Case Report

Primary Mucosa-Associated Lymphoid Tissue (MALT) Lymphoma Involving Cecum A Case Report From Saudi Arabia

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ABSTRACT

MALT Lymphoma is now recognized as a distinct subtype of non-Hodgkin's Lymphoma (NHL) with distinguishable immunopathological characteristics. MALT lymphoma of GI tract is rare and most of the cases are found in the stomach, other parts of GI tract are affected very rarely. Here we present a case of a sixty-four years old Saudi female diagnosed with cecal MALT lymphoma, presented to gastroenterology clinic with chronic mild nonspecific abdominal pain for several years. She was treated initially as irritable bowel syndrome, subsequently underwent CT abdomen and colonoscopy, which revealed subepithelial cecal mass. Biopsy and immunohistochemistry were consistent with cecal MALT lymphoma.

Key words: Cecal MALT Lymphoma, Extra nodal Non-Hodgkin's lymphoma, Intestinal Marginal Zone Lymphoma (I-MZL).

الملخص

يعتبر الآن سرطان الغدد الليمفاوية المرتبطة المخاطية الانسجة كنوع فرعي متميز من سرطانات الغدد الليمفاوية غير هودجكن مع خصائص مناعية مميزة. وهو من الأورام نادرة الحدوث بالجهاز الهضمي ومعظم الحالات وجدت في المعدة، تتأثر الأجزاء الأخرى من الجهاز الهضمي نادرا جدا. ونحن نقدم حالة من النساء السعوديات تبلغ من العمر ٦٤ سنة تم تشخيصها بسرطان الغدد الليمفاوية الشعير بالمعي الأعور، قدمت إلى عيادة أمراض الجهاز الهضمي مع ألم مزمن بالبطن غير محدد وبسيط استمر لعدة سنوات. عولجت المريضة في البداية كمتلازمة القولون العصبي، وخضعت في وقت لاحق لأشعة البطن وتنظير القولون، والتي كشفت عن كتله ورمية في القولون الأعور، وكانت نتائج الخزعة متسقة مع سرطان الغدد الليمفاوية الشعير.

INTRODUCTION

MALT Lymphoma is a sub type of Non- Hodgkin Lymphoma involving mucosa associated Lymphoid Tissue (MALT). It is labeled as a separate entity because it involves lymphoid proliferation in mucosa associated lymphoid tissue that lines the body organs or cavities rather than lymph nodes, this includes GI tract, lungs, eyes, skin, salivary glands, thyroid and breast. MALT lymphomas are usually indolent but with a potential to transform into a high-grade B cell lymphoma. MALT lymphoma represents only 5% of all non-Hodgkin lymphomas and majority of

Gastrointestinal MALT lymphomas affect the stomach. Intestinal lymphomas have not well investigated compared to stomach MALT lymphoma. Cecal MALT lymphoma is a very rare entity, we present a case of MALT lymphoma involving the cecum, probably the first case to be reported from KSA.

CASE REPORT

A 64-year-old Saudi female initially presented to a primary health clinic with chronic, mild and nonspecific abdominal pain and altered bowel habits for several years. Pain was relieved by defecation and not associated with other GI symptoms. She was treated as irritable bowel syndrome (IBS) initially, then she was referred to GI clinic for further evaluation. CT scan abdomen/ pelvis showed asymmetric circumferential wall thickening of the cecum measuring 3.1 X 4 cm (Fig.1). The Colonoscopy showed a subepithelial mass involving cecal base and appendicular orifice (Fig.2). histopathology and immunochemistry were consistent with diagnosis of MALT lymphoma of the cecum (Fig.3). We referred her to haemato-oncology team as a case of cecal MALT lymphoma for further management. They further investigated her with bone marrow aspiration which was normal. The consensus plan was closed observation and follow up, given that she has a limited disease (stage 1-2) with a potential plan to start chemotherapy if she develops invasive disease, becoming symptomatic or transformed to other type of lymphoma. Patient is being regularly followed with hemoto/ oncology for almost 2 years since the diagnosis and fortunately, the disease is still in same stage without progression.

