

Tumors of the Sinonasal Region Posing Diagnostic Difficulties: Secondary Lymphoma and SMARCB1- Deficient Carcinoma Case Series

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1. Introduction

Sinonasal malignancies are uncommon neoplasms that constitute approximately 1% of all human malignancies and around 5% of head and neck cancers. They predominantly affect men in the fifth and sixth decades of life, with an estimated incidence of 1 per 100,000 individuals, although this rate may vary across geographic regions. Timely diagnosis is often difficult because the presenting symptoms including nasal obstruction, epistaxis, and rhinorrhea, frequently resemble those of benign inflammatory sinonasal disorders. These tumors typically arise within air-filled cavities that the tumor infiltrates before the development of overt signs and symptoms; therefore, most patients present with advanced-stage disease at diagnosis, often demonstrating extensive involvement of adjacent structures. The prognosis is with a 5-year overall survival ranging from 60% for early tumors (T1–2) to 20% for advanced disease [1].

As a result, many patients are diagnosed at a locally advanced stage. Several environmental and occupational exposures, particularly to wood dust, leather dust, and metal and textile industry by-products, together with tobacco consumption, have been implicated in the pathogenesis of these tumors [2].

From a histopathological perspective, sinonasal malignancies encompass a broad and heterogeneous spectrum of epithelial and non-epithelial neoplasms, including squamous cell carcinoma, adenocarcinoma, mucosal melanoma, olfactory neuroblastoma, and lymphoma. Owing to their biological heterogeneity and the intricate anatomy of the sinonasal region, optimal management

generally necessitates a multidisciplinary and multimodal treatment strategy incorporating surgery, radiotherapy, and, where appropriate, systemic therapy [3].

Hodgkin lymphoma (HL) is a hematopoietic malignancy arising from germinal center B cells. Patients are typically diagnosed in their 20s or 30s and commonly present with supradiaphragmatic lymphadenopathy, often accompanied by systemic B symptoms. Even in advanced stages, HL is highly curable with combination chemotherapy, radiotherapy, or combined-modality treatment. Approximately 90% of patients with HL achieve long-term survival [4].

Several studies have demonstrated an increased risk of second (and multiple) malignancies following treatment for HL; both hematologic neoplasms and solid tumors collectively represent the leading cause of mortality among long-term survivors of HL. Survivors of HL are also at a 13-fold increased risk of developing a second primary non-Hodgkin lymphoma (NHL). A Dutch study with a 40-year follow-up estimated a standardized incidence ratio of 4.2 for any solid cancer, corresponding to an absolute excess risk of 100 cancers per 10,000 person-years and a 30-year cumulative incidence of 29%.³ Despite therapeutic advances in HL treatment, this increased cumulative incidence of second solid malignancies did not differ between patients treated in the 1960s and those treated in the 1990s [5].

Plasmablastic lymphoma (PBL) is a rare and highly aggressive subtype of B-cell lymphoma, most commonly seen in HIV-positive individuals, although it has also been reported in HIV-neg-

ative patients.⁵ Clinically, it frequently arises in extranasal sites such as the oral cavity and paranasal sinuses [6].

SMARCB1 (INI1) is one of the core components of the SWI/SNF chromatin remodeling complex and functions as a tumor suppressor gene involved in the regulation of cellular proliferation and differentiation. Sinonasal carcinoma characterized by SMARCB1 loss is a rare malignancy with an aggressive clinical course. This tumor poses diagnostic challenges, particularly due to its high propensity for local invasion and its potential to be initially mistaken for benign lesions. Its prognosis is poorer compared to other subtypes of sinonasal carcinoma. SMARCB1 immunostaining was performed on 142 sinonasal carcinomas [7].

In this case series, we present two rare and diagnostically challenging sinonasal malignancies with distinct histopathological backgrounds but overlapping clinical features of aggressive local invasion and potential initial misinterpretation as benign disease. The first case involves an HIV-negative patient who developed sinonasal plasmablastic lymphoma three decades after successful treatment and remission of Hodgkin lymphoma, highlighting the possibility of late secondary lymphoid malignancies in long-term survivors. The second case describes SMARCB1-deficient sinonasal carcinoma that was initially interpreted as an inflammatory polyp at an external center but was subsequently diagnosed as an invasive malignant epithelial tumor following repeat biopsy and comprehensive immunohistochemical evaluation. Through these cases, we aim to emphasize the importance

of maintaining a high index of suspicion in patients with persistent sinonasal symptoms, radiologically aggressive features, or discordance between clinical, radiological, and pathological findings. Early repeat biopsy, broad immunohistochemical assessment, and multidisciplinary management are essential for accurate diagnosis and timely treatment of these uncommon but highly aggressive sinonasal tumors.

2. Case 1

A 72-year-old male patient presented to our clinic with complaints of nasal obstruction, speech difficulty, and dyspnea persisting for approximately one month. His past medical history included hypertension (HT), diabetes mellitus (DM), and hyperlipidemia (HPL). Notably, he had been diagnosed with Hodgkin lymphoma (nodular sclerosing subtype, stage IIIS) in 1995 and achieved remission following six cycles of ABVD chemotherapy.

On physical examination, a violaceous mass was observed filling both nasal cavities, destructing of the nasal floor and extending to the hard palate, occupying 3×2 cm on the hard palate.

Paranasal computed tomography and MRI scans revealed a large, intensely contrast-enhancing solid mass destructing the nasal septum, extending to the anteromedial orbit and maxillary sinuses, and eroding the hard palate and F-18 FDG PET scan demonstrated uptake in the mass with an SUVmax of 6.02 (All of the findings showing in Figure 1).

