Research Article

Combination of MALT Lymphoma of the Nasopharynx and the Rectum: a Case Report

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Abstract: Mucosa-associated lymphoid tissue (MALT) is an extra-nodal marginal zone B cell lymphoma, it accounts for about 7-8% of all non hodgkinian lymphomas. It occurs most frequently in the stomach (70% of the cases), but can also occur on non gastric tissue especially the salivary glands, orbit, thyroid, lungs. However, colorectal and nasopharyngeal involvements are extremely rare. We report the case of a combination of these unusual locations in a 51-years-old patient in whom MALT lymphoma was diagnosed in 2006. The patient had rectal bleeding. Colonoscopy has shown the presence of large ulcerated folds of the rectum. Pathological and immunohistochemical study confirmed the diagnosis of MALT lymphoma. No extra intestinal involvement was found on the staging evaluation. The patient had a pelvic radiation at the dose of 40 Gy with endoscopic and histological remission. Eight months after the end of treatment, clinical examination showed an oropharyngeal mass. The computed tomography revealed a thickening of the posterior nasopharyngeal wall extended to the oropharynx. The diagnosis of MALT lymphoma was confirmed by the biopsy. The patient had radiotherapy on the Wadayer ring at the dose of 44 Gy. Currently, five years after the end of treatment, the patient is on complete remission.

Key words: Malt lymphoma, rectum, nasopharynx
1. Introduction

Mucosa-associated lymphoid tissue (MALT) lymphoma is a low-grade extra-nodal B-cell lymphoma including marginal zone. It represents 7-8% of all non-Hodgkin lymphomas (1). It occurs on native MALT (Lung, Small Bowl) or on acquired one resulting from a chronic inflammatory disorder (Stomach). Many authors suggest that it has its origin from an antigenic stimulation of autoimmune disorders (2). Stomach is the commonest site of MALT lymphoma followed by orbit, salivary gland, thyroid and superior respiratory tract (3). Rectal and nasopharyngeal locations have been rarely described in the literature. We report a case of a female patient followed for both rectal and nasopharyngeal MALT lymphoma, and we propose, through a literature review to discuss the etiopathogenic factors and therapeutic modalities of both locations.

2. Case Report

Mrs. H.T is a 51-year old woman, explored in 2006 for rectal bleeding associated with abdominal pain. The digital rectal examination revealed a circumferential and non-stenosing tumor. No further abnormalities were found in the clinical examination. Blood chemistry was normal. A colonoscopy found a large ulcerated rectal fold which was biopsied and the pathological study revealed a gastric B-cell lymphoma of MALT type with no Helicobacter pylori. The staging including an upper gastrointestinal endoscopy, a small bowel transit, a thoraco-abdominal-pelvic scan, a bone marrow and a nasopharyngeal biopsy revealed no abnormalities. The patient underwent a pelvic external beam radiation at 40 Gy. The clinical and endoscopic post-therapeutic controls were normal.

Eight months after a complete remission, Mrs. H.T complained from a swallowing pain. The oropharyngeal exam revealed a right-sided mass of the oral cavity. No cervical nodes were found. A head and neck scan found a thickened right side wall of the posterior nasopharyngeal mucosa with a choanal and an oropharyngeal extension (Figure 1). The Nasopharyngeal biopsy revealed a B-small cell lymphoma of the MALT-type with no evidence of Helicobacter pylori.

The patient underwent an external beam radiation therapy of the Waldeyer Ring at 44 Gy. Currently, five years after the end of treatment, the patient is in clinical, radiological and histological remission.

3. Discussion

The MALT lymphoma is a special pathological entity; it’s an extra nodal non-Hodgkin lymphoma. It consists on a monoclonal proliferation of centrocytes-like B lymphocytes from the mucosa of lymphoid tissue organs. These cells involve the epithelium and lead to lymphoepithelial lesions (1, 2). It occurs in adults with a median age at diagnosis of 61 years. It occurs in men as in women with a female predominance for thyroid and salivary glands involvements (2).

The gastrointestinal tract is the first location of MALT lymphoma interesting 50% of cases. In fact, the stomach represents by far the most common site reported in 70% of cases, followed by the small intestine representing 20 to 30% of cases (4). The colo-rectal location is rare, 60 to 74% of colorectal lymphoma occurs on the caecum which is rich in lymphoid tissue (5).

Rectal MALT lymphoma is exceptional and only a dozen cases have been reported in the literature. They are often revealed by non-specific clinical signs, including gastrointestinal bleeding and abdominal pain. On endoscopy, lesion of rectal MALT lymphoma appears to be indurated, budding, rarely ulcerative or infiltrative (6, 7, 8, 9).