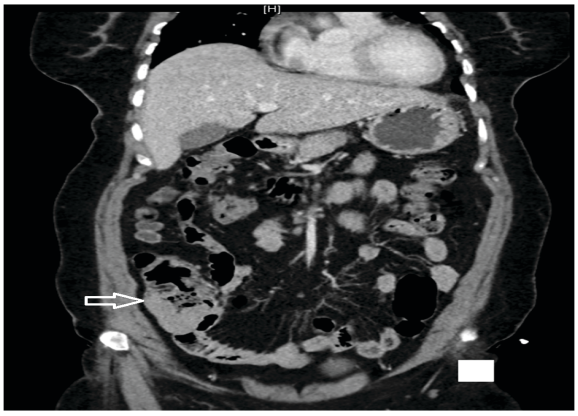

Fig (1) Abdomen

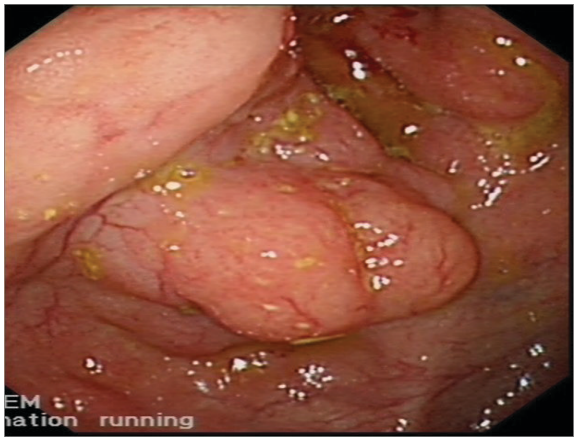

Fig (2) Colonoscopy

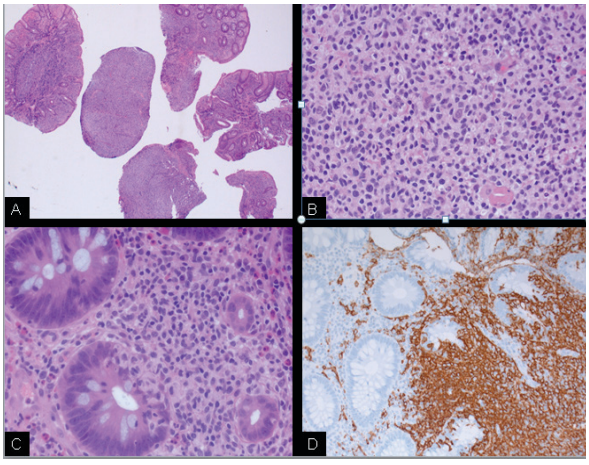

Fig (3) Histopathology

DISCUSSION

The concept of distinctive type of extra nodal B-cell lymphoma arising from MALT-derived lymphocytes in the gastro-intestinal tract was first time introduced in 1983 by Isaacson P and Wright DH [1]. MALT lymphoma is considered a sub type of non-Hodgkin lymphoma [2]. Mucosa-associated lymphoid tissue (MALT) is found along with the mucosal linings in the human body [3, 4]. MALT-type lymphomas are usually found in organs originally devoid of lymphoid tissue, which may be acquired by persistent antigenic stimulation by infectious or autoimmune processes [5].

MALT lymphoma is considered very rare and constitute only about 5% of all non-Hodgkin lymphoma [6]. In GI tract, two-thirds of all cases occur in stomach [7, 8] followed by small intestine, cecum, colon and rectum [9]. Increasing evidence suggests that etiology of MALT lymphoma may be related to chronic stimulation of infectious agents or autoimmune stimuli [10] Gastric MALT lymphomas are associated with H. Pylori infection in more than 90% of cases [11]. However, the etiology of non-gastric MALT lymphomas is not clear. The clinical presentation of MALT lymphoma is heterogeneous. It is quite nonspecific and is related to the organs involved. Pain, loss of appetite, and weight loss are the most common symptoms, while bleeding occurs more commonly with gastric involvement [12, 13]. Our patient initially presented with non-specific symptoms followed by altered bowel habits.

We searched the literature and found only few cases of MALT lymphoma involving cecum reported from different countries. This may be the first case of cecal MALT Lymphoma to be reported from Kingdom of Saudi Arabia. Previously few reported cases of cecal MALT lymphoma had various presentation notably with intestinal obstruction [14] and intussusception [15]. Our patient had just nonspecific symptoms and on further follow-up with haemato-oncology for approximately two years, she is asymptomatic without any specific treatment. It may reflect that her symptoms initially might be related to IBS and MALT lymphoma was just an incidental finding. The data about cecal and other part of the colon are scarce, and more data are needed to guide future management about this rare disease.

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