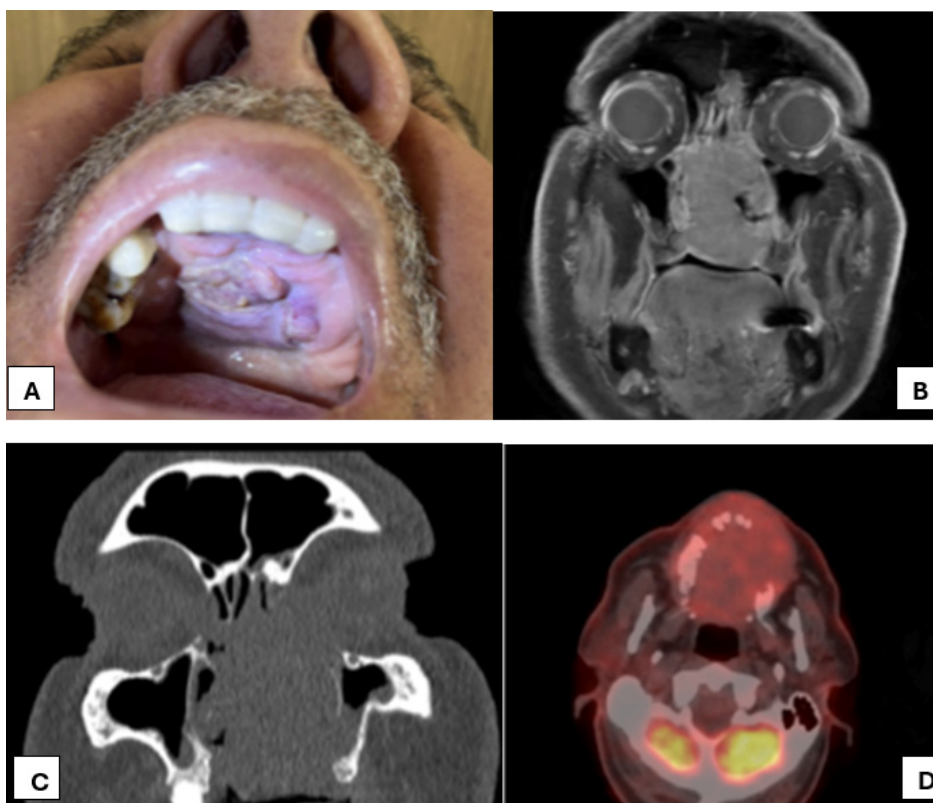


Figure 1 **A:** showing the lesion destructing the nasal floor and extending to the hard palate, occupying approximately 3×2 cm area. **B and C** MRI (B) and Paranasal CT (C) imagings revealed a large, intensely contrast-enhancing solid mass destructing the nasal septum, extending to the anteromedial orbit and maxillary sinuses, and eroding the hard palate and **D** F-18 FDG PET scan demonstrating uptake in the mass with an SUVmax of 6.02.

The patient's laboratory findings were unremarkable, and he tested negative for infectious diseases, including HIV.

Histopathological examination of the biopsy obtained from the mass filling the left nasal cavity was reported as "high-grade hematolymphoid neoplasm." Immunohistochemical analysis demonstrated positivity for CD38, CD138, MUM1, Bcl-2, cyclin D1, CD56, EBER and c-MYC in neoplastic cells while HHV-8 and ALK were negative. Considering these histopathological findings along with the lesion being located on head and neck region, the patient was diagnosed with plasmablastic lymphoma.

The patient then referred to the Hematology Department. Chemotherapy with the EPOCH protocol was initiated. The patient's treatment is currently ongoing.

3. Case 2

A 65-year-old male patient presented to our outpatient clinic with a complaint of right-sided nasal obstruction persisting for approximately 6 months. His medical history was significant for diabetes mellitus, hypertension, coronary artery disease, and asthma. He had undergone carotid artery stenting 2 months prior and was actively receiving anticoagulant and antiplatelet therapy.

Rigid endoscopic nasal examination revealed a polypoid mass with a vascular appearance completely obliterating the right nasal cavity.

At the time of presentation, paranasal sinus computed tomography (PNCT) performed at an external center demonstrated a soft tissue density lesion filling the right maxillary sinus, ethmoid sinus, and nasal cavity, reported as a possible polyp. An incisional biopsy obtained at the same center was reported as an "inflammatory polyp."

Further imaging performed at our institution revealed a mass completely filling the right maxillary, frontal, and sphenoid sinuses as well as the ethmoid cells, causing destruction of the maxillary sinus walls and extending into the premaxillary area and masticator space. The lesion showed invasion of the pterygoid muscles, pterygoid plates, and the pterygopalatine fossa. Additionally, destruction of the medial orbital wall with extension into the extraconal fat was observed. Destruction of the hard palate and maxillary alveolar process was also noted, with extension into the oral cavity. These radiological findings were considered suggestive of malignancy (All of the findings showing in Figure 2).

Incisional re-biopsy performed at our center revealed an "invasive malignant epithelial tumor." Immunohistochemical analysis demonstrated loss of the SWI/SNF complex, consistent with loss of SMARCB1 (INI1) expression, leading to the diagnosis of sinonasal carcinoma.

Subsequent positron emission tomography-computed tomography (PET-CT) imaging showed no evidence of distant metastasis.

