



Case report

Mantle cell lymphoma of palatine tonsil: A case report

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1. Background

Mantle cell lymphoma (MCL) is an uncommon B cell non Hodgkin lymphoma (NHL) and comprises about 3–10% of adult onset of NHL [1]. It affects mainly lymph nodes. Extranodal sites that are commonly involved include the gastrointestinal tract, Waldeyer's ring, lungs and pleura [2]. Primary MCL of palatine tonsil is rare. Extensive review of English literature review revealed 17 previously reported cases [3–6]. We report a rare case of primary MCL of the palatine tonsil.

2. Case presentation

A 53-year-old female presented in early February 2020 with a six-month history of hoarseness of voice, dysphagia and loss of weight. Clinical examination showed an enlarged left tonsil. There was no cervical lymphadenopathy. The patient then underwent left tonsillectomy. Gross examination showed fragments of brownish tissue measuring 6 cm in aggregate with pale-tan fleshy cut surface. On microscopic examination, the parenchyma of the tonsil was replaced by neoplastic lymphoid nodules of varying sizes and diffuse infiltrate of atypical lymphoid cells. The lymphoid cells were of medium size with round to oval nuclei and scanty cytoplasm. No germinal centres were seen (Fig. 1). Immunohistochemically, the tumour cells were positive for CD 20, CD 5, CD 43 and cyclin D1 and negative for CD 23, CD 3, CD 10 and *bcl-6*. MIB-1 activity was seen in about 40% of the cells (Fig. 2).

Fluorescence in situ hybridization (FISH) was also performed using dual fusion probe (mixture of the LSI CCND1 probe labelled with spectrum orange and the LSI IGH probe labelled with spectrum green). The t(11;14)(q13;q32) translocation was detected in 98.5% (197/200) of the interphase nuclei scored. Positron emission tomography-computed tomography (PET-CT) scan showed residual tumour in the left palatine tonsil (2.4 cm in greatest dimension) with involvement of the left cervical node (measuring 1.3 cm). There were no conclusive Fluorodeoxyglucose (FDG)-PET-CT avid nodal or extra-nodal diseases demonstrated elsewhere. Peripheral blood examination was unremarkable. Bone marrow biopsy or gastroscopy was not performed. The clinical staging was considered as Stage IIB. The patient is being treated with chemotherapy. The drugs included rituximab (R) CHOP (cyclophosphamide, vincristine, doxorubicin, prednisone) alternating with R + high dose cytarabine. Patient is in remission at present, four months following therapy and PET-CT scan shows no residual tumour.

3. Discussion and conclusion

Mantle cell lymphoma (MCL) was recognized as a distinct entity by World Health Organization in 2001 [2]. It occurs more often in males, the male to female ratio of 2.5 [7]. The incidence of MCL in Asian countries is lower than in Western countries and the patients present at an earlier age [7,8]. The median age at initial presentation is 68 years extensive literature review by Dereci [3] revealed 15 cases of primary

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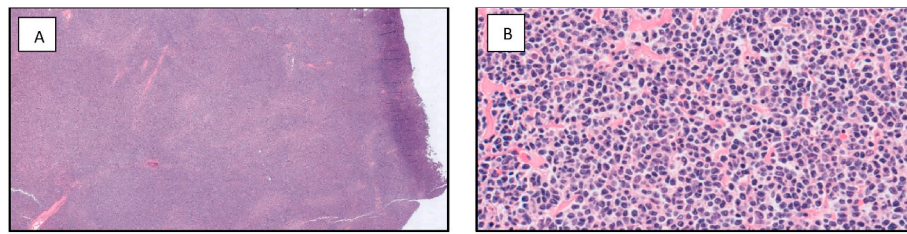


Fig. 1. (A) Effaced tonsillar architecture, replaced by diffuse sheets of atypical lymphoid cells (H&E x1). (B) A homogenous population of medium sized lymphocytes resembling centrocytes is seen, with irregular nuclear contours, condensed nuclear chromatin, inconspicuous nucleoli and scant cytoplasm (H&E x40).

