CASE REPORT

RJME
Romanian Journal of Morphology & Embryology
http://www.rjme.ro/

Non-Hodgkin's lymphoma, rare localization at the base of the tongue – case presentation

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Abstract

The extranodal malignant lymphomas occur mainly in the region of the head and neck, after the gastrointestinal tract. The most common site of the head and neck lymphoma is at the lymphatics ring of Waldeyer, followed by the nasosinusal region. Localization at the base of the tongue is extremely rare. The authors reported a case of a 50-year-old male with a non-Hodgkin's lymphoma, which developed at the base of tongue. An unusual sensation described as a foreign body at the hypopharyngeal region associated with progressive dysphagia were the main symptoms for which the patient was referred to our Hospital. The fibroscopic exam has revealed an oval tumor of the base of tongue, with large dimensions, sessile emergent base and smooth superficial mucosa. The tumor was removed using an endoscopic transoral approach, followed by the histopathological and immunohistochemical examination, which have been suggestive for non-Hodgkin's lymphoma with small B-cells. The treatment continued with chemotherapy, while radiotherapy was not necessary due to the fact that the tumor was completely removed.

Keywords: non-Hodgkin's lymphoma, base of tongue, extranodal lymphoma.

Introduction

Lymphomas are malignant tumors that develop from lymphocytes. There are two main categories of lymphoma: Hodgkin's lymphoma (HL) and non-Hodgkin's lymphoma (NHL). The most frequent site of this kind of tumor is the lymphatic nodes or spleen, but there are some extra-nodal forms. The extranodal malignant lymphoma occurs mainly in the gastrointestinal tract and then in the region of the head and neck [1]. Head and neck lymphomas are the second malignant tumors as a frequency after carcinomas [2]. The most common site of the head and neck lymphoma is at the lymphatics ring of Waldeyer, followed by the nasosinusal region [1]. From the lymphatic ring of Waldeyer, the palatine tonsil is the mainly site for the non-Hodgkin's lymphoma. The base of tongue is extremely rare affected by this type of malignant tumor, being more common in man than in women, with the main age of appearance being 59-year-old [3, 4]. Until now, the reported cases of non-Hodgkin's lymphoma of the base of tongue have appeared under the clinical form of ulcerative lesions associated with latero-cervical lymphadenopathy [5]. Therefore, the aim of this paper is to report a particular case of non-Hodgkin's malignant lymphoma due to its extremely rare tumor site, endoscopic aspect, therapeutic protocol (endoscopic removal of the entire tumor through transoral approach) and its immunohistochemical assessment.

Case presentation

After signing the informed consent regarding the treatment and publication of medical data, we reported the case of S.A., a 50-year-old male, which was admitted in the Department of ENT (Ear, Nose and Throat) at "Elias" University Emergency Hospital, Bucharest, Romania, in September 2016 (Chart No. 37623), for an unusual sensation described as a foreign body at the hypopharyngeal region associated with dysphagia. The symptoms have evolved during the last three months. The patient was a nonsmoker, without dyspnea or dysphonia. His medical history revealed no other significant pathologies. The clinical exam was normal and so were the results of the routine blood test (complete blood count, coagulation, liver transaminases, urea and creatinine tests).

The fibroscopic examination of the pharynx showed the presence of a large tumor at the right side of the base of tongue, with large emergent base, significant amputation of the pharynx lumen, but with smooth superficial mucosa (Figure 1). At palpation, the tumor had a solid consistency.

The palpation of the lymphatic nodes did not revealed any adenopathies, the rest of the clinical and otorhinolaryngology examination being normal.

Magnetic resonance imaging (MRI) examination of the cervical region has revealed the presence of a solid tumor, with sessile emergent base at the right side of the base of tongue, measuring 25/20/15 mm, which was causing significant amputation of the pharynx lumen (Figures 2–4).

After the approval of the Local Ethics Committee of the "Elias" University Emergency Hospital, Bucharest, regarding the treatment protocol, the patient underwent surgical excision of the tumor through endoscopic transoral approach, under general anesthesia with nasotracheal intubation (Figure 5). The tumor's resection was made with the electrocautery. After the tumor was removed, the intraoperative aspect of the surgical site reveals a large tumor implantation base (Figure 6).

Postoperatively, the evolution was favorable; the patient resumed oral feeding next day, initially with liquid and semisolid foods. The patient was dismissed after three days.

