

CASE REPORT

## A case of mucosa-associated lymphoid tissue lymphoma of the parotid gland

Parotis bezinin mukoza ile ilişkili lenfoid doku lenfoması: Olgu sunumu

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Non-Hodgkin's lymphomas of the salivary glands are rare. Most of these tumors occur in the parotid gland. The most common subtype is extranodal B-cell lymphoma, which arises from mucosa-associated lymphoid tissue. A 63-year-old-woman with a painless swelling in the right parotid gland was examined with magnetic resonance imaging and fine-needle aspiration cytology. The mass was excised via superficial parotidectomy. Histological examination revealed a low-grade B-cell lymphoma originating from the lymphoid tissue associated with malignant mucosa. Lymphoma should be considered in the differential diagnosis in patients having a mass in the parotid gland and a systemic or autoimmune disease.

**Key Words:** Lymphoma, mucosa-associated lymphoid tissue/diagnosis/surgery; parotid neoplasms/diagnosis/surgery.

Tükürük bezlerinin non-Hodgkin lenfoması nadirdir. Bu tümörlerin çoğu parotis bezinde görülür. En sık görülen alttipi, mukoza ile ilişkili lenfoid dokudan köken alan, ektranodal B-hücreli lenfomadır. Sağ parotis bezinde ağrısız şişlik nedeniyle başvuran 63 yaşındaki bir kadın hasta manyetik rezonans görüntüleme ve ince iğne aspirasyon sitolojisi ile incelendi. Kitle yüzeysel parotidektomi ile çıkarıldı. Histolojik incelemede düşük evreli, malign mukoza ile ilişkili lenfoid doku kökenli B-hücreli lenfoma saptandı. Parotis bezinde şişlik ile başvuran, özellikle sistemik ya da otoimmün hastalığı olan olgularda ayırıcı tanıda lenfoma da göz önüne alınmalıdır.

**Anahtar Sözcükler:** Lenfoma, mukozayla ilgili lenfoid doku/tanı/cerrahi; parotis neoplazileri/tanı/cerrahi.

Lymphoma is classified in two categories, Hodgkin's lymphoma (HL) and non-Hodgkin's lymphoma (NHL), and both types arise from lymphoid tissues. The vast majority of lymphomas occur within the lymph nodes; however, these tumors can also present at extranodal sites. Approximately 25-30% of NHL cases and 1% of HL cases present as extranodal disease.<sup>[1]</sup> The majority of extranodal lymphomas

occur in the gastrointestinal tract, and the head and neck are the next most common sites.<sup>[2]</sup>

### CASE REPORT

A 63-year-old-woman presented with a painless mass in the right preauricular area. The lesion had been present for 2 years, but had grown in the 2 months prior to presentation. There was no history of

