CASE REPORT

Symmetric palatal swelling as the first clinical manifestation of a mantle cell non-Hodgkin's lymphoma: A case report and review of literature

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ABSTRACT

The mantle cell lymphoma (MCL) is a rare (3.7%) low-grade non-Hodgkin lymphoma originating from the B-cell precursor-subpopulation. The clinical appearance in the oral cavity is rare. Since 1980, nine cases have been reported. A 41-year-old patient showed a MCL presenting with a symmetric, painless palatal swelling without any other clinical symptoms. Histological sections revealed malignant monotonous lymphoid cells (CD20+, CD43+, Ki67+) and the typical cyclinD1 over-expression by the chromosomal translocation t(11;14)(q13;q32). The proliferating cells weekly expressed CD5, kappa- and lambda-light chains and no EMA, CD10, bcl-6, CD30, and CD23. The patient was treated according to the European MCL younger study, and the MCL is regressive. The high incidence of dento-alveolar abscesses, inflammations, or benign tumor-formations leads to associate any maxillary or palatal swelling with this clinical condition. Considering the serious consequences of a missed therapy a histological examination of any untypical "swelling" is demanded.

Key words: Immunophenotyping, mantle cell lymphoma, symmetric palatal swelling

INTRODUCTION

The mantle cell lymphoma (MCL) is a relatively uncommon yet distinct type of malignant lymphoma whose clinical and pathological characterization is limited by the small number of cases published to date. The MCL is a lymphoid malignancy of B-cells of the mantle zone or primary lymphoid follicles. It was first classified in the revised European–American Classification of Lymphoid Neoplasms (REAL-classification,[1]).

MCL typically occur in the middle-aged to older adults with a marked male predilection. The involvement of the oral cavity is almost rare and only nine cases have been described in literature so far.[2] This report is unique, because of the uncommon, not previously described symmetric appearance on the hard palate.

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The few previously described cases with oral manifestation presented all with an all unilateral localization on the palate or the tongue.[2]

The primary clinical manifestation of a non-Hodgkin lymphoma (NHL) and especially the MCL in the oral cavity is very rare and uncommon. Estimation of the right diagnosis was impossible because of the absence of any other clinical symptoms besides the growing symmetric tumor-formation on the palate.

The exact differentiation and classification of this NHL subtype is very important for therapeutic investigations and for the outcome and variable prognosis [Table 1].

Histological sections of MCL classically describe a mantle zone, a nodular or a diffuse pattern of architecture. The definitive diagnosis of the MCL is predicted on appropriate immunohistochemical staining with an over-expression of cyclinD1.[3] Other immunohistochemical features are strong expression of CD20+, CD43+, Ki67+ and no expression of bcl-6, CD30, and CD23.

Cytological examinations show predominantly small to intermediately sized slightly cleaved B cells. The

Table 1: Immunohistochemical differential diagnosis of different low-grade non-Hodgkin's lymphomas by the commonest immunophenotyping according to Swerdlow and Williams[8]

Immunological markers	Small lymphocytic lymphoma	Marginal zone lymphoma	Follicular lymphoma	Mantle cell lymphoma
CD5	+	−	−	+(79−96%)[3,4]
CD10	−	−	+	−[5]
CD20	+	+	+	+
CD23	+	−	−	−(86−100%)[6]
CD43	+	−	−	+(91%)[7]
CD44	+	−	−	+[7]
Cyclin D1	−	−	−	+[5]
Cytogenetics				t(11;14)(q13;q32)[4]

genomic analysis typically demonstrates a chromosomal translocation t(11;14)(q13;q32) with a CCND1/IGH fusion.[4] This translocation implicates the cyclinD1gene with an over-expression of cyclinD1 in the pathogenesis of MCL.

The aim of this paper was to review the important clinical and pathological features of MCL with a clinical obvious manifestation in the oral cavity. It assembles the important histological findings that are necessary to give an accurate diagnosis for the right therapeutically features and the posttherapeutic outcome.

