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Follicular Lymphoma Development in Primary Sjögren's Syndrome: A Case Report

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Abstract

This paper reports a malignant transformation of Sjögren's syndrome to extraglandular follicular lymphoma in a 33-year-old female. The developing lymphoma has manifested itself in the cervical lymph nodes after seven years of diagnosing Sjögren's syndrome. To date, several hundred cases have suffered a malignant transformation of which few cases have developed follicular lymphoma. Given the overall transformation potential, the necessity of the close follow-up in patients with Sjögren's syndrome cannot be overemphasized.

Keywords: Sjögren's syndrome; Malignant potential; Follicular lymphoma

Introduction

Sjögren's syndrome (SS) is an idiopathic systemic autoimmune disease, characterized, histologically, by lymphocytic infiltrates, substantial acinar atrophy, and dysfunctional exocrine glands—inducing a general status of "*autoimmune epithelitis*" [1]. Manifesting itself classically in xerophthalmia, keratoconjunctivitis sicca and xerostomia, SS also shows numerous systematic manifestations that hit almost every organ. Moreover, the underlying idiosyncrasy of the autoimmune system may develop, unluckily, a malignancy. In this affect, the incidence of developing lymphomas and squamous cell carcinomas is the most frequent [1,2]. Follicular lymphoma (FL) is a mature B-cell neoplasm of non-Hodgkin lymphoma, with a morphologically follicular pattern. Grading of FL ranges, according to the number of centroblasts per high-power field, from grade 1 - comprising low numbers of centroblasts (0-5)- to grade 3b, marked by solid sheets of these same cells. However, FL relates, most often, to a clinically indolent form. As such, FL is characterized by slow progression and high response rates to therapy [3-5]. This paper reports a progressive course of SS in a young female; developing a follicular lymphoma (Grade 1) in a 7-year-interval of follow up.

Case Presentation

A 33-year-old female presented to our hospital in 2008 with undiagnosed disease. Her signs and symptoms included dry eye and mouth, swollen parotid gland, numb lower lip, and general fatigue. The serological findings characterized a remarkable increase in the ESR, mild anemia, lymphcytopenia, positive ANA as well as strongly positive Anti-Ro and anti-La. The sonographical study revealed miliary cystic cavitation, set against a heterogeneous parenchyma with sporadic calcifications. The histological examination of the minor salivary gland biopsy displayed impressive lymphocytic foci which effaced a significant part of the glandular architecture (Figure 1). By the same token, there was a conspicuous acinar degeneration and few epimyoepithelial islands were also seen (Figure 2). Based on this clinic-pathological picture, the diagnosis of primary Sjögren's syndrome was made. Accordingly, the patient was educated about her medical condition and advised to comply with a regime of close follow-up. In September 2015, the patient presented with bilaterally swollen cervical lymph nodes with increased reactivity seen on PET CT. The biopsy of the right cervical lymph node showed mostly a monotonous population of small lymphoid cells, in follicular arrangements, with irregular, angulated nuclei, inconspicuous nucleoli, and scant cytoplasm (Figures 3 and 4). The follicular proportion was conspicuous. There were neither focal areas of grade 3 nor diffuse large B-cells in the examined specimens. The patient was scheduled for chemotherapy.

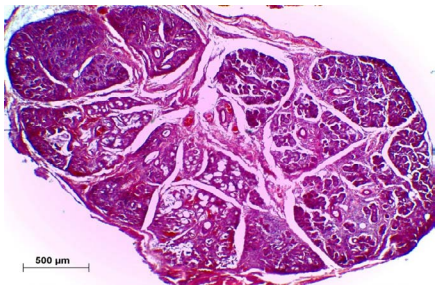

Figure 1: The histological examination of the minor salivary gland biopsy displayed impressive lymphocytic foci.

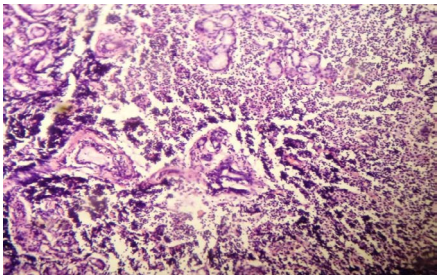

Figure 2: Conspicuous acinar degeneration and few epimyoepithelial islands.

Discussion

Follicular lymphoma is mostly a low-grade B-cell neoplasm encountering, mostly *de novo*, in adults, with a median age of 59 years

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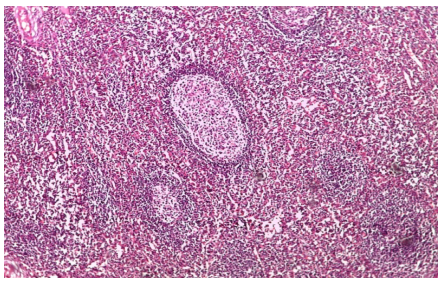

Figure 3: The biopsy of the right cervical lymph node showed mostly a monotonous population of small lymphoid cells, in follicular arrangements, with irregular, angulated nuclei, inconspicuous nucleoli, and scant cytoplasm.

