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Primary MALT Lymphoma of Appendix: A Rare Case Report.

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ABSTRACT

Malignant lymphomas comprise 1-4% of the malignant neoplasm's of the gastrointestinal tract but the appendiceal lymphomas are exceedingly rare with nonspecific clinical findings leading to delay in diagnosis. The incidence of primary appendiceal lymphoma is 0.015% of all gastrointestinal lymphomas. This is a case report of primary MALT lymphoma of appendix in a 35 year female with presentation of acute appendicitis and right lower abdominal pain which presented as appendicular mass on CT examination.

Keywords: MALT Lymphoma, appendicitis, laproscopic

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INTRODUCTION

The gastrointestinal tract is the most common site involved by extra nodal non Hodgkin's Lymphoma's and it accounts for 30-45% of all cases [1]. Most commonly involved organ is stomach followed by small intestine, colon & esophagus organs [2]. Primary lymphomas of appendix are extremely rare tumors with incidence of 0.015% of all gastrointestinal lymphomas [1,3]. Here we report a case of primary MALT lymphoma of the appendix which presented as with complain of appendicular mass in our institution, which was excised and sent for histological examination.

CASE REPORT

A 35 year old female was admitted in our institute with right lower abdominal pain, nausea and vomiting for last 24 hours. Her laboratory values included hematocrit 32.5%(normal 40%-52%), Hb 12 g/dl (14-18 g/dl), white blood cell count 8500/μl (3800-10,500/μl), (with 49% neutrophils (45-75%), 51% lymphocytes(20-51%), platelet count 95000/μl (1,50,000-4,50,000/μl), ESR 15mm/h (0-20mm/h). On examination there was tenderness in the right iliac fossa. The clinical diagnosis was made as acute appendicitis. The abdominal ultrasound showed appendicular mass. On CT, there was marked homogenous enlargement of the appendix with thickened walls. The patient underwent laproscopic appendicectomy.

Grossly, The appendix was symmetrically and markedly enlarged and measured 8.5cm in length and 3.0 cm in maximum diameter. On cut section, the wall of appendix was grossly thickened with fish flesh like consistency; lumen was compressed and obliterated (Fig 1).

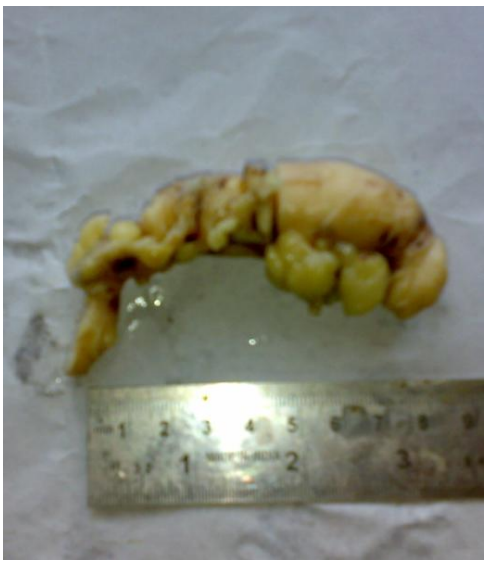

Fig.1

Microscopy: The section showed diffuse transmural infiltration by atypical lymphoid cells, which were small to medium sized with slightly irregular nuclei, inconspicuous nucleoli and relatively moderate cytoplasm (Fig.2).

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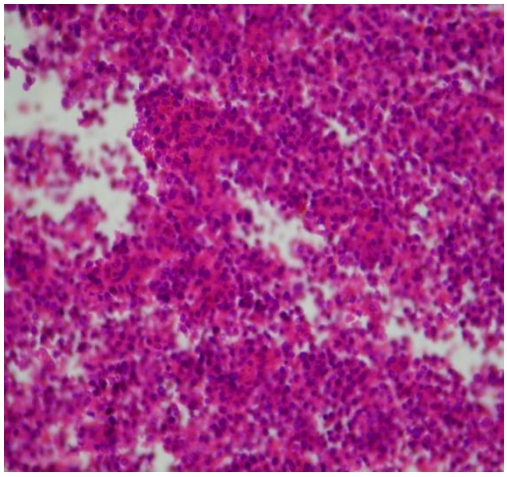

Fig.2

Immunohistochemical study showed positively for CD20 (Fig. 3) where as it was negative for CD5 & CD10. Thus the diagnosis of Marginal zone B-cell lymphoma was established. The postoperative course of the patient was uneventful and the patient was discharged without any complication.

