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Case Report

Simultaneous MALT lymphoma of the thymus and parotid gland: independent lymphomas or metastatic spread?

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ABSTRACT

Background: Mucosa associated lymphoid tissue (MALT) lymphoma of the thymus is rare and generally associated with autoimmune disorders. It is generally suspected in middle-aged asian women with Sjogren's syndrome or other autoimmune diseases presenting a multicystic mediastinal mass. Contrary to other MALT lymphomas, the etiology of these tumors is still uncertain especially those affecting patients with no autoimmune diseases. Thymic MALT lymphoma with simultaneous salivary gland involvement is extremely rare, with only few cases described in literature.

Case presentation: We present a case of a 33 years-old male, with a recent history of MALT lymphoma of right parotid gland, affected by thymic mass. The patient underwent a right video-assisted thoracoscopy with radical thymectomy. The histological examination revealed a MALT lymphoma of the thymus. The molecular analysis on both thymic and parotid MALT lymphoma for clonal rearrangement of the immunoglobulin heavy chain gene suggests the origin from the same lymphomatous clone.

Conclusions: It is still unclear if multiple localizations of MALT lymphomas are attributable to the development of different primary lymphomas or can be caused by lymphatic metastatic spread. Our analysis reveal that the hypothesis of metastatic spread cannot be excluded in patients with simultaneous MALT lymphoma of the thymus and parotid gland, especially in non-autoimmune related MALT lymphomas.

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Background

Mucosa associated lymphoid tissue (MALT) lymphoma is an indolent non-Hodgkin lymphoma that generally arises in the gastro-intestinal tract. Salivary glands localization represents the most common non-gastric localization often arising in chronic inflamed glands such those of patients affected by Sjogren's syndrome or other autoimmune disorders. Thymic localization is rare with only few cases reported. The association with autoimmune disorders is well reported in literature. It is believed that continuous B-cells stimulation during autoimmune diseases could produce the development of primary thymic MALT lymphomas such as other mucosal sites. Conversely, it is unclear the etiology of non-autoimmune related thymic MALT lymphomas. We report a case of simultaneous MALT lymphoma of the thymus and parotid gland in a patient with no evidences of autoimmune disorders.

Case Presentation

A 33 years-old male came to our department after a surgical exeresis of the right parotid gland for Lymphoepithelial sialadenitis-associated MALT lymphoma of the parotid gland three months before. The post-operative computed tomography (CT) scan revealed an irregular mediastinal mass with two branches of 77x32 mm and 74x33mm placed above the right atrium and anteriorly to the ascending aorta (Figure 1). No compression of vascular structure has been demonstrated at both CT-scan and doppler-echocardiography. Suspecting a thymus primary neoplasm, the patient underwent a right Video Assisted Thoracoscopy (VATS) in general anesthesia. The mediastinal mass appeared partially cystic, elastic, in contact with the left anonymous vein and pericardium with no evidence of infiltration. Cautiously dissection of the tumor was performed, and the mass was completely removed and sent for

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histological examination. Histologically the neoplasm consisted of a lymphoid infiltrate of small-to-medium sized lymphocytes, positive in B cell markers (CD20+) with plasmocytic differentiation and k- light chain restriction. Whereas CD3, CD5 and CD10 were negative. Ki-67 expression was < 10% of the extra-follicular lymphocytes. The diagnosis of thymic MALT lymphoma was made. The postoperative course was uneventful, and the patient was discharged on post-operative day 3. In order to assess the correlation between the neoplasm and the right parotid gland lymphoma, molecular analysis for clonal rearrangement of the immunoglobulin heavy chain (IgH) gene was performed. After DNA extraction through Wizard SV Genomic DNA Purification System, the presence of clonal rearrangement was assessed using PCR amplification methods and specific primers for conserved genomic regions FR1, FR2 and FR3. Both samples were positive for the clonal rearrangement of the IgH gene, suggesting an origin from the same lymphomatous clone.

