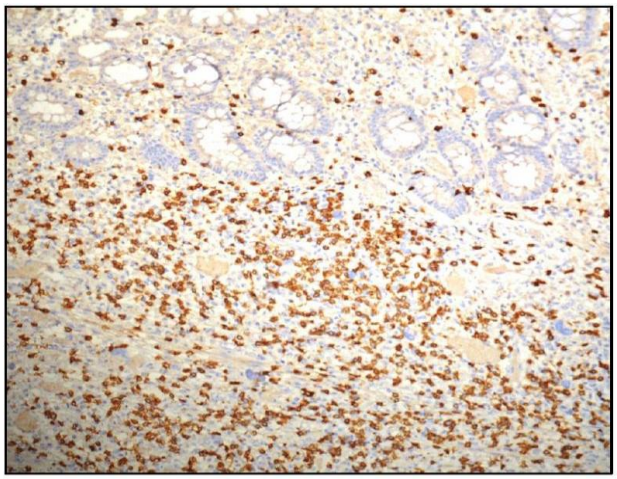

Figure 8: Photomicrograph showing positive immune staining for CD 68 (40X).

DISCUSSION

THRLBCL is a rare morphologic variant of DLBCL with limited number of pan B (CD20 or Pax5) positive neoplastic B-cells with abundant population of the non-neoplastic CD3 positive T-cells and CD68 positive histiocytes.[6-8] It causes diagnostic dilemma since it mimics the NLPHL. Morphologically diffuse architecture without rosette around atypical cells, numerous histiocytes and immunophenotypically pan B cell positivity in scant tumor cells along with CD 68 and CD3 expression in histiocytes and T lymphocytes respectively favors diagnosis of TCRLBCL than NLPHL.[9] There is also a hypothesis that the NLPHL progress to this subtype of DLBCL.[10] Gottesman et al, identified that the background population is distinct in both, as THRLBCL shows strong positivity of reactive T-cells in the background whereas the NLPHL shows reactive B cells.[9] Hartmann S et al, have analysed the gene expression profiling of these entities and found BAT 3/BAG 6, HIGD1A AND FAT10/UBD expression in both.[11] Using comparative genomic hybridization Franke S et al, concluded that TCHRLBCL possibly originates from the same precursor cell of NLPHL.[12]

Correct diagnosis is essential for management as limited field radiation therapy is administered for NLPHL in contrast to THRLBCL which is treated primarily by chemotherapy. Siricilla et al have reported a case of chemotherapy refractory THRLBCL like transformation of NLPHL which was then successfully treated with lenalidomide.[13] Surgical debulking followed by adjuvant chemotherapy (R-CHOP) is the mainstay for localized and non-metastatic gastrointestinal disease. El Weshi et al, has reported a 5-year overall survival rate of 46% with aggressive course and poor outcome.[14]

CONCLUSION

THRLBCL in colon is very rare and it can be diagnosed appropriately with the help of ancillary studies like immunohistochemistry and molecular diagnostics. Morphologically it consists of abundant CD 3 positive non neoplastic T cells and CD 68 positive histiocytes with limited number of CD 20 and PAX 5 positive neoplastic B cells. Identification of this entity is crucial since it very closely mimics NLPHL and treatment is entirely different. Chemotherapy (CHOP-R) is the primary treatment modality. Prognosis as reported in literature is very poor.

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