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Chłoniak strefy brzeżnej krtani. Problemy diagnostyczne – opis przypadku

B-Cell marginal zone lymphoma of the larynx. Diagnostic difficulties – case report

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STRESZCZENIE: Chłoniak strefy brzeżnej krtani należy do grupy wolno rosnących, nie-Hodgkinowskich chłoniaków wywodzących się z limfocytów typu B. Umiejscowienie tego nowotworu w krtani zdarza się niezwykle rzadko. Pierwszy raz chłoniak strefy brzeżnej krtani został opisany w 1990 roku [2], a od tego czasu w literaturze zostało opisanych mniej niż 50 przypadków tego nowotworu [1]. W pracy opisujemy przypadek 66-letniej kobiety przyjętej w kwietniu 2012 roku na oddział otorynolaryngologiczny w Specjalistycznym Centrum Medycznym w Polanicy-Zdroju w celu wykonania biopsji guza krtani. Pacjentka skarżyła się na chrypkę od 2 lat i narastającą duszność. Historia wskazywała również na nawracające epizody zapalenia krtani i tchawicy. Histopatologiczna ocena wycinka pobranego z krtani wykazała chłoniaka strefy brzeżnej. Pacjentka została skierowana na oddział hematologiczny, gdzie otrzymała leczenie chemioterapeutyczne, które zakończyło się całkowitą remisją. Biopsja wykonana w kwietniu 2012 roku na naszym oddziale była już trzecią z kolei biopsją zmiany. W 2010 roku na innym klinicznym oddziale otorynolaryngologicznym w Polsce zmiana była poddawana histopatologicznej ewaluacji dwukrotnie. Wynik badania w obu wcześniejszych przypadkach wskazywał na zmianę zapalną, za każdym razem wskazywano również jednak na trudność oceny materiału. Opis przypadku, który prezentujemy, wskazuje na problemy diagnostyczne w rozpoznawaniu chłoniaka strefy brzeżnej w obrębie krtani.

SŁOWA KLUCZOWE: guz krtani, pozawęzłowy chłoniak, MALT

ABSTRACT: Marginal zone lymphomas are a group of slow-growing non-Hodgkin B-cell lymphomas. This type of lymphoid tissue tumors occur extremely rare in larynx. Since the first description by Diebold et al in 1990, less than 50 cases marginal zone lymphomas of the larynx have been reported [1, 2]. We present a case report of a 66-year-old woman, admitted in April 2012 to an ENT Ward in Polanica Zdroj hospital in order to perform a biopsy of lesion present in larynx since at least 2 years. Patient presented with hoarseness, mounting dyspnoea and history of recurrent inflammation of larynx and trachea. Histopathologic examination revealed B-Cell Marginal Zone Lymphoma. The patient was sent to undergo further hematological treatment (R-CHOP regimen chemotherapy), which was followed by a complete remission. It is important to emphasize that it was the 3rd biopsy of the lesion taken in 2 years time (since 2010) preceded by two such procedures in another clinical ENT Ward in Poland. The microscopic examination results of the previous biopsies were described as indicative more of an inflammatory lesion although both of them were said to be hard to evaluate. This shows the diagnostic difficulties that biopsy itself or its microscopic interpretation might bring.

KEY WORDS: laryngeal tumor, extranodal lymphoma, MALT

INTRODUCTION

Marginal zone lymphoma is a type of extranodal non-Hodgkin B Cell Lymphoma also called MALToma (MALT stands for Mucosa Associated Lymphoid Tissue). The most common location of marginal zone lymphoma in larynx for the already described cases is supraglottic [4]. Majority of the malignant tumors of the larynx are squamous cell carcinomas (90%) while malignant hematopoietic tumors of the larynx are estimated to account for 1% of laryngeal malignant tumors [3]. This particular case of laryngeal marginal zone lymphoma underlines diagnostic difficulties and outlines the morphological features that might suggest hematopoietic malignant tumor of the larynx.

CASE REPORT

A 66-year old female LL was admitted to the ENT Ward in Polanica Zdroj on April 3rd 2012 in order to perform further diagnostic of laryngeal tumor. CT scan showed right lateral supraglottic tumor (2,7 cm x 2.7 cm x 4,0 cm) obstructing the lumen of the pharynx and larynx, clamping the piriform fossa with no signs of metastatic infiltration of neighbouring tissues nor lymph nodes. The biopsy was taken for further microscopic evaluation. The histopathological specimen was described as lymphoid tissue predominantly made up of small B-cells (CD 20 > CD3, CD5). The BCL-2 mutation was positive. Biopsy sample was described as characteristic to marginal zone lymphoma.