CASE REPORT

A 41-year-old man with no medical history presented himself to the department of Oral Maxillofacial Plastic Surgery of the University Halle–Wittenberg with a symmetric, painless swelling in the palatal molar region (10/2008). He reported the first appearance 10–12 weeks ago and a slow growth over this time [Figure 1a]. Other clinical symptoms such as dysphagy or odynophagy were not noticed. Familiy history was unremarkable.

The local examination of the palate showed a 1.5 × 3 cm swelling on both sides of the hard palate in the molar to premolar region with symmetrical appearance. This mass was covered by nonulcerated mucosa [Figure 1a] and had a firm and elastic consistence. No palpable cervical lymph nodes were present. The serum biochemistry values including the LDH (3.95 µmol/l) and the albumin serum level (61%) were normal.

The polymerase chain reaction (PCR) for hepatitis A, B, and C virus infection was negative.

During surgery, no involvement of the palatal bone was seen. The postoperative primary wound healing was delayed over 4–5 weeks.

The histological examination showed a population of small lymphocytes with slightly irregular intended nuclei and moderately dispersed chromatin (HE-staining, Figure 2a and b). Giemsa-staining [Figure 2c] and PAS-reaction detected a follicular pattern in the glandular tissue of the palate.

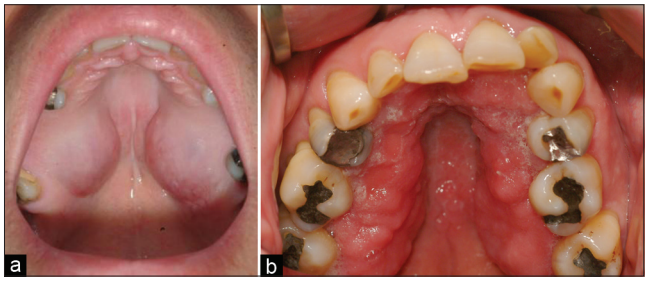

Figure 1: Clinical appearance of two different symmetric palatal tumor. The MCL (a) and the palatal epithelial hyperplasia (b) show both a symmetric appearance. They have a tight elastic consistence and are covered with a normal mucosa. Similarity in clinical appearance demands a patho-histological analysis for correct diagnosis

Immunohistochemical analyses showed a clear positive reaction for CD20 [Figure 2d], CD43 [Figure 2e] and the proliferation marker Ki67 [Figure 2f].[3]

The most significant reference for the MCL was the strong over-expression of cyclinD1 [Figure 2g].[3] Although Aguilera *et al*.,[5] described a positive reaction in 79–96% of the analyzed MCL cases for Leu-1 (CD5), only a weak expression was seen in the immunochemical examinations of the proliferating cells in this case. Immonochemical staining showed almost no expression of the kappa- and lambda-light chains and no expression for EMA, CD10, bcl-6, CD30, and CD23. There was no evidence of any blastic variant, such as centro- or immunoblasts, in tumor cells.

The hallmark for MCL: The chromosomal translocation t(11;14)(q13;q32) revealed many copies and in cytogenitical analyses the typical amplification of the CCND1/IGH fusion gene was found.[4]

After the histological analysis confirmed the diagnosis of a MCL the staging procedures (computed tomography, abdominal sonography) showed a disseminated disease at stage Va with axillary, pulmonal, iliac, inguinal, and liver manifestation.

The patient underwent chemotherapy according to the European MCL-Younger Study (www.lymphome.de/Projekte/MCL/Protokolle.jsp) and radiotherapy with a whole body-dose of 10 Gy (fractions of 2 Gy five times a day).

