MALT lymphoma of initial gastric site relapsing as simultaneous bilateral palpebral and salivary glands localizations

Rechute simultanée au niveau des deux paupières supérieures et des glandes salivaires d’un lymphome du MALT de site initial gastrique

Case report

A 67-year-old male patient presented with a bilateral upper palpebral ptosis associated with infiltration of external parts of superior eyelids (figure 1a). This symptom appeared gradually over a few months. There was a past history of gastric *Helicobacter pylori* positive MALT non-Hodgkin lymphoma diagnosed 9 years ago. This man had received chlorambucil as first-line therapy and thereafter four cycles of rituximab and gastric irradiation (30 grays) for two relapses which had occurred respectively after 6 and 8 years of clinical course.

His general condition was excellent (PS-ECOG = 0). At the physical examination, no lymphadenopathy was detected nor hepatomegaly. There were small nodules at the buccal mucosa surface. The hemogram showed 3.3 giga/L leukocytes with 71% neutrophils and 15% lymphocytes, 131 g/L hemoglobin and 177 giga/L platelets. LDH and CRP levels were normal. An IgM monoclonal gammopathy at 28 g/L was seen on the serum electrophoresis. At the CT-scan both right and left lacrimal glands were enlarged (35 mm diameter) but no abnormality was noted in the thorax or in the abdomen. Histological examination of eyelid and mouth biopsies disclosed MALT lymphoma invading the lacrimal and minor salivary glands respectively. The trephine bone marrow biopsy did not detect any abnormality.

This patient was treated with six courses of R-CVP (rituximab, cyclophosphamide, vincristine and prednisone) which induced a rapid disappearance of both mouth and palpebral tumours (figure 1b). About 2 years after the end of this treatment, he remains free of detectable disease.

Discussion

MALT lymphomas are indolent lymphoid neoplasms which may develop in various sites, the stomach being the most frequent. They are thought to be related to B-cells demonstrating excessive response to chronic infections or autoimmune processes [1]. This case is interesting because of the synchronous occurrence of two relatively rare localizations: the eyelid and minor salivary glands [2,3]. The eyelid accounts for only about 3% of non-gastrointestinal sites of MALT lymphomas [2] and such a symmetrical bilateral palpebral localization has been rarely described. The aspect of eyelids shown here (figure 1a) resembles that reported...
by Shikino et al. [4]. This type of presentation may mimic sarcoidosis, a granulomatous inflammation of still uncertain pathophysiology [5]. In the largest series of MALT lymphomas of the salivary glands published so far including 63 patients from 10 different countries, minor salivary glands were very uncommonly involved (3 cases) in comparison with parotid (49 cases) or submandibular (15 cases) glands [3]. A good prognosis has been associated with these unusual lymphoma sites using adapted therapeutic approaches [2,3]. Here, in this multisite relapsing patient, we obtained a rapid, complete and still ongoing response using a chemoimmunotherapy regimen including rituximab.

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References


