Primary Mucosa-Associated Lymphoid Tissue of the Gallbladder: Case Report

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Abstract

Primary gallbladder Mucosa-Associated Lymphoid Tissue (MALT) lymphoma is exceedingly rare. Less than 50 cases have been reported in the English literature. The majority of cases are found incidentally in the context of acute cholecystitis. Here, we present a rare case of primary gallbladder MALT lymphoma, found incidentally as a mass on abdominal computed tomography (CT) scan.

ABBREVIATIONS

MALT: Mucosa-Associated Lymphoid Tissue; CT: Computed Tomography; PET: Positron Emission Tomography

INTRODUCTION

Malignant lymphoma presents 0.1-0.2% of malignant tumors of the gallbladder [1]. Most of the literature regarding primary gallbladder lymphoma therefore consists of case reports, to the best of our knowledge about 50 cases have been reported in the English literature so far [1-6]. The mean age of presentation ranges between 35 and 69 years with a female prevalence [2]. Symptomatic acute cholecystitis represents the most common clinical presentation of the disease, followed by asymptomatic patients (incidental finding on CT). Incidental operative findings of MALT of the gallbladder, however, are most common in cases where the tumor mass in the gallbladder wall is not detected pre-operatively. In the event of pre-operative identification of a gallbladder mass, the distinction between gallbladder adenocarcinoma and lymphoma is hardly achieved without histological confirmation post-operatively [2,3].

CASE PRESENTATION

86-year-old-female with a past medical history of hypertension, presented with vague history of right upper quadrant pain. Computed Tomography (CT) was performed and revealed an incidental finding of a 2 cm mass in gallbladder wall which was read as a gallbladder polyp by radiology. The patient underwent a laparoscopic cholecystectomy. Intra-operative findings were significant for an area of discoid plaque on the exterior surface of the gallbladder wall. The specimen was removed en-bloc with the gallbladder and sent for pathologic analysis. The patient’s post-operative course was unremarkable and the patient was discharged on post-operative day 1. Pathology findings revealed, grossly, a 24x2x6 mm firm, homogenous, ovoid nodule without necrosis, ulceration or hemorrhage, confined to the sub-serosa. Microscopically, it showed diffuse proliferation of small lymphocytes and reactive germinal centers with inconspicuous nucleoli, clear cytoplasm (centrocytes), plasma cells with negative surgical margins. The immune-phenotype screening was positive for: CD20, CD43, and BCL-2 and negative for: CD3, CD5, CD10, CD23, BCL-6 and cyclin-D1. Pan-cytokeratin (AE1/3) stain highlighted the epithelium, further supporting the diagnosis of primary B-cell MALT lymphoma.

Post-operative lymphoma staging included esophagogastroduodenoscopy (EGD), Pan CT scan, PET-CT, and bone marrow biopsy. All of these studies were negative for any metastasis or primary MALT tumor. The CT scans were repeated at the one year follow up which did not reveal any evidence of recurrence.

DISCUSSION

MALT lymphomas were initially reported by Isaacson [7]
when he described a particular type of B cell lymphoma that arose from the gastrointestinal tract, which exhibited distinct histopathologic characteristics. Three main components make up the histological pattern: 1) centrocyte-like cells, 2) small lymphoid cells, and 3) plasma cells [7].

The etiology of MALT lymphomas involving the gallbladder is still unclear as the gallbladder itself does not contain lymphoid tissue. It has been suggested that bacterial infection and chronic inflammation cause lymphocytes to migrate to the gallbladder mucosa, forming secondary follicles [8]. The continuous antigenic stimulus may cause a chromosomal translocation, resulting in a fusion protein that inhibits apoptosis and causes antigen-independent proliferation [6]. This transformation, specific for MALT lymphomas, has been proposed for other sites normally devoid of lymphoid tissue, such as the stomach [9-11].

The radiological features of gallbladder lymphoma depend upon their pathological classifications. High-grade lymphomas, such as diffuse large B cell type, tend to form a solid and bulky mass in the gallbladder or have marked and irregular wall thickening. Most low-grade lymphomas display submucosal, homogeneous, wall thickening of the gallbladder which correlates with the pathological findings of homogeneous tumor cell infiltration within the submucosa [1,3]. Findings of homogeneous submucosal thickening of the gallbladder wall with a preserved mucosal surface on CT scan or magnetic resonance imaging (MRI) might also be suggestive of lymphoma [1]. Ultrasonography may also be sufficient for pre-operative identification of wall thickening with limited data reported. Proceeding with diagnostic cholecystectomy for histological diagnosis is still considered best practice for definitive diagnosis.

Primary MALT tumors of the gallbladder have an excellent prognosis with complete resection. All patients were reported to be alive at the time of this publication. Some authors reported, and some recommended, the use of adjuvant chemotherapy and/or radiation therapy; even for MALT confined to the gallbladder with negative margins. Recent evidence is now suggesting that excellent local control is achieved with surgical resection alone, without documented local recurrences and no effect on long term disease-free survival [12].

Postoperative staging including whole body PET CT and bone marrow aspiration should be completed as per lymphoma-staging workup recommendations in all patients.

SUMMARY

Primary MALT lymphoma of the gallbladder is exceedingly rare. The majority of cases are found incidentally in symptomatic elderly females with symptoms mimicking cholecystitis. Gallbladder polyps, non-homogeneous or patchy thickening of the gallbladder wall seen on imaging (with or without symptoms of cholecystitis), particularly in the elderly, should be investigated for possible lymphoma or other malignancy, even when no risk factors are present. These findings are an indication for cholecystectomy for definitive diagnosis.

REFERENCES