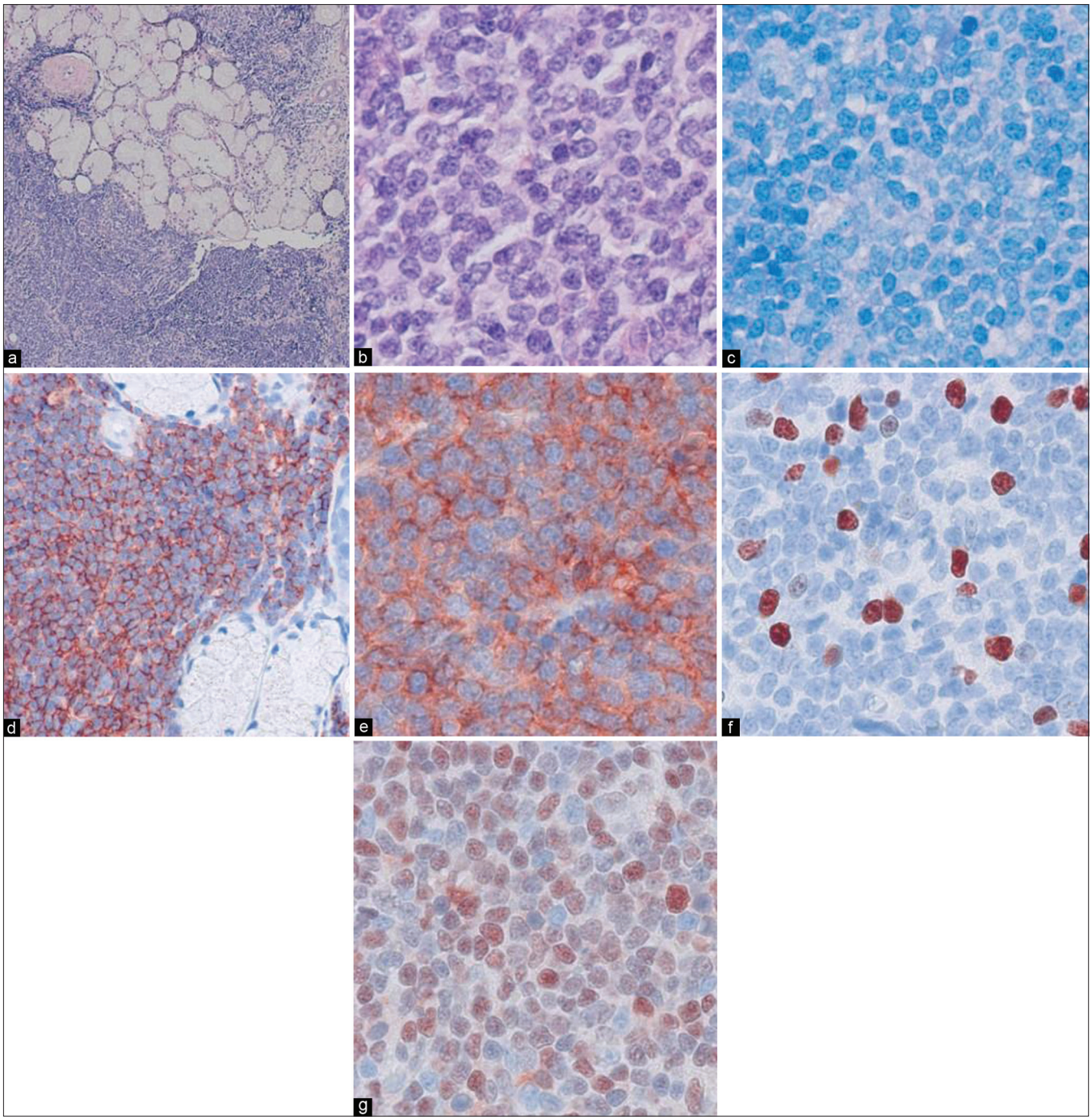

Figure 2: Histological findings. The overview in H.E. (a, b) and Giemsa-stain (c) detected the typical follicular growth pattern of the lymphocytic tumor. Immunohistochemical-staining showed a positive reaction for the proliferation marker Ki67 (f), the CD43 (e) the CD20-antigen (d) and the typical cyclinD1-expression in the tumor (g)

The therapy showed a clear regression of the MCL and the patient is still alive till 48 months after diagnosis without any detection of lymphoma-manifestation.