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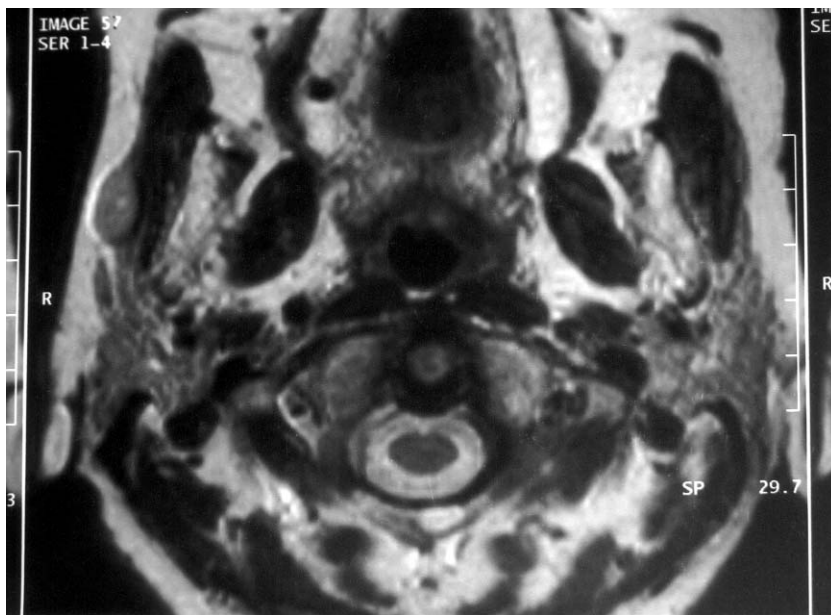
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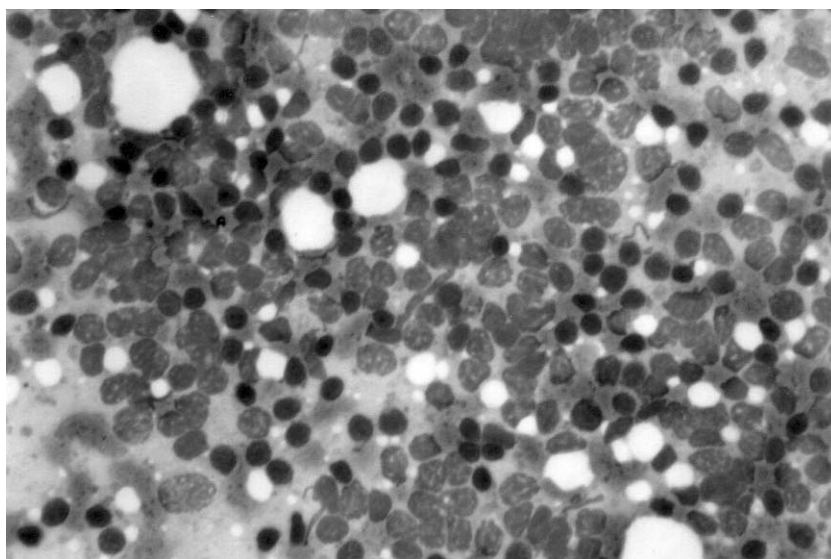
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systemic disease. Physical examination revealed a firm, fixed 1x1.7 cm mass located anterior to the right ear. No cervical lymphadenopathy was detected on neck palpation. Magnetic resonance imaging demonstrated a solid 17x10 mm mass anterior to the right parotid gland. The mass was located in the subcutaneous fatty tissue overlying the masseter muscle (Fig. 1). Fine-needle aspiration biopsy (FNAB) was performed, and cytological examination revealed large numbers of lymphoid cells and scant plasma cells and histiocytes (Fig. 2). Based on these findings, the two main diagnostic rule-outs were Sjögren's syndrome and mucosa-associated lymphoid tissue lymphoma.

Superficial parotidectomy was performed to remove the lesion. Macroscopically, the tumor was a smooth solid mass that measured 1.8x1x1 cm. On cut section, it was tan to white in color and appeared homogeneous. Histologically, there was marked infiltration of lymphoid cells within the parotid gland parenchyma (Fig. 3). The tumor was composed of fairly uniform, monomorphic, medium-sized lymphoid cells with pale nuclei and scant eosinophilic cytoplasm. Infiltrate filled the acini and ducts within the affected portion of the gland, and was also present in the adipose tissue and the interlobular and periglandular connective tissues. Immunohistochemical testing showed that the



*Fig. 1 - As seen in the axial T2W image above, the mass which is approximately 17 mm in size with smooth contour in the anterior of parotid gland shows isointens signal characteristic with right parotid gland.*



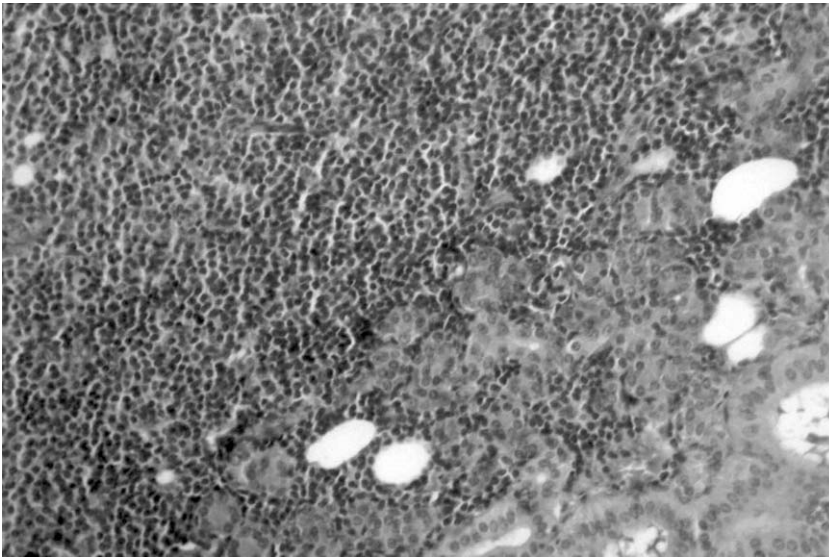
*Fig. 2 - A section of the tumor shows small to medium-sized cells with round nuclei and sparse cytoplasm (MGG x 200).*