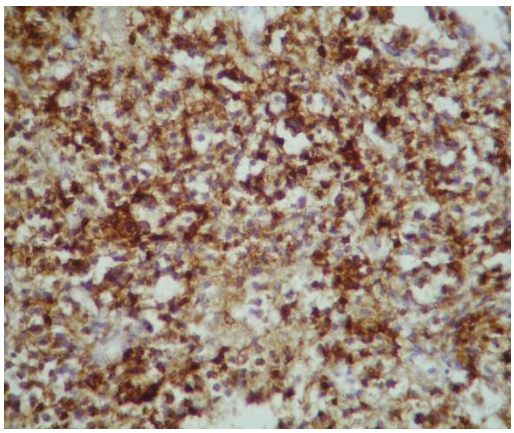

Fig.3

DISCUSSION

Gastrointestinal tract are frequently involved by non Hodgkin's lymphoma with the incidence of 4-20% of all NHL and 30-45% of extra nodal cases [2]. Information on appendiceal lymphoma is limited to few scattered case reports and letters. It has been inconsistently reported in large series of gastrointestinal lymphomas, but the frequency of involvement ranges from 1-3%only [4, 5]. Malignant lymphomas comprise 1-4%of the malignant neoplasm's of the gastrointestinal tract but the appendiceal lymphomas are exceedingly rare with nonspecific clinical findings leading to delay in diagnosis [6,8].

The reported incidence of primary appendiceal lymphoma is 0.015% of all appendectomy specimens they usually present in the 2nd or 3rd decade of life. The patient most of the time presents clinically as acute appendicitis. In a review of 68 patients of

appendiceal lymphoma, 43 patients were presented with acute or sub acute right lower quadrant pain and others were either incidentally found or presented with nonspecific symptoms [6]. Another report of 46 patients with appendiceal lymphoma showed that 31 patients presented with acute or sub acute right lower quadrant pain where as five lymphomas were incidentally discovered [1,2]. The overall incidence of non-Hodgkin's lymphoma of the gastrointestinal tract has increased over the last two to three decades with the use of CT imaging [2].

Armed Forces institute of pathology reported 5 cases of Non–Hodgkins lymphomas of appendix retrospectively discovered on CT . Three of the patients had the initial diagnosis of acute appendicitis, one patient presented with lower gastrointestinal bleeding and the other had relapse of lymphoma diagnosed six year earlier [1,2].

According to previous reported cases the diameter and length of appendix involved by lymphoma varied from 2.5-4 cm and 7-17cm respectively [2,8]. Diffuse enlargement of the appendix due to lymphomatous infiltration was noted in all patients with average diameter of 3.0cm [5, 9]. In the present case the diameter of the appendix was 2.5cm and length was 8.5cm.

In a series of 19 cases of appendiceal lymphomas reported in a last three decades, the most commonly reported appendiceal lymphoma was Burkitt's lymphoma (9 cases) followed by diffuse large B cell lymphoma (4cases), T cell lymphoma (3 cases), mantle cell lymphoma (2 cases), and marginal zone B-cell lymphoma (1 case) [1,8]. Another mucosa associated lymphoid tissue lymphoma (MALT) comprise a group of indolent B cell Non-Hodgkin's lymphomas, which is a rather uncommon pathology in appendix and to our knowledge there are only few reports with appendiceal MALT lymphoma in the world literature [7]. In majority of cases MALT lymphoma involve the stomach. Clinical symptoms of MATL lymphoma are vague and varied with abdominal pain being the most common presenting complaints followed by nausea vomiting or bleeding [10]. Yoshino et al [11] found only 2% of MALT lymphoma with multiorgan involvement. Overall survival of localized and disseminated MALToma are similar [10]. MALT lymphoma of appendix is very rare entity; it was first reported by Ramazan K et al [12] with appendiceal intussusceptions. As some cases are incidentally discovered it is mandatory that all appendicectomy specimens are examined histologically.

We reported primary appendiceal MALToma in a middle aged female. To the best of our knowledge it is very rare entity.

CONCLUSION

MALT lymphoma, which is very rarely involved appendix, was incidental finding in histological examination in our case. The involved appendix was markedly and entirely enlarged, soft and fish flesh like in consistency. Immunohistochemical examination done for the confirmation of diagnosis.

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