The histological assessment of the surgical specimen showed palatine tonsils tissue associated with malignant lymphoid proliferation with nodular pattern, consisting of small cells, polymorphous, with rounded or incised nucleus, pulverulent chromatin and isolated histiocytes (Figures 7–9).

The immunohistochemical assessment revealed that the tumor proliferation is suggestive for non-Hodgkin's lymphoma with small B-cells, strongly positive for CD20 (L26, Immunologic®, 1:1000 – Figure 10), expressing cyclin D1 (EP12, Dako®, 1:100) and SOX11 markers (MRQ-58, Cell Marque®, 1:50) (Figure 11), with low Ki-67 proliferation index <10% (MIB1, Immunologic®, 1:500) (Figure 12). The tumor proliferation is strongly negative for CD10 (56C6, Leica®, 1:100) and Bcl-6 markers (LN22, Leica®, 1:50) and also negative for CD5 (4C7, Leica®, 1:100) (Figure 13).

The histopathological and immunohistochemical assessment concluded that the basilingual tumor is a rare form of non-Hodgkin's mantle cell B-lymphoma, with nodular pattern of growth and negative CD5 staining.

Postoperatively, a positron emission tomography–computed tomography (PET–CT) scan was performed which confirmed the total tumor resection and the absence of secondary sites. The patient underwent chemotherapy, being treated with four cycles of Cyclophosphamide, Adriamycin, Vincristine and Prednisone. During chemotherapy, the patient developed bone marrow aplasia, therefore, a bone marrow transplant was required. After seven months, the patient was presenting without any tumor recurrence.

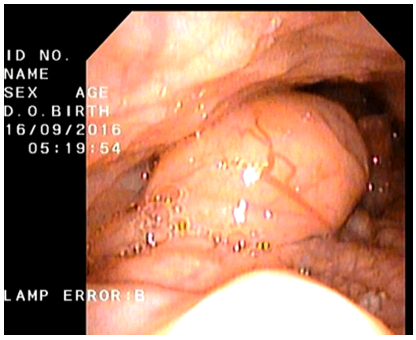

Figure 1 – ***The video-fibroscopy reveals a tumor with normal aspect of the mucosa and a large insertion on the right hemibase of the tongue, which causes the partial obstruction of the laryngeal inlet.***

Figure 2 – ***MRI aspect of the round-shaped well-demarked basilingual tumor.***

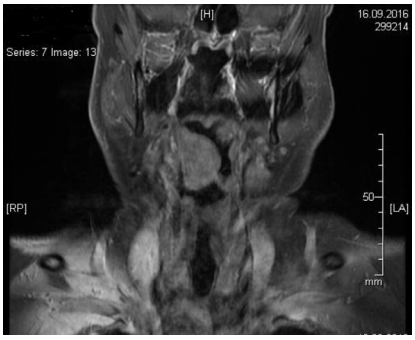

Figure 3 – ***MRI aspect of the tumor, which lies between the palatine tonsils and the lingual surface of the epiglottis.***

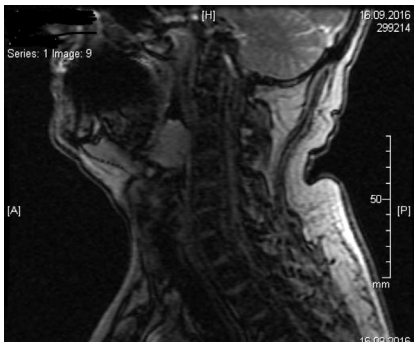

Figure 4 – ***MRI aspect revealing the anterior limit of the tumor represented by the tongue's base and the posterior limit represented by the close contact without infiltrating the posterior wall of the oropharynx and hypopharynx.***

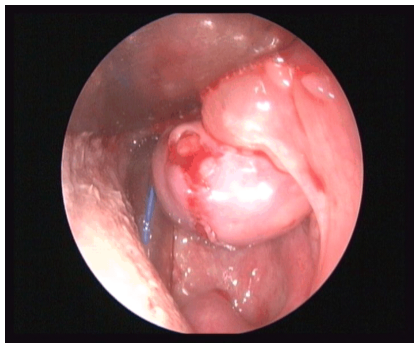

Figure 5 – ***Intraoperative endoscopic aspect at 0° angle, which reveals the entire tumor after pushing the base of the tongue and the intubation nasal cannula behind the uvula, which facilitated this surgical approach.***