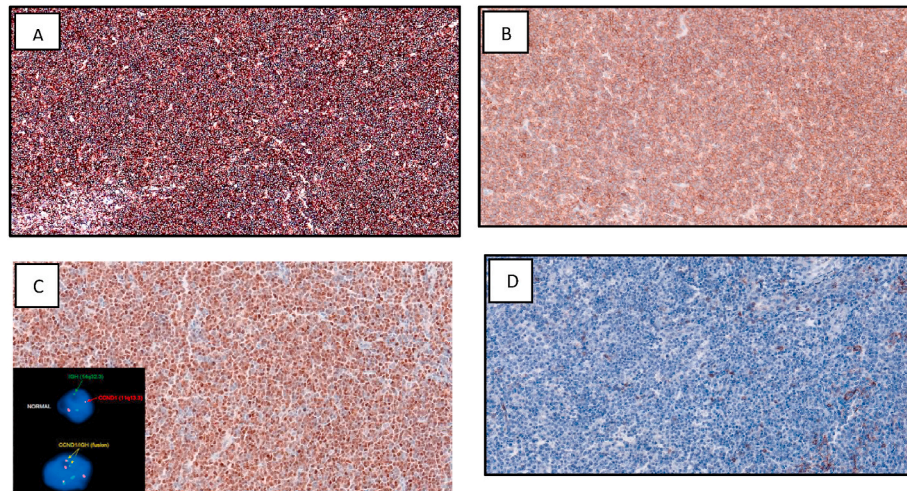


Fig. 2. Immunohistochemistry. (A) The tumor cells showing positivity for CD 20, (B) CD 5 and (C) Cyclin D1. Inset shows FISH for CCND1/IGH shows dual fusion signal with CCND1/IGH translocation. (D) The cells are negative for CD 23.

palatine tonsil MCL including cases from an earlier review by Guggisberg et al. [4]. Since then single case reports have been published [5,6].

The classical microscopic appearance of MCL is that of a proliferation of monomorphic small to medium sized cells with irregular nuclear contours harbouring inconspicuous nucleoli. The infiltrative pattern may be diffuse, nodular or the cells may exhibit mantle zone or follicular pattern. Blastoid and pleomorphic variants are biologically more aggressive [2]. Since MCL shares similar histological features with other small cell lymphomas such as small lymphocytic lymphoma, marginal zone lymphoma and follicular lymphoma, an immuno-histochemical panel is required for an accurate diagnosis. Typical cases of MCL express relatively intense surface IgM/IgD (more frequently with lambda than kappa restriction), pan B-cell antigen, Bcl-2, CD5 and CD43. The cells are negative for CD10 and *bcl-6*. A characteristic feature of MCL is the overexpression of nuclear cyclin D1 protein, expressed by more than 95% of MCL, as well as in the minority of cases that are CD5-negative [2]. Supplementary testing for FISH studies to demonstrate *CCND1/IGH* fusion is an important ancillary evidence in confirming the diagnosis. The chromosomal translocation of t (11; 14) (q13; q32) between an *IGH* gene and *CCND1* (encoding cyclin D1) is present in >95% of cases, rendering it to be the primary genetic event. This translocation results in deregulated overexpression of *CCND1*, leading to overexpression of cyclin D1 which is then assumed to overcome the suppressive effect of RB and p27 in the cell cycle, causing progression of cells from G1 to S phase [2]. Cyclin D 1 negative cases of MCL can be diagnosed by detection of expression of cyclin D 2. The clinical and biological behaviour is similar to that of cyclin D1 positive MCL [9].

Although MCL is not graded, evaluation of the proliferative fraction is critical as it has prognostic significance. High mitotic rate (>10–37.5 mitoses per 15 high-power fields or > 50 mitoses/mm²) and high Ki67 proliferation index (>30%; currently accepted cut-off point) are

associated with an adverse prognosis. Cases exhibiting less than 10% Ki-67-positive cells are considered to have a more indolent behaviour [2, 10]. Ki 67 expression has been included in the biological, prognostically important Mantle cell lymphoma Lymphoma International Prognostic Index (MIPI) score which incorporates patient's age, Eastern Cooperative Oncology Group performance score, lactate dehydrogenase level and white blood cell count [11] Typically, MCL has an aggressive clinical course that is resistant to therapy, with a median survival of 3–5 years [1].

In conclusion, MCL of the oral cavity is uncommon with a general predilection for the elderly. Immunohistochemical profiling is necessary for a precise diagnosis with an emphasis on overexpression of cyclin D1 nuclear protein. Fluorescent in situ hybridization study for detection of t (11; 14) (q13; q32) translocation also plays a big role as an adjunct in the diagnosis of MCL.

Declaration of competing interestDoCI

The authors have no conflicts of interest to disclose.

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CRedit authorship contribution statement

Jayalakshmi Pailoor: Diagnosing, designing and, Writing - original draft. **Venupriya Ramasamy:** Data curation, involved in collecting the data. **Yousoof Saira Bahnu:** Formal analysis. **Cheng-Eng Koay:** Management of the patient. **Kian-Meng Chang:** Management of the patient.

Pathmanathan Rajadurai: Diagnosing, designing and, Writing - original draft.

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