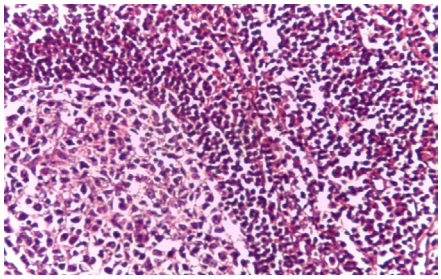

Figure 4: The biopsy of the right cervical lymph node showed mostly a monotonous population of small lymphoid cells, in follicular arrangements, with irregular, angulated nuclei, inconspicuous nucleoli, and scant cytoplasm.

[3]. However, the incidence of developing malignant lymphomas in the patients of SS [6], especially if germinal centers organization is histologically evident, is said to be as risky as 44 times the healthy population [7]. Patients with SS are at higher risk of developing lymphoproliferative neoplasms than patients with other autoimmune disorders [7,8]. Of these, recent population-based case-control study found that marginal zone lymphoma was most strongly associated with SS, followed by diffuse large B-cell lymphoma (DLBCL) and MALT lymphoma [9]. The possible mechanisms of such transformation include, among many, T-cell dysregulation, abnormal B- cell biology (distribution, mutagenesis and clonal expansion), cytokines, additional oncogenic events such as the inactivation of proto-oncogenes and some liable viruses. Suspiciously-oncogenic viruses, in developing lymphomas, comprise herpes virus 6, cytomegalovirus, Epstein-Barr virus, human T lymphotropic virus type I, human immunodeficiency viruses, human intracisternal A-type retroviral particle, human retrovirus 5, and coxsackie virus 6. Such affectation, either primarily or secondarily, are controversially leveled at progressing SS into lymphomas [10,11]. In this reported case, the exact pathogeneitc pathway of transformation could not be detected.

Conclusion

Close follow-up for patients with Sjögren's syndrome is warranted due to the potential risk of malignant transformation. Follicular lymphoma may be considered a possible risk of SS.

References

1. Ottoman B, Ali R (2015) A rare case report of Sjögren's Syndrome in a young female with clinicosonographical characterization. Am J Ultrasound 2: 1-9.
2. Vitali C, Bombardieri S, Jonsson R, Moutsopoulos HM, Alexander EL, et al. (2002) Classification criteria for Sjögren´s syndrome: a revised version of the European criteria proposed by the American-European Consensus Group. Ann Rheum Dis 61: 554-558.
3. Fisher RI, LeBlanc M, Press OW, Maloney DG, Unger JM, et al. (2005) New treatment options have changed the survival of patients with follicular lymphoma. J Clin Oncol 23: 8447-8452.
4. Kridel R, Sehn L, Gascoyne R (2012) Pathogenesis of follicular lymphoma. J Clin Invest 122: 3424-3431.
5. Pereira D, Vilela V, Dos-Santos T, Pires F (2014). Clinical and laboratorial profile and histological features on minor salivary glands from patients under investigation for Sjögren's syndrome. Med Oral Patol Oral Cir Bucal 19: e237-241.
6. Kassan SS, Thomas TL, Moutsopoulos HM, Hoover R, Kimberly RP, et al. (1978) Increased risk of lymphoma in sicca syndrome. Ann Intern Med 89: 888-892.
7. Theander E, Vasaitis L, Baecklund E, Nordmark G, Warfvinge G, et al. (2011) Lymphoid organisation in labial salivary gland biopsies is a possible predictor for the development of malignant lymphoma in primary Sjögren's syndrome. Ann Rheum Dis 70: 1363-1368.
8. Tincani A, Andreoli L, Cavazzana I, Doria A (2013) Novel aspects of Sjögren's syndrome in 2012. BMC Med 11: 93.
9. Dong L, Chen Y, Masaki Y, Okazaki T, Umehara H (2013) Possible Mechanisms of Lymphoma Development in Sjögren's Syndrome. Curr Immunol Rev 9: 13-22.
10. Anderson L, Gadalla S, Morton L (2009) Population-based study of autoimmune conditions and the risk of specific lymphoid malignancies. Int J Cancer 125: 398-405.
11. Tapinos NI, Polihronis M, Moutsopoulos HM (1999) Lymphoma development in Sjogren's syndrome: novel p53 mutations. Arthritis Rheum 42: 1466-1472.