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LETTER TO THE EDITOR

The hidden oncological challenge in Sjögren's syndrome with a focus on pharyngeal cancer

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Recent studies have revealed a link between autoimmune diseases and a heightened risk of developing cancer, notably in patients with autoimmune rheumatic diseases.[1,2] This is particularly true for Sjögren's syndrome, which is well-documented to increase the risk of lymphoma.[3] However, recent findings suggest that individuals with Sjögren's syndrome might also be at an elevated risk for a wider range of cancers, including the less commonly reported pharyngeal cancer. This type of cancer often mimics sialadenitis, making accurate diagnosis challenging.

A 49-year-old male patient with a history of Sjögren's syndrome, previously treated tuberculosis, and chronic obstructive pulmonary disease presented with the complaint of a mass in the left jaw region, resembling sialadenitis but lacking tenderness or typical inflammatory signs. The diagnosis of Sjögren's syndrome was established one and a half years prior, adhering to the 2016 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria. This diagnosis was supported by the presence of a positive anti-SSA (Ro) antibody (50.1 U/mL), a positive Schirmer test (3 mm/5 min in both eyes), and a reduced unstimulated whole saliva flow rate (0.2 mL over 5 min). The treatment for the patient focused on alleviating sicca symptoms, with no extraglandular manifestations reported.

Upon admission, laboratory analyses revealed anemia, indicated by a hemoglobin concentration of 10.8 g/dL, and raised C-reactive protein levels, which were measured at 54.05 mg/L. A computed tomography scan indicated necrotic lymph nodes, leading to a diagnosis of metastatic carcinoma confirmed by fine-needle aspiration. Subsequent magnetic resonance imaging (MRI) and positron emission tomography (PET)/computed tomography (CT) confirmed the presence of left hypopharyngeal cancer with ipsilateral lymph node metastasis involving the left submandibular gland and sternocleidomastoid muscle (5.2×4.8 cm; Figures 1a-c). Following radical neck dissection, squamous cell carcinoma with metastasis, including invasion into the submandibular gland was confirmed (Figures 2a, b). The patient was diagnosed with Stage IVA hypopharyngeal cancer and underwent three months of cisplatin and radiation therapy. However, restaging after treatment revealed a new nasopharyngeal mass with skull base destruction and lung metastasis. Despite undergoing an additional five months of chemotherapy, treatment was discontinued due to his deteriorating condition. Subsequently, the patient received four months of symptomatic conservative care before being lost to follow-up.

The rising incidence of pharyngeal carcinoma, which includes nasopharyngeal, oropharyngeal,

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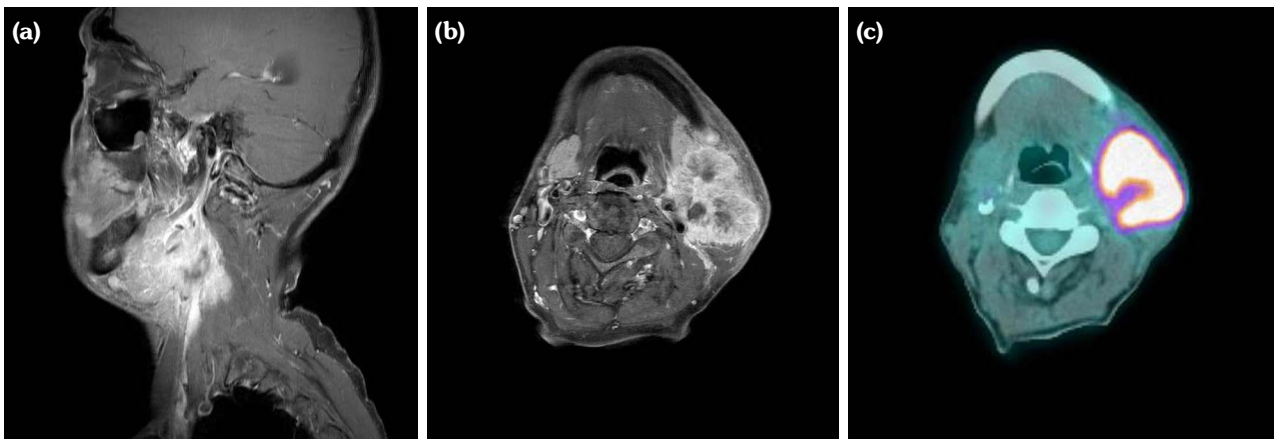

Figure 1. MRI and PET/CT visualization of lymph node enlargement and direct tissue invasion in the left cervical region **(a)** Contrast-enhanced, T1-weighted sagittal MRI and **(b)** axial MRI illustrating extensive lymph node enlargement and conglomeration along the left jugular chain at level II, with direct invasion into the left submandibular gland and sternocleidomastoid muscle. **(c)** PET/CT imaging reveals a cluster of lymph nodes at the left cervical level II with intense fluorodeoxyglucose uptake, forming a conglomerated mass measuring 5.0×3.4 cm. The mass displays heterogeneous internal fluorodeoxyglucose distribution, indicative of pathological involvement, and is closely adjacent to the left submandibular gland.
MRI: Magnetic resonance imaging; PET: Positron emission tomography; CT: Computed tomography.

and hypopharyngeal cancers, poses a significant concern.[4] These cancers often present without symptoms or mimic benign conditions, making early detection difficult.[5,6] The prognosis for hypopharyngeal cancer remains poor, with a five-year survival rate of 30 to 40%.[7]

