



## Mucosa Associated Lymphoid Tissue Lymphoma (Maltoma) of Parotid Gland

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### Abstract

Mucosa-associated lymphoid tissue (MALT) is a non-encapsulated cluster of lymphocytes that is thought to have an important role in mucosal immunity. Maltoma is an extranodal lymphoma either of B- or T-cell origin that may most commonly involve the stomach, followed by other gastrointestinal organs, respiratory tract, skin and others containing lymphoid tissue parts. Maltoma in salivary gland is a rare entity because it is not a part of salivary gland tissue, but chronic inflammatory processes can cause lymphocyte accumulation in the gland and these cells can be the cause of Maltomas.

Tumors located in the parotid gland are reported to be malignant in 26-32% of cases. Physical examination is not effective to rule out malignancy in all cases. Preoperative studies including imaging modalities such as ultrasonography (US) and magnetic resonance imaging (MRI), and histopathological studies (fine needle aspiration cytology (FNAC)) are used to have accurate preoperative diagnosis.

In this case, we report a rare tumor of the parotid gland in a patient with a breast cancer history that is a confusing clinical situation and the insufficiency of preoperative and postoperative diagnostic studies to determine the pathology in the parotid gland.

Despite improvement of diagnostic studies, personal experience and evaluation of the clinician remain the most important part of planning of treatment strategies for parotid gland neoplasms.

### Introduction

Mucosa-associated lymphoid tissue (MALT) is a non-encapsulated cluster of lymphocytes that is thought to have an important role in mucosal immunity [1]. In 1983 Isaacson and Wright described lymphoma arising from MALT (Maltoma) and it was recognized as a discrete entity in 1994 by the Revised European-American Lymphoma classification [2]. Maltoma is an extranodal lymphoma either of B- or T-cell origin that may most commonly involve the stomach, followed by other gastrointestinal organs, respiratory tract, skin and others containing lymphoid tissue parts [3-7]. Any local chronic inflammatory process (autoimmune diseases, infections) may induce lymphoid proliferation and is suspected to be in the pathogenesis of Maltoma [8,9]. In general Maltoma is a low grade, localized tumor and usually does not metastasize at early periods [10].

Maltoma in salivary gland is a rare entity because lymphoid tissue is not a part of salivary gland, but chronic inflammatory processes like Sjogren's Disease and Hashimoto's Thyroiditis can cause lymphocyte accumulation in the gland and these cells can be the cause of Maltomas [8,10,11]. In Sjogren's disease (SD) periductal lymphoepithelial islands consist of reactive, polyclonal cells are seen in the parotid gland and normal lobular appearance of the gland is preserved [11]. There is an increased risk of malignant lymphomas in chronic autoimmune disorders and in SD this risk is approximately 44 times higher than the normal population [12]. It is seen in biochemical analysis of gland biopsies that immunoglobulin M bearing plasma cells are increased in glands and this suggests the possibility of monoclonal expansion of lymphocytes in SD. If a monotypic plasma cell is found in gland tissue the patient has a higher risk of lymphoproliferative disorders [13].

Tumors located in the parotid gland are reported to be malignant in 26-32% of cases [14]. Physical examination is not effective to rule out malignancy in all cases. Preoperative studies including imaging modalities such as ultrasonography (US) and magnetic resonance imaging (MRI), and histopathological studies (fine needle aspiration cytology (FNAC)) are used to have accurate preoperative diagnosis [14].

Metastasis to the parotid gland is a common reason for parotid gland masses. In a recent study it

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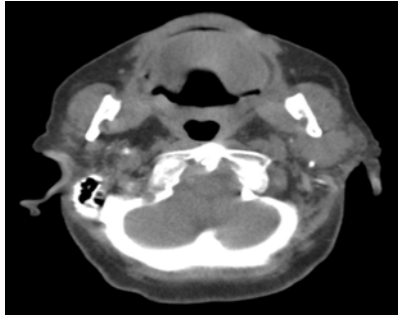
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**Figure 1:** CT image of parotid mass extending to the deep lobe.

was reported that in 89 patients with malignant tumors of parotid gland, 39 (44%) were metastatic [15]. Although majority of primary tumor in these cases located in head and neck region, metastasis of renal cell carcinoma, breast carcinoma and carcinoma of bronchus was reported in this study.

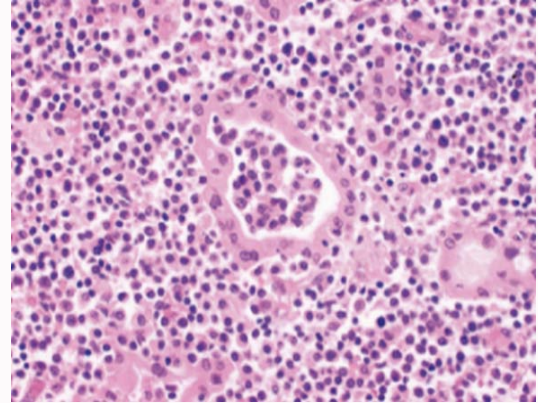
In this case, we report a rare tumor of parotid gland in a patient with breast cancer history that is confusing clinical situation and the insufficiency of preoperative and peroperative diagnostic studies to determine the pathology in parotid gland.