DISCUSSION

The high incidence of dento-alveolar abscesses and inflammations leads many dental surgeons to associate a swelling or tumor formation of the maxilla, palate, or lower jaw with these usual and common clinical conditions. In this case, the slow growth, the symmetric palatal appearance, and the normal covering mucosa [Figure 1a] led to the clinical diagnosis of a benign tumor formation such as hyperplasia [Figure 1b] or a lipoma and an excision with histological examination was demanded.

The diagnosis of a MCL within the oral cavity was made after histological and immunohistochemical analysis.

The wide group of NHL is the second-most common malignancy presenting within the head and neck region by painless cervical lymph node swelling.[1] For the subgroup, the MCL, the localization in the head and neck area is unusual and only a few cases with primary manifestation in the oral cavity can be found. Guggisberg *et al*.,[2] were able add two new cases of MCL appearing within the oral cavity to the 7 in English-literature presented cases since 1980 and assembled the clinical features of these orally manifested MCL.

The case reported here has lot in common with the nine earlier reported cases, but shows some differences in histology, clinical presentation, and outcome.

The majority of MCL's is predominant in male (male: female = 3:1,[2,9]) while the patients with a primary oral manifestation does not show this gender-related distribution (ratio male: female = 5:4,[2]). The MCL within the oral cavity were usually found in elderly patients (median age at 71 years) with a preferred localization on the palate (four of nine cases) like in this case.[2] But until now, the youngest patient reported within this clinical feature of an oral manifested MCL was 62 years.[2]

Respecting all case reports of an oral manifested MCL the stadium of disease at the beginning of therapy is not carefully documented. In general, it is known that the most patients with a MCL present at an advanced stage of disease (95% in Stage III–IV,[9]) and an extra-nodal advancement can be found in 92% of the patients.[8,9] For the MCL manifested in the oral cavity only three carefully documented patients could be identified in all published cases. According to the findings of the MCL patients, two of these patients with orally manifested MCL presented at a clinical stage IV/IVa and one at stage Ia.[2] The 41-years-old patient described here presented in an advanced stage IVa of the systemic disease, without any other clinical symptom.

Since the diagnostic criteria of MCL have been defined there is less known about the clinical characteristics and outcome of the patients.[1] According to the findings of Swerdlow and Williams,[8] the patient described here had a good prognostic feature by presenting a nonblastic and histological typical variant of a MCL. In all described MCL within the oral cavity, the diffuse classic variant was dominant.[2] This kind of MCL seems to have a better behavior and an advanced survival-period with 48 month after chemo- and radiotherapy.[6,9]

The presence of the translocation t(11;14)(q13;q32) and the over-expression of cyclinD1 does not have any impact on patient survival and prognosis and is only a good diagnostic criteria.[4,7]

A low LDH level at the time of therapy and the age under 65 years were found to be a good prognostic factor.[6,10] Other clinical studies of Norton *et al*.,[10] showed that the age over 70 years, a low albumin serum level, splenomegaly and an advanced stage of disease were associated with a short survival time, even if there was a nodular growth pattern in the histological examination, that actually would be a good factor.

This case demonstrates different behavior of an MCL within the oral cavity in a younger patient. The patient presented all prognostic factors that lead to a good survival: A low LDH-level, a normal serum albumin level and a histological not-blastic but classic variant of the MCL and an age under 65 years. Compared to the prior described cases of MCL within the oral cavity he shows good outcome with 48 month of remission.

So far, it was only unusual that the CD5-expression in our case was weak, while the CD5-expression is described to be typically beside the expression of CD20 and CD43. The negative reactions with CD23, CD10 were even typical and led to the right diagnosis.[6,10]

All in all the palate is a very rare location for the clinical manifestation of a MCL. The symmetrical appearance and the age of the patient are not very common. Even here, the right diagnosis was delayed by a wrong interpretation of the palatal swelling that could have been accelerated by a probe-excision and histological examination. An early diagnosis is necessary to help the patient to overcome the many challenges of this malignant disease.

The survival-period of 48 months after diagnosis was certainly associated with good prognostic factors [Figure 2].

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