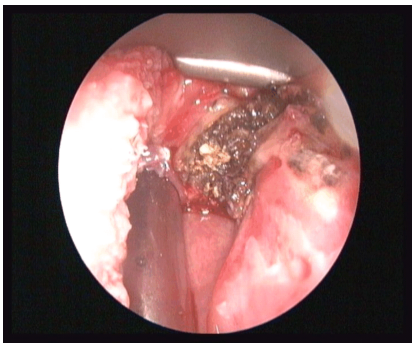

Figure 6 – ***Intraoperative endoscopic aspect of the cauterized large tumor site after removing it using CO_2 laser.***

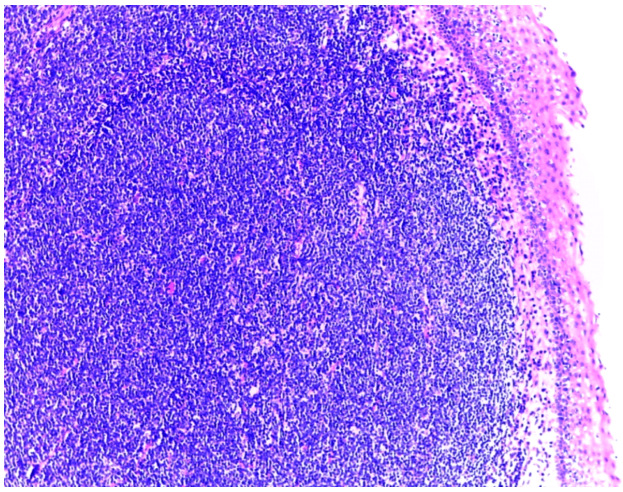

Figure 7 – ***Lingual tonsil reveling malignant lymphoid diffuse proliferation (HE staining, ×100).***

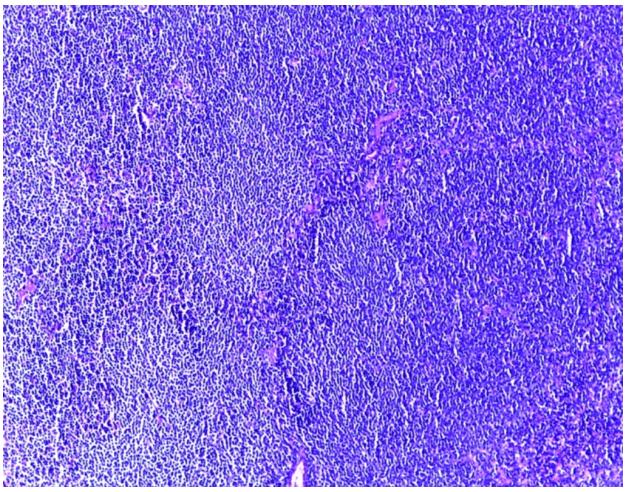

Figure 8 – ***Areas with nodular pattern of the malignant lymphoid diffuse proliferation (HE staining, ×100).***

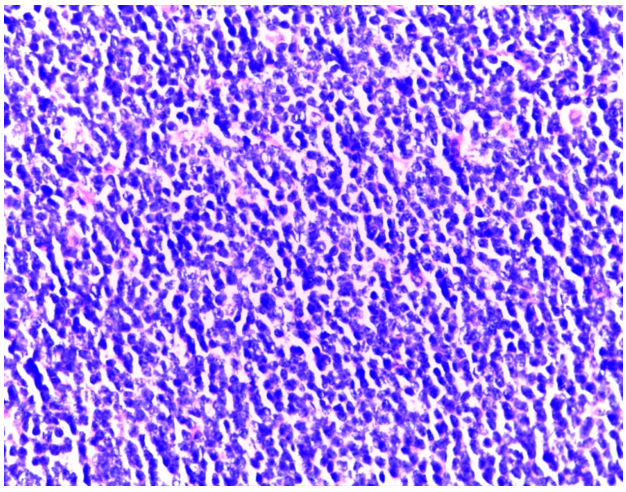

Figure 9 – ***Pleomorphic small cell proliferation with round or cleaved nuclei and hardly visible nucleoli (HE staining, ×400).***