tumor cells were positive for leukocyte common antigen (LCA; Klon CD45 Ab-3, Neomarkers) and CD20 (PAN-B; Klon L-26, DAKO) (Fig. 4), and negative for CD45 (PAN-T; Klon UCHL1, DAKO) The morphological and immunohistochemical findings were compatible with low-grade malignant mucosa-associated lymphoid tissue B-cell lymphoma. There were no problems in the postoperative period, and computed tomography (CT) scans of the abdomen and thorax revealed nothing abnormal. The patient was discharged and rechecked at monthly intervals.

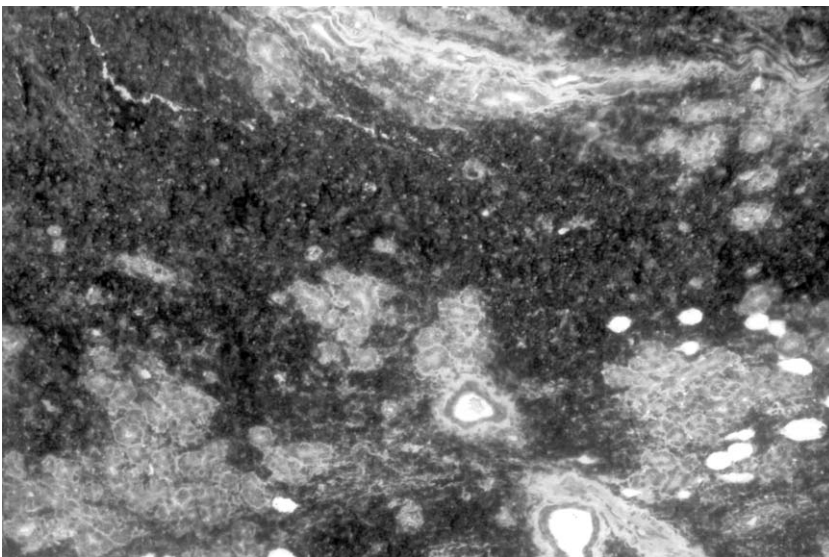
### DISCUSSION

Eleven to 33% of extranodal lymphomas occur in the head and neck area,<sup>[2]</sup> and more than 50% of

cases in this region arise from Waldeyer's ring.<sup>[3]</sup> Primary malignant lymphomas account for only 1.7-3.1% of all salivary gland neoplasms, and the parotid gland is the most common site of occurrence.<sup>[4]</sup> This category of lymphoma represents 3-4% of all parotid gland tumors.<sup>[5]</sup> Malignant lymphomas of the parotid gland may arise from mucosa-associated lymphoid tissue in the gland parenchyma and/or in intraglandular lymph nodes, or they may be disseminated from an existing lymphoma.<sup>[1]</sup> Malignant lymphomas linked with mucosa-associated lymphoid tissue were first described in 1983 as gastrointestinal tract lymphoma. Malignant mucosa-associated lymphoid tissue lymphomas are extranodal B-cell NHLs that are primarily low



**Fig. 3** - The severely atrophic gland tissue was infiltrated with large numbers of lymphoma cells featuring irregular nuclei and pale cytoplasm (H-E x 200).