### Case Presentation

A 69-year-old woman presented with a growing mass over left parotid gland for about a month. Patient had a breast cancer and right partial mastectomy history, 10 years ago. After the operation she had been treated by adjuvant radiotherapy and chemotherapy. Six years after the first treatment a local recurrence observed during routine follow ups and right radical mastectomy procedure had been added. Adjuvant chemotherapy (trastuzumab) had been given after surgery for one year. Patient still uses letrozole since her last operation. A computed tomography (CT) scanning had been done in another medical center before she referred to our clinic. CT showed a lobulated, hyperdense parotid gland mass, containing microcalcification areas and extending to deep lobe (Figure 1). US and MRI scanning and FNAB offered to patient, but she refused MRI. US showed bilateral atrophy in submandibular gland, and a solitary, hypoechoic 30 mm in diameter mass with hyperechoic foci in left parotid gland. FNAB reported as Warthin's tumor. After these results surgical procedure was planned.

In operation main trunk of facial nerve was found. While dissecting superficial lobe of gland it was seen that buccal branch of facial nerve was wrapped by tumor and tumor was going through the deep lobe. The buccal branch was sacrificed and remaining branches of facial nerve dissected carefully and superficial lobe taken out. Incisional sample taken from deep lobe tumor and send for frozen section evaluation together with superficial lobe. After total dissection of facial nerve over the tumor, deep lobe of the parotid was removed. A frozen section result showed no malignancy and operation was concluded. She had grade IV (House Brackmann Scale) facial nerve palsy developed in early postoperative period and after 6 months facial palsy completely recovered.

Pathology department reported "extranodal marginal zone lymphoma (maltoma)", with immunohistochemical stains finding the lymphoid cells to be positive for CD20, bcl-2 after paraffin block evaluation. The Ki67 was estimated at only 7% of the lesion. Tumor free areas of gland showed changes suggesting SD (Figure 2).



**Figure 2:** Microscopic appearance of tumor.

After pathological results patient reexamined for symptoms of SD and we found that the patient had complaint about dryness of eyes for one year and this symptom had been attributed to medication history for breast cancer by her oncologist.

The patient was informed about her pathological result and we offered her chemotherapy for maltoma. She refused chemotherapy because of her treatments for breast cancer. She comes for routine follow up physical examinations and as a radiological examination she accepts US alone and she has not any problem in 14th month of postoperative period.

### Discussion

Because of the slow-growing nature of maltoma expected survival rates can be observed even in advanced cases [16]. This survival rates mandates surgeon to select lower morbidity procedures in this case. Although we need pathological proof of histology of the mass before the decision of treatment modality, this may not be available in some cases for parotid gland. Incisional biopsy is contraindicated for parotid gland masses because of the risk of tumor in-semination and facial nerve injury [17]. FNAC is an evaluation study that can be easily done with almost no pain to determine whether the mass is benign or malignant preoperatively. In the literature the sensitivity and specificity of FNAC is reported in range of 54-92% and 86-100% respectively [18-20]. Cytological analysis of hypercellular tumors is particularly difficult. Frozen section (FS) is a study, performed preoperative and reported sensitivity and specificity are between 77-93% and 95-100% respectively [14].

Like as other soft tissue masses, MRI is the imaging method of choice for the evaluation of parotid tumors. In the recent literature, the sensitivity of parotid FNAC in the diagnosis of malignancy has been reported to be in the range of 54-92%, with specificity in the range of 86-100% [18,19]. The false-negative rate ranges from 2% to 31% and the false-positive rate from 0% to 7% [14]. In our case we could not have MRI because of patient's refusal.

FS is a good diagnostic tool to have information on the nature of the tumor whether it is malignant or not during surgery [14]. Despite it is a very effective method in most body parts its accuracy in salivary gland masses is controversial. Therapeutic decision should not be based on FS solely. Therefore surgeon must consider frozen section in conjunction with the preoperative clinical findings, diagnostic tests and patient's medical history.

Like as other tissues intraparotid metastases can occur many

years after primary tumor treatment, and can be the first clinical manifestation of a recurrence. Parotidectomy with complete excision of the tumor can be curative [15] and is necessary for exact diagnosis in most cases. In our case we could have pathological diagnosis after total excision of gland.

In this case we reported fail of both preoperative and peroperative diagnostic studies in a patient with a confusing previous malignancy history. In our case we decided to perform total parotidectomy because of patient's breast cancer history and infiltrating feature of the mass made us think that it could be malignant. Perineurium is a well-known barrier to tumor invasion, and this allowed us to protect facial nerve because of we had no certain diagnosis of malignancy. FS result confirmed our decision and we protected patient's facial nerve branches those were not affected. In fact our patient refused further therapies for Maltoma and we follow up her by physical examination and US alone according to her request.

## Conclusion

Despite improvement of diagnostic studies personal experience and evaluation of clinician remains most important part of planning of treatment strategies for parotid gland neoplasm.

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