Case Report

Waldenstrom’s Macroglobulinemia - A coincidental finding in a case of carcinoma of buccal mucosa

Mahjabeen Salma1, Sudha Madhuri2, S. J. Nandish Kumar3 and S. Karimuddin Abdullah4

1Consultant Pathologist, Yashoda Hospitals, Somajiguda, Hyderabad.
2Consultant Pathologist, Yashoda Hospitals, Somajiguda, Hyderabad.
3Consultant Oncologist, Yashoda Hospitals, Somajiguda, Hyderabad.
43rd year MBBS student, Shadan Institute of Medical Sciences, Peerancheru, Hyderabad.

*Correspondence Info:
Dr. Mahjabeen Salma,
Associate Professor,
Department of Pathology,
Dr. VRK Women’s Medical College, Aziznagar, R. R. District, Andhra Pradesh, India
E-mail: drsalsa786@yahoo.com

Abstract
Waldenström's Macroglobulinemia (WM) is a clonal disorder of small lymphocytes that show maturation to plasma cells synthesizing IgM. It is the biological activity of IgM protein that determines most of the clinical manifestations of the disease. WM most closely corresponds to lymphoplasmacytic lymphoma or immunocytoma of the REAL classification of lymphoma. Here, we report a case of a 50 year old male with squamous cell carcinoma of buccal mucosa. On routine investigations we found autoagglutination of RBCs; on workup with this feature we had suspicion of Waldenström's Macroglobulinemia. The diagnosis of WM was established after cytomorphology and immunohistochemistry (IHC) analysis of the bone marrow which revealed the presence of a lymphoid/lymphoplasmacytid-like bone marrow infiltrate along with an elevated serum IgM level. Waldenström's Macroglobulinemia and its co-existence with autoimmune diseases and non hematological malignancies has already been described in the literature, but the present case was asymptomatic for WM with carcinoma of buccal mucosa, which is the first case reported so far.

Keywords: lymphoplasmacytic lymphoma, REAL classification, autoagglutination, immunohistochemistry analysis

1. Introduction
WM is a pleomorphic lymphoproliferative disorder characterized by the elevated levels of monoclonal immunoglobulin (IgM) protein secreted by malignant B-cells and paratrabecular lymphoplasmacytic infiltration in the bone marrow. The classical presentation of WM’s patient includes anemia, hepatosplenomegaly, lymphadenopathy and hyperviscosity. Infiltration of the bone marrow and extramedullary sites by malignant B-cells and elevated IgM levels account for the symptoms associated with this disease. Patients may develop constitutional symptoms, pancytopenia, organomegaly, neuropathy, and symptoms associated with immunoglobulin deposition or hyperviscosity. WM is incurable with current therapy, and half of the patients die of disease progression; median survival is approximately 5 years. The incidence of development of second cancers was not significantly different between asymptomatic and symptomatic WM and between treated and untreated patients.

1. Case Report
A 50 year old male, known patient of hypertension and diabetes mellitus, on treatment; non-smoker, occasional alcoholic, had undergone TURP 3 years ago, now presented with buccal mucosal lesion. On examination of oral cavity, a cystic lesion found on right side, inside the cheek. Biopsy of the lesion performed and diagnosis of squamous cell carcinoma was made. On correlating all the findings with IHC results, thus a final diagnosis of Waldenström's Macroglobulinemia was made.

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2. Discussion

The WHO includes Waldenstrom Macroglobulinemia in lymphoplasmacytic lymphoma category and excludes other subtypes of specific lymphoma with plasmacytic differentiation.63 Secretion of monoclonal IgM is common but IgA and IgG or an immunoglobulin light chain is also seen in some patients.6 In our case, we report a sharp M-band seen in gammaglobulin region detected by protein electrophoresis. The incidence of WM is approximately 5 cases per 1 million persons per year, and this disease accounts for approximately 1% to 2% of hematologic cancers.6,7,8,9 The incidence of WM is more commonly seen in white people compared to other groups of population.10 The median age at diagnosis varies between 63 and 68 years, and slightly more common in males.11 Our patient was 50 year old male, which is slightly younger than the cases described in other studies. The disease was originally described by Waldenstrom in 1944.12 In the original description of WM, Waldenstrom described two patients with oronasal bleeding, lymphadenopathy, anemia and thrombocytopenia, and an elevated ESR.13 Bone marrow and lymph nodes show infiltration of pleomorphic B–lineage cells at different stages of maturation.14,15 The cells express pan B-cell markers (e.g., CD19 and CD20) and typically test negative for CD3 and CD103.15 In the described case, IHC analysis of bone marrow biopsy showed infiltration of lymphoplasmacytoid CD20 positive cells.

A study by Alexanian et al. was done to evaluate the frequency and history of Waldenstron’s Macroglobulinemia. They found 27% patients to be asymptomatic with slow disease progression. They concluded that the outcome was similar for both asymptomatic and symptomatic WM.16 This case differs from the cases reported in literature, as here, the patient is having pre-existing WM without any symptoms but he presented with cystic lesion of buccal mucosa. This association is not a therapy related development of second malignancy (carcinoma of buccal mucosa) because the patient received neither chemotherapy nor radiotherapy except for the biopsy of lesion.

Waldenstrom’s macroglobulinemia is a lymphoid malignancy and the association between lymphoid malignancies and solid malignancies has been discussed in the literature.17 Coexistence of hepatic carcinoma, lung carcinoma, and some other malignancies with WM have been reported previously.18,19,20 Most of these case reports belong to patients in whom a second malignancy developed during the course of WM except the case reported by Hasegawa et al.13 An alternative explanation for the preponderance of second cancers in WM patients is disease-related immune suppression. The immunologic impairment associated to lymphoproliferative disorders could contribute to the pathogenesis of WM as well as to the development of additional malignancies.9 There might be a common genetic mutation or an immunomodulatory role of the first malignancy predisposing to the second. This could suggest that the existence of two malignancies in a patient could be a co-occurrence. Regardless of the cause and the pathogenetic mechanism, the awareness of an increased risk of second malignancy needs a careful oncohematological examination of WM patients.

References