Head and neck is the second most common location of MALT lymphomas (2). It occurs, in descending order, on orbit, on salivary glands and on Waldeyer Ring. MALT lymphomas represent less than 20% of Waldeyer Ring lymphomas: it occurs...
on palatine tonsils in 50-60% of cases, and nasopharyngeal mucosa in 25-30% of cases (10, 11).

As for the rectal MALT lymphoma, nasopharyngeal involvement is not frequent. There is no specific endoscopic or radiological aspect to this location: an asymmetrical thickening of the nasopharyngeal wall is most often described in endoscopy and computed tomography (12). Despite its low-grade, MALT lymphoma tends to be multifocal and disseminated. In fact two large studies about MALT lymphomas, including 158 and 108 patients, reported rates of spread respectively by 34% and 32% (13, 14). This is particular to head and neck MALT lymphoma: 50% of patients treated for head and neck MALT lymphoma, had at least one further location against 20-30% for gastric MALT lymphoma (15,16). In our case, the patient had two rare locations of MALT lymphoma.

The MALT lymphomas develop from the mucosa-associated lymphoid tissue in most cases acquired from chronic inflammation by antigenic stimulation or by an autoimmune disorder. The pathogenic agent has been identified for some locations, but remains poorly understood for others. Thus, the link between gastric MALT lymphoma and chronic gastritis Helicobacter pylori (HP) was established on pathological, epidemiological and therapeutic arguments (17, 18).

For non-gastric locations, the role of infectious agents has been identified. In fact, cutaneous lymphoma has been associated with infection by Borrelia burgdorferi (19), and the ocular lymphoma has been associated with infection by psittacii chlamydia (20).

The etiopathogenic link could, also, be established with some autoimmune diseases such as Hashimoto thyroiditis, in case of MALT lymphoma of the thyroid and Sjogren and sjogren lymphomes in the salivary glands MALT lymphoma (2).

For nasopharyngeal MALT lymphoma, the role of Epstein-Barr virus has been mentioned, but was never been clearly demonstrated (10, 11). As for colorectal locations, the role of Helicobacter pylori remains controversial. Some authors have shown a positive association with a regression of colorectal MALT lymphoma after HP eradication therapy, while others have described an inefficiency of tritherapy and suggest the role of other infectious agents present in the intestinal flora which are remaining to be identified (21, 22, 23). Histological study of our patient did not show the presence of HP.

The treatment of MALT lymphoma is standardized for the stomach location and it consists in the eradication of HP (17, 18). Radiotherapy is given in case of failure of tritherapy. Outside the gastric location, there are no therapeutic standards of MALT lymphoma. For locations in the head and neck, local control is the mainstay treatment and it consists on surgery and / or radiotherapy which is used as a first line treatment. In these cases, retrospective studies evaluating radiotherapy, at doses between 25 and 40 Gy, have objectified a 98% local control rate and a 5 years survival rate at 70% (24, 25). The matter of dose has been resolved in the last years and a dose of 30 Gy was adopted consensually as sufficient for the treatment of MALT lymphoma as for the other low-grade lymphomas (24, 26). However, the definition of target volumes to irradiate remains debated. Some authors include all the affected organ, others include GTV with a margin of 20 mm and include the affected organ and the first lymph node (24,26). In our case, the patient underwent an external beam radiation of the whole Waldayer Ring.

Due to the propagation and the recurrence of head and neck MALT lymphoma, systemic therapy, either chemotherapy or targeted therapy, has been discussed. Two retrospective studies, but with low enrollment, have shown no benefit with the addition of anthracycline-based chemotherapy in terms of survival, but these results remain to be validated by randomized trials (27).

Similarly, the combination of anti-CD20 monoclonal antibody (rituximab) to radiotherapy did not improve patient outcomes; in this case also, the study had a low enrollment and a short follow (28, 29).

Due to their rarity, the colorectal MALT lymphomas treatment is not standardized, and it’s very heterogeneous. Surgical resection or endoscopic mucosal resection is considered to be the treatment of choice by some authors because the disease remains, a long time, localized at the initial site (6, 7). In other reported cases, patients were treated with chemotherapy, most often a cyclophosphamide-based chemotherapy. The eradication of HP was
also tested and the reported results are controversial (21, 22, 23). The role of radiotherapy is poorly defined. Some authors have indicated radiotherapy in combination with chemotherapy, other in case of tritherapy failure. Only two cases of rectal MALT lymphoma treated with radiotherapy alone were reported, and as for our patient, complete remission was obtained. As in our case, dose levels were 40 and 45 Gy (8.30).

4. Discussion
Our reported case takes its particular feature from the Association of two unusual locations of MALT lymphomas and the choice of exclusive radiotherapy as a treatment modality for both locations that allowed control of the disease 5 years back.

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