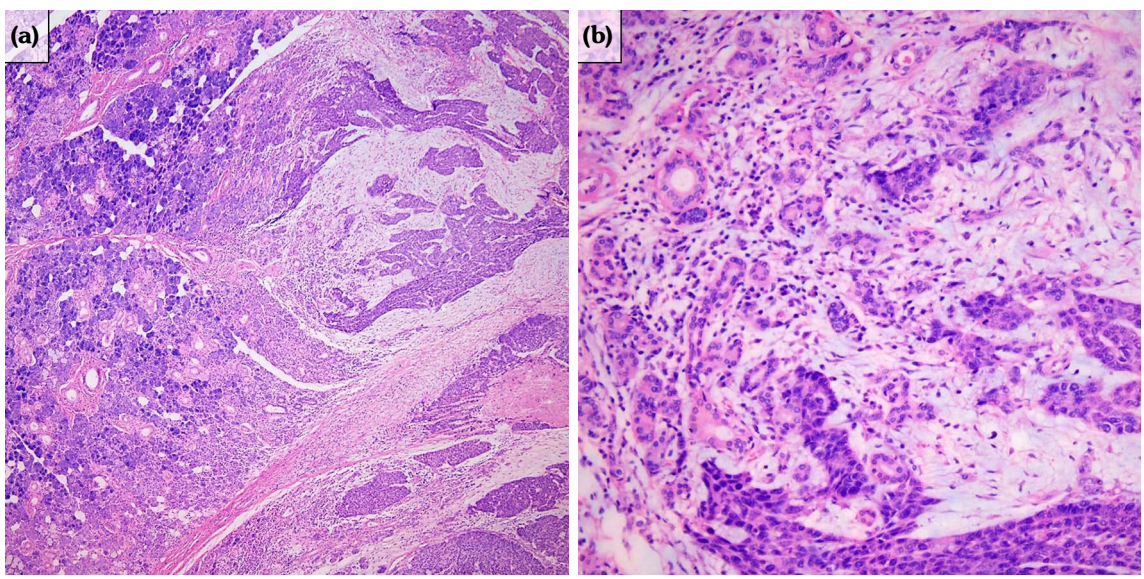

Figure 2. Histopathological findings of metastatic nasopharyngeal carcinoma infiltrating salivary gland tissue **(a)** Low-power view of a histological section of metastatic nasopharyngeal carcinoma. The section illustrates tumoral tissue with irregular islands of malignant cells penetrating the salivary gland parenchyma. Adjacent nonneoplastic salivary gland tissue appears to be destructed by the infiltrating tumor. Staining with hematoxylin-eosin, original magnification ×40. **(b)** High-power histological examination of metastatic nasopharyngeal carcinoma. The image reveals a dense infiltrate of malignant epithelial cells with hyperchromatic nuclei and irregular nuclear membrane. The carcinoma cells are observed infiltrating the fibrous stroma. Staining with hematoxylin-eosin, original magnification ×200.

Research from Taiwan's Health Insurance database suggests a 45% increased risk of head and neck cancer among individuals with Sjögren's syndrome.[8] The cause of this increased risk may be related to chronic inflammation and immune dysregulation associated with autoimmune diseases.[9] Notably, the Epstein-Barr virus, a known risk factor for pharyngeal cancer, has also been implicated in the pathophysiology of autoimmune diseases, including Sjögren's syndrome.[10] The similarity of symptoms between pharyngeal cancer and sialadenitis, often encountered in Sjögren's syndrome, underscores the necessity for vigilance in diagnostic and therapeutic approaches in clinical practice.

Patient Consent for Publication: A written informed consent was obtained from patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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REFERENCES

1. Bernatsky S, Ramsey-Goldman R, Clarke A. Malignancy and autoimmunity. Curr Opin Rheumatol 2006;18:129-34. doi: 10.1097/01.bor.0000209423.39033.94.
2. Giat E, Ehrenfeld M, Shoenfeld Y. Cancer and autoimmune diseases. Autoimmun Rev 2017;16:1049-1057.
3. Mariette X, Criswell LA. Primary Sjögren's syndrome. N Engl J Med 2018;378:931-9. doi: 10.1056/NEJMcp1702514.
4. Aupérin A. Epidemiology of head and neck cancers: An update. Curr Opin Oncol 2020;32:178-86. doi: 10.1097/CCO.0000000000000629.
5. Davis ED. The early diagnosis of carcinoma of the oral and laryngeal pharynx, including the diagnosis of inoperable cases. Proc R Soc Med 1920;13:202-3.
6. Koivunen P, Rantala N, Hyrynkangas K, Jokinen K, Alho OP. The impact of patient and professional diagnostic delays on survival in pharyngeal cancer. Cancer 2001;92:2885-91. doi: 10.1002/1097-0142(20011201)92:11<2885::aid-cncr10119>3.0.co;2-g.
7. Garneau JC, Bakst RL, Miles BA. Hypopharyngeal cancer: A state of the art review. Oral Oncol 2018;86:244-50. doi: 10.1016/j.oraloncology.2018.09.025.
8. Yang TH, Cheng YF, Chen CS, Lin HC. Increased prevalences of head and neck cancers in patients with Sjögren's syndrome. Head Neck 2023;45:2874-81. doi: 10.1002/hed.27518.
9. Carsons S. The association of malignancy with rheumatic and connective tissue diseases. Semin Oncol 1997;24:360-72.
10. Toussirot E, Roudier J. Epstein-Barr virus in autoimmune diseases. Best Pract Res Clin Rheumatol 2008;22:883-96. doi: 10.1016/j.berh.2008.09.007.