**Fig. 4** - Immunoreactivity for CD20 in the lymphoid cells of the MALT lymphoma. (IHC, CD20 x 200).

grade.<sup>[6]</sup> In most cases, these tumors arise in combination with benign lymphoepithelial lesions or myoepithelial sialadenitis associated with Sjögren's syndrome.<sup>[7]</sup> Patients with this autoimmune disease are at increased risk (as high as 6% per year) for developing B-cell lymphomas, including mucosa-associated lymphoid tissue lymphomas.<sup>[8]</sup>

Sixty percent of low-grade lymphomas exhibit trisomy 3.<sup>[9]</sup> Most mucosa-associated lymphoid tissue lymphomas are indolent neoplasms that tend to remain localized for a long time, even without treatment; however, some disseminate or transform to a higher grade.<sup>[9]</sup> It has been suggested that Epstein-Barr virus or human herpesvirus 6 might be involved in inducing mucosa-associated lymphoid tissue lymphoma.<sup>[5]</sup> Interestingly, recent work has demonstrated an association between the chromosome translocation t(11;18) (q21;q21) and low-grade malignant mucosa-associated lymphoid tissue NHL, indicating that this abnormality might predict this tumor.<sup>[5]</sup> It has also been shown that mucosa-associated lymphoid tissue lymphomas lack rearrangements of the bcl-2 gene (t[14;18]).<sup>[5]</sup>

The gastrointestinal tract is the most common site of mucosa-associated lymphoid tissue lymphoma occurrence.<sup>[9]</sup> Of the 43 cases described by Berger et al.<sup>[10]</sup> 27 were in the gastrointestinal tract and the remaining 16 were in the orbit (n=5), parotid gland (n=3), lungs (n=3), tonsils (n=2), thyroid gland (n=2), and mammary glands (n=1). Takahashi et al.<sup>[11]</sup> investigated 10 cases of mucosa-associated lymphoid tissue lymphoma of the salivary glands. Seven of these cases involved the parotid gland, two involved the submandibular gland, and one involved the sublingual glands. Shi et al.<sup>[12]</sup> studied 32 cases of mucosa-associated lymphoid tissue lymphoma of the salivary glands, 17 in the parotid gland and 15 in the submandibular gland. Hacıhanefioglu et al.<sup>[13]</sup> reported two cases of mucosa associated lymphoid tissue lymphoma in salivary and lacrymal gland.

The reported ages of patients with mucosa-associated lymphoid tissue lymphoma range from 29-53 years, and there is a strong bias towards females (ratio 6:1).<sup>[10]</sup> The most common symptom is painless swelling of the parotid gland,<sup>[10]</sup> and 30% of patients also show features of sicca syndrome.<sup>[11]</sup> Our patient was a 63-year-old woman with no systemic disease who had a painless swelling in her right parotid gland.

Mucosa-associated lymphoid tissue lymphomas are diagnosed based on histological examination of a biopsy specimen. Microscopic examination reveals B-cells surrounding lymphoid follicles and selectively infiltrating the epithelium to form the characteristic lymphoepithelial lesion. Characteristic mucosa-associated lymphoid tissue lymphoma cells are small to medium-sized and contain a moderate amount of cytoplasm. The cytoplasm sometimes stains pale, and the nuclei are typically irregular, resembling those of centrocytes (small cleaved cells).<sup>[6]</sup> Fine-needle aspiration is not an ideal diagnostic tool because it is difficult to diagnose lymphoma on cytological grounds alone. The preferred approach is to obtain an incisional biopsy using facial nerve monitoring, or to perform parotidectomy and evaluate the specimen histologically.<sup>[9]</sup> In our case, we performed superficial parotidectomy, and the histopathological diagnosis was mucosa-associated lymphoid tissue lymphoma.

The treatment in these cases depends on the stage of the disease. Mucosa-associated lymphoid tissue lymphomas of the parotid gland are staged according to the Ann Arbor classification.<sup>[10]</sup> In stages 1 and 2, the recommended therapy is curative surgery or radiotherapy, whereas in stages 3 and 4 chemotherapy is required.<sup>[10]</sup> However, the optimal treatment for low-grade (stage 1 or 2) mucosa-associated lymphoid tissue derived NHLs of the salivary glands remains unclear.<sup>[14]</sup> Since our patient was categorized stage 1, we performed superficial parotidectomy and followed her with monthly rechecks.

This case is of interest because it is rare to diagnose mucosa-associated lymphoid tissue derived parotid gland lymphoma. When a patient presents with unilateral or bilateral parotid swelling, it is important to be able to differentiate lymphoma from the more common parotid tumors. The possibility of mucosa-associated lymphoid tissue lymphoma should always be considered in such cases, particularly if the individual has systemic or autoimmune disease.

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