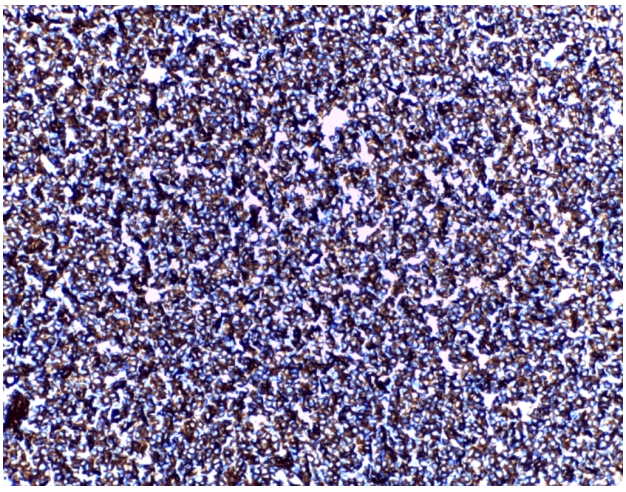

Figure 10 – ***Positive staining for CD20 immunohistochemistry (×200) in B cells.***

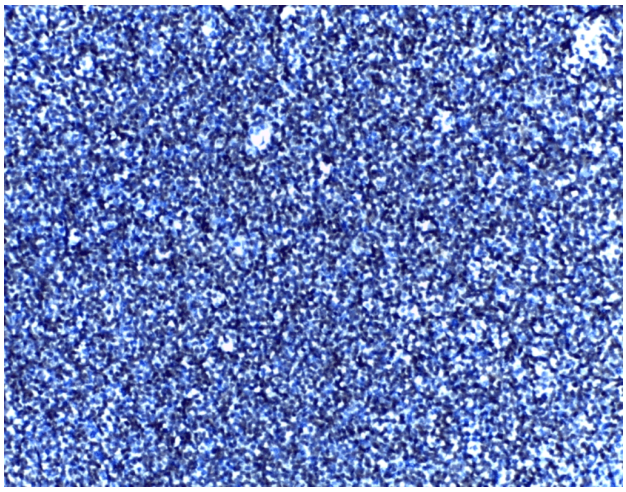

Figure 11 – ***Positive staining for cyclin D1 immunohistochemistry (×200) in tumor cells which is a marker for mantle B-cell lymphoma.***

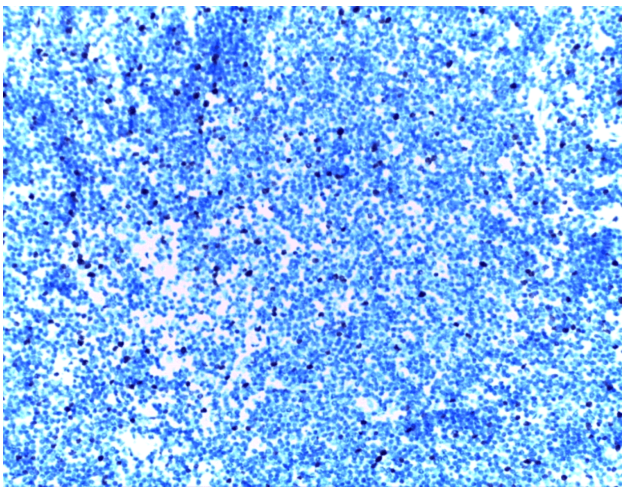

Figure 12 – ***Positive staining for Ki-67 immunohistochemistry (×200) with a low Ki-67 index (less than 10%).***

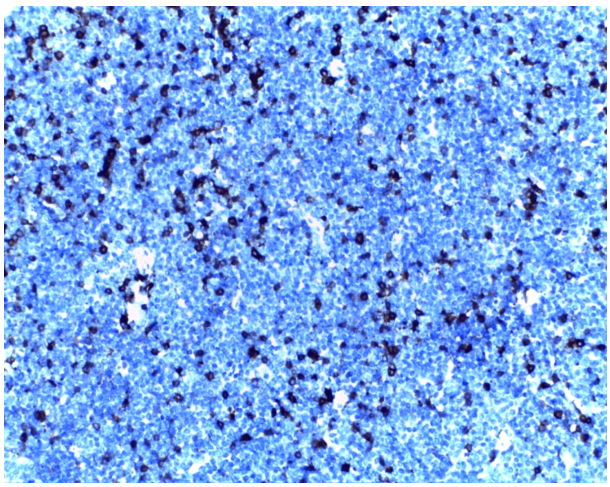

Figure 13 – ***Negative staining for CD5 immunohistochemistry (×200) in tumor cells revealing the absence of T cells.***

Discussion

Lymphomas represent a group of malignant solid tumors that develop from proliferation of B- and T-lymphocytes. The most frequent sites for this tumors are the lymphatics nodes, but it has been reported a lot of cases with extranodular sites, in the recent years. Tongue and kidney sites are some of the most rare involved sites, renal lymphomas being usually associated with kidney disease, which requires hemodialysis [6–8]. According to some authors, 30% of the lymphomas are extranodal, the majority being non-Hodgkin's lymphomas [5]. Among these, lymphoma with small B-cells is associated with a much more aggressive behavior, consisting of increased tendency to local and regional dissemination [9–11].

The etiology of lymphomas is not well known. However, the chromosomal mutations, the infection with Epstein–Barr virus, the human herpesvirus 8 (HHV 8), the retrovirus and the human immunodeficiency virus (HIV) remain the most important risk factors [12, 13].

The malignant lymphomas are the most frequent non-epithelial neoplasia located in the region of the head and neck [14]. The lymphatics ring of Waldeyer, especially the palatine tonsil, are the sites most usually involved for this type of tumors [14]. Recently, Turner & Zitsch have reported a case of oropharyngeal lymphoma, which was clinically presenting significant bleeding [15].

The diagnosis of the basilingual lymphomas requires endoscopic examination of the region. In most of the cases, this are identified as tumoral masses with a smooth superficial mucosa, but there are some reported cases with ulcerations of the mucosa, thus being confused with carcinoma. The CT scans of the cranial, cervical, thoracic, abdominal and pelvic region are useful in order to establish the full classification of the tumor. To assess the response of the treatment, the PET–CT scan is preferred [16].

The diagnosis of certainty is established through biopsy followed by histopathological and immunohistochemical assessment. The last one is mandatory to identify the phenotype of the mantle cell lymphoma, which is usually positive for CD5, CD19, CD20, cyclin D1 and Bcl-1 and negative for CD23, CD10, Bcl-6, TdT and CD11C [17]. In our case, strong positive expression of CD20, cyclin D1 and SOX11 associated with negative expression of CD10 and Bcl-6 supported the diagnosis. However, negative expression of CD5 (which a T marker) classified this mantle cell lymphoma as an aberrant phenotype. Other aberrant phenotypes are associated with positive expression of CD10 and Bcl-6 or negative expression of cyclin D1 [17]. Low Ki-67 proliferation index supports the less aggressive behavior of this tumor, even if it resembled some histological aspects of the pleomorphic aggressive variant (polymorphous cells, irregular or incised nuclear contours, pale chromatin) [18, 19].

The differential diagnosis should be made with other tumors located at the base of the tongue: metastasis, melanomas, low differentiated squamous cell carcinoma or rare tumors like neuroblastoma, rhabdomyosarcoma or Ewing's sarcoma [18, 19]. After identifying the histological aspects of lymphoma, immunohistochemical assessment is performed in order to classify the tumor. In our case, positive expression of cyclin D1 and negative expression of CD10 excluded marginal zone lymphoma (CD5-, CD10-, cyclin D1-) and follicular lymphoma (CD5-, CD10+, cyclin D1-) [17, 18], while strong expression of CD20 and negative expression of CD5 excluded chronic lymphocytic leukemia and small lymphocytic lymphoma (cyclin D1-, low expression of CD20, CD5+, CD23+, p27 negative) [18, 19]. Even if positive expression of cyclin D1+ may be encountered in hairy cell lymphoma and myeloma, the histological aspects in this case supported the mantle cell lymphoma diagnosis [17–19].

The absence of metastasis, the localization in a singular extranodal site and the age of the patient less than 60 years old influenced positively the prognosis of our patient.

The therapeutic protocol for the extranodal lymphomas consists in chemotherapy followed by radiotherapy of the affected zone [20]. The resection of the tumor being complete, the multidisciplinary therapeutic team decided that the radiotherapy is no longer necessary; therefore, the patient was treated with four cycles of chemotherapy.

Conclusions

The basilingual malignant lymphomas are very rare, but it should be considered whenever a tumoral mass is identified in this area. The diagnosis of certainty is established only by histopathological and immunohistochemical assessment. The outcome is better for the localized forms and for the patients younger than 60 years old. Although the complete resection of the tumor is not included in the standard protocol, in this case, using this technique has improved the quality of life for our patient, because the radiotherapy was no longer necessary along with its negative effects.

Conflict of interests

There is no conflict of interests.

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Received: March 5, 2017

Accepted: August 4, 2017