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CASE REPORT
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UNCOMMON ENCOUNTERS: A CASE SERIES OF FOLLICULAR LYMPHOMA PRESENTING IN THE PAROTID GLAND

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ABSTRACT

INTRODUCTION

Follicular lymphoma (FL) is a slow-growing B-cell non-Hodgkin lymphoma that predominantly involves lymph nodes. Primary extranodal involvement is uncommon, particularly in the salivary glands, where it may mimic benign lesions and lead to delayed diagnosis. This case series highlights two patients with parotid gland involvement, demonstrating varied clinical presentations and management strategies.

METHODS

We report two cases of patients presenting with parotid gland swelling who were subsequently diagnosed with follicular lymphoma. Clinical evaluation, imaging, histopathological examination, and immunohistochemical analysis were utilized to establish the diagnosis. Treatment approaches and clinical outcomes were reviewed.

RESULTS

Both patients presented with parotid swellings of differing characteristics and durations. Initial clinical impressions suggested benign salivary gland pathology. However, further diagnostic workup confirmed follicular lymphoma in both cases. The patients exhibited distinct clinical courses and were managed with different therapeutic strategies based on disease extent and individual factors.

DISCUSSION

Primary follicular lymphoma of the parotid gland is rare and can be easily misdiagnosed due to its resemblance to benign salivary lesions. These cases emphasize the importance of maintaining a high index of suspicion and performing comprehensive diagnostic evaluations in persistent or atypical salivary gland swellings. Early and accurate diagnosis is essential to guide appropriate management and improve outcomes.

KEYWORDS

Case series, non-Hodgkin lymphoma, Follicular lymphoma, Salivary gland tumor, Parotid gland swelling.

INTRODUCTION

Salivary gland swellings are commonly encountered in ENT clinical practice. While most of them are benign conditions, including pleomorphic adenoma, Warthin's tumor or reactive lymphoid hyperplasia, there are

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instances where they can be indicative of more serious conditions. Lymphomas that primarily develop in salivary gland tissue are quite rare, accounting for only about 2–5% of all tumors found in these glands. Most cases of lymphoma affect the major salivary glands, particularly the parotid gland, which accounts for about 1-4% of cases.[1]. These neoplasms may originate from an intraglandular lymph node or within the gland itself. Follicular lymphoma (FL) is the second most common subtype of non-Hodgkin lymphoma (NHL), comprising about 35% of NHL cases [2]. However, its occurrence in the parotid gland is relatively rare, accounting for only around 25.8% of parotid gland NHL cases [15]. The clinical presentation of the condition often manifests subtly, usually characterized by a painless and progressively enlarging mass. This case series presents two patients with parotid gland follicular lymphoma, each with a distinct clinical course. The series underscores the importance of biopsy and a multidisciplinary evaluation in cases of persistent or unusual salivary gland swelling.

Case 1

A 57-year-old gentleman, a former smoker with a history of Diabetes Mellitus and Dyslipidemia, presented to the ENT clinic with a noticeable, painless swelling on the right side of his neck that had been steadily increasing in size over the past few months. He reported no accompanying symptoms such as fever, weight loss, or changes in appetite, and no history of trauma to the neck. Furthermore, the patient denied any family history of malignancy.

A clinical examination revealed a firm swelling in the right upper neck area, measuring 2 cm, with no overlying skin changes. Both the neck and oral examinations were unremarkable. A flexible nasoendoscopy was performed and showed normal results, with no medialization observed.

An ultrasound of the neck was performed, revealing a 3 x 2 cm lesion in the right parotid gland that showed vascularity. Fine needle aspiration cytology (FNAC) results indicated that the lesion is non-neoplastic and suggested the possibility of sialolithiasis. A Contrast-Enhanced Computed Tomography (CECT) scan of the neck demonstrated bilateral intraparotid lymph nodes and neck lymph nodes with mild sub centimeter enlargement.

Given the enlarged neck mass, he was subjected to right superficial parotidectomy. Histopathological examination confirmed that the overall findings are consistent with follicular lymphoma, grade 1. Immunostaining was typical: CD20+, CD10+, BCL2+, with a low proliferation index (Ki-67 <20%). Positron Emission Tomography (PET) scan revealed supra and infra-diaphragmatic nodal deposits, classifying his lymphoma as Stage III (advanced). Bone marrow examination revealed no marrow involvement.

He was referred to a hematologist for further management. Due to the indolent nature of his disease and the presence of non-aggressive symptoms, the treatment options were discussed. These included a watch-and-wait approach or starting Rituximab-based therapy. He opted to begin with monoclonal antibody therapy and received four cycles of weekly Rituximab. Following this, he continued with maintenance therapy for two years, during which the disease remained stable.

Case 2

A 63-year-old gentleman with a history of dyslipidemia visited our clinic complaining of swelling below his left ear that had persisted for four months. The swelling was painless and had remained consistently the same size. He did not

report any additional swellings or constitutional symptoms. Notably, he has a strong family history of malignancies, including lung cancer in both his father and elder sister.