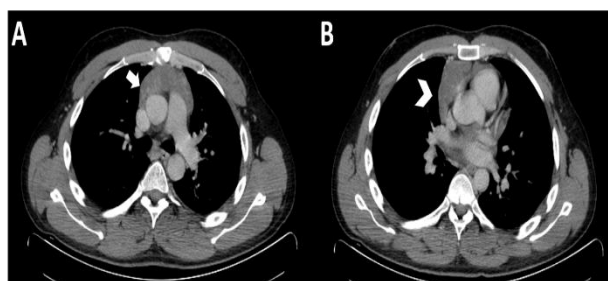


Figure 1: Preoperative CT-scan showing an irregular mediastinal mass placed above the ascending aorta (1A – arrow) and right atrium (1B – arrowhead)

Discussion

MALT lymphoma is an extranodal low-grade B-cell lymphoma that arises from Mucosa Associated Lymphoid Tissue (MALT) and represent 5% of all non-Hodgkin lymphomas [1]. It is localized more common into the stomach, but the most frequent non-gastric MALT lymphoma develops into the salivary glands. It is believed that MALT lymphoma is associated with continuous B-lymphocyte system stimulation in organs with a chronic inflammation background. *Helicobacter pylori*-induced gastritis is the most common cause of gastric MALT lymphoma, while non-gastric MALT lymphomas arise often in patient with autoimmune diseases especially Sjogren's syndrome (SjS) [2]. Lymphoepithelial sialadenitis (LESA) is a benign lesion often found in the salivary glands of patients with SjS. It is characterized by marked lymphocytic epeitheliotropism associated to parenchymal atrophy and ductal hyperplasia [3]. Its association with MALT lymphoma of the salivary glands is well known, described for the first time by Schmid et al. in 1983 [4]. Primary MALT lymphoma of the thymus gland is a rare condition, firstly described by Isaacson et al. in 1990 [5]. It occurs characteristically in asian middle-aged women with a well circumscribed mediastinal mass containing cystic lesions. Such as other MALT lymphomas, it is associated to autoimmune disorders, in particular SjS. It is believed that continuous polyclonal B cell activation of Sjogren's syndrome may cause lymphoma even if a precursor lesion has not been properly described. However, some authors suggest the existence of a LESA-like thymic hyperplasia that could promote the development of primary low-grade B-cell lymphomas in thymic gland [6]. Simultaneous MALT lymphoma of the thymus and salivary gland has been only reported in three cases previously [7-9]. There are no studies that prove the histopathologic relationship between thymic and salivary MALT lymphoma localization. Di Loreto et al. suggested that lymphomas originating from the same original clone assume a lymphatic spread from one mucosal side to another [8]. Otherwise, Nagasaka et al. asserted that two different primary lymphomas could develop in patients with autoimmune disorders [10].

In our case, the similar histological population of the neoplasms, especially the same k- light chain restriction and IgH rearrangement, and the absence of chronic autoimmune disorders suggest a development from the same lymphatic clone. There is no evidence of a possible metastatic spread between MALT lymphomas of the thymus and salivary glands, however in our opinion that possibility cannot be excluded, especially in patient without history of chronic autoimmune disorders.

According to literature, the thymus gland was completely resected [11].

Conclusions

The histopathologic relationship between simultaneous thymic and salivary glands MALT lymphomas is still unclear. The current evidences suggest that patient with an history of autoimmune disorders could present independent primary lymphomas while the hypothesis of metastatic spread cannot be excluded in non-autoimmune related MALT lymphomas.

List of abbreviations

CT - computed tomography
LESA - Lymphoepithelial sialadenitis
MALT - Mucosa associated lymphoid tissue
SjS - Sjogren's syndrome
VATS - Video Assisted Thoracoscopy

Consent for publication

Not applicable.

Conflict of interests

None.

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