The patient was immediately referred to hematologist. R-CHOP regimen chemotherapy (Rituximab, Cyclophosphamide, Vincristine, Prednisolone) was initiated. A PET-CT scan performed before chemotherapy showed an increased glucose metabolism in the region of laryngeal tumor, tonsils, adenoid and the cervical lymph nodes (CIII CIV). First dose of chemotherapy was administered on June 6th 2012, the last one on November 9th 2012. One month later a control PET-CT was performed revealed a complete remission of the lesions. Another PET-CT scan dated November 18th 2014 confirmed complete remission. ENT control examination performed on March 27th 2015 showed no abnormalities in the region of the larynx.

It is important to emphasise that the tumor was only correctly classified during the 3rd biopsy. The patient LL was first admitted in June 2010 to an ENT ward in Polish clinical hospital presenting persistent hoarseness since 5 months. Examination revealed a laryngeal tumor. Lesion was then biopsied twice, with both histopathological evaluations describing the material as non-representative with features pointing to rather an inflammatory lesion. In 2012, however, the problem intensified, as the patient started suffering from dysphonia and persistent

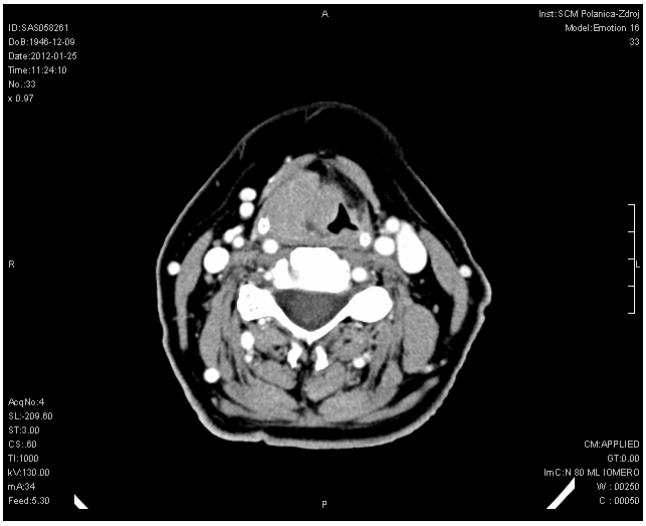

Fig.1 Mass of the tumor in the right glottic and supraglottic region extensively obturating the lumen of the larynx]

cough. In February 2012 patient was admitted to a hospital in Germany. The endoscopy revealed an inflammated trachea and larynx and persistent tumor present in larynx. An MR confirmed the presence of supraglottic tumor. The patient was treated at the German Clinic for laryngotracheitis with a 10-day treatment of Clindahexal and Decortin-H. Although the volume of the tumor was reduced by 1/3rd after the treatment, patient was advised to extend the examination, which led to the admittance to the ENT Ward in Polanica Zdroj.

DISCUSSION

B- Cell marginal zone lymphoma of the larynx is a rare kind of malignant tumor. It develops from MALT, which by some authors is described as specific laryngeal type of lymphoid tissue - LALT (Laryngeal Associated Lymphoid Tissue) [7].

Pathogenesis of B-Cell Marginal Lymphoma is considered to be associated, as in other MALT tumors, with either a chronic inflammation of the larynx (as it is in stomach – H.Pylori infection with chronic inflammation) or with an autoimmune process (as it is in salivary glands – with Sjogren's Syndrome). The 5-year survival rate of marginal zone lymphoma is high, exceeding 80% [6].

Although clinical manifestation is no different than other lesions in this regions there are some imaging characteristics which may indicate a lymphomatic type of the tumor. These are: the supraglottic localization and lack of calcification in the tumor. Abnormalities within the tissue are usually non-necrotic and larger in size which differs them from squamous cell carcinoma

[4]. Macroscopically these tumors present as smooth or polypoid, rather than ulcerated masses [5]. The main symptoms thus are universal for laryngeal tumors including dysphagia, dysphonia, dyspnoea and stridor. Our case report suggests that there are difficulties with either microscopical assessment of the biopsy or the technique of the biopsy itself. Awareness of the two factors can contribute to an earlier diagnosis: one must be aware of the diagnostic difficulties and be familiar with the characteristic morphology of the tumor's tissue.

CONCLUSION

Marginal zone lymphoma of the larynx is a rare malignant tumor with less than 50 cases reported during 25 years since the first description in 1990[2]. Moreover this is the first report of such tumor described in Poland. Complete remission in this case and high survival rate among patients with marginal zone lymphoma suggest that the challenge is not the management of the disease but the proper biopsy and its microscopic evaluation.

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