A clinical examination revealed swelling in the left lower ear measuring 3 x 2 cm. The swelling was firm, mobile, and showed no changes in the overlying skin. Both the oral and neck examinations were normal. A flexible nasoendoscopy was performed, which did not show any signs of medialization. A Contrast-Enhanced Computed Tomography (CECT) scan of the neck indicated a lesion in the left parotid gland, with differential diagnoses being either a lymph node or a salivary gland tumor. A Fine Needle Aspiration Cytology (FNAC) was then conducted, which yielded atypia of undetermined significance, characterized primarily by a lymphocyte-rich aspirate.

He underwent a left superficial parotidectomy a few weeks later. The histopathological examination confirmed the diagnosis of follicular lymphoma, grade 2. Immunostaining results were characteristic, showing positivity for CD20, CD10, and BCL2, along with a low proliferation index (Ki-67 between 5% and 10%). A Positron Emission Tomography (PET) scan revealed infradiaphragmatic nodal deposits, classifying his lymphoma as Stage III (advanced). Given the localized and inactive nature of his disease, along with the absence of symptoms, a shared decision was made to adopt a watch-and-wait strategy. At the six-month follow-up, he remained asymptomatic and continued under surveillance.

DISCUSSION

Salivary gland lymphoma is a type of tumor that shows a wide range of symptoms and imaging results. This variety makes it difficult to distinguish it from other tumors in the salivary glands. Lymphomas represent 3% to 4% of malignant tumors and are the most common head and neck malignancies after squamous cell carcinoma and thyroid cancer [3]. Primary malignant lymphomas of the salivary glands are mainly B-cell lymphomas, including mucosa-associated lymphoid tissue (MALT) lymphoma, follicular lymphoma, and diffuse large B-cell lymphoma [4].

Among salivary gland lymphomas, the most typical subtype is MALT lymphoma, followed by follicular lymphoma [5]. Primary parotid gland lymphoma is a limited segment of head and neck NHL. The overall incidence is just about 0.3% of all tumors and 5% of extranodal lymphomas [6]. In comparison to other extranodal sites affected by NHL, involvement of the parotid gland tends to manifest as a low-grade form of the disease. Patients with NHL in the parotid gland generally experience a more favorable prognosis than those whose lymphoma affects other extranodal areas [6].

This growth typically occurs without any accompanying systemic symptoms, making it difficult to differentiate from more frequently encountered benign salivary gland lesions. As a result, the patient may remain unaware of the underlying issue until the mass becomes more pronounced. Diagnosis of FL typically necessitates a comprehensive approach that includes imaging, histopathological examination, and immunophenotyping. Fine-needle aspiration cytology (FNAC), although frequently used as a first-line diagnostic tool for parotid masses, often falls short in diagnosing FL accurately due to its architectural subtleties [7]. As a result, a core needle biopsy (CNB) or excisional biopsy is often required to obtain a definitive diagnosis and accurate grading. CNB

for salivary gland lesions has shown lower rates of nondiagnostic results and superior diagnostic performance in identifying malignant tumors compared to fine needle aspiration cytology (FNAC)[14].

Histologically, FL is defined by the presence of neoplastic follicles composed of centrocytes and centroblasts. Furthermore, immunohistochemical analysis reveals positivity for CD20, CD10, and BCL2 markers [8]. Imaging techniques like ultrasound, computed tomography (CT), or magnetic resonance imaging (MRI) are typically employed to evaluate the mass. The literature suggests that CT and MRI findings of MALT lymphoma in the parotid gland can exhibit various appearances in the contours and internal structures of the masses. These include a solitary solid mass, a solitary solid-cystic mass, a diffusely solid-cystic lesion, or multiple solid nodules or masses. Among these, solitary and diffusely solid-cystic lesions are more commonly observed [9].

The outlook for patients with primary parotid FL is very positive, especially when the condition is diagnosed early. MacManus and Hoppe (1996) reported that patients with localized FL who underwent radiotherapy achieved long-term disease-free survival. The five-year survival rates for Stage I disease exceed 90%, and there are low rates of transformation to more aggressive subtypes [10]. Treatment strategies for parotid FL depend on the stage of the disease and the symptoms experienced by the patient.

For localized disease (Stage I or II), the typical approach is surgical excision, such as a superficial parotidectomy, followed by involved-field radiotherapy. FL is known to be sensitive to radiation, and the local control rates with radiotherapy alone are high.[11]. In asymptomatic patients with low-burden disease, a “watch-and-wait” approach may be reasonable, as shown in randomized trials for low-grade NHL [12]. For advanced or symptomatic disease, systemic treatment with anti-CD20 monoclonal antibodies (rituximab), alone or in combination with chemotherapy (e.g., R-CHOP or R-CVP), is the mainstay. Treatment selection should be individualized based on disease burden, comorbidities, and patient preference [13].

CONCLUSION

Follicular lymphoma of the parotid gland is a rare and often underrecognized presentation of NHL. Its clinical and radiologic similarity to benign salivary gland tumors necessitates a high index of suspicion. Histopathological confirmation, supported by immunohistochemistry, is essential for accurate diagnosis. With appropriate staging and treatment, the prognosis of localized parotid FL is excellent. Recognition of this rare entity allows for timely management and avoidance of overtreatment or misdiagnosis.

Authors Declarations

Ethical Approval: Obtained

Consent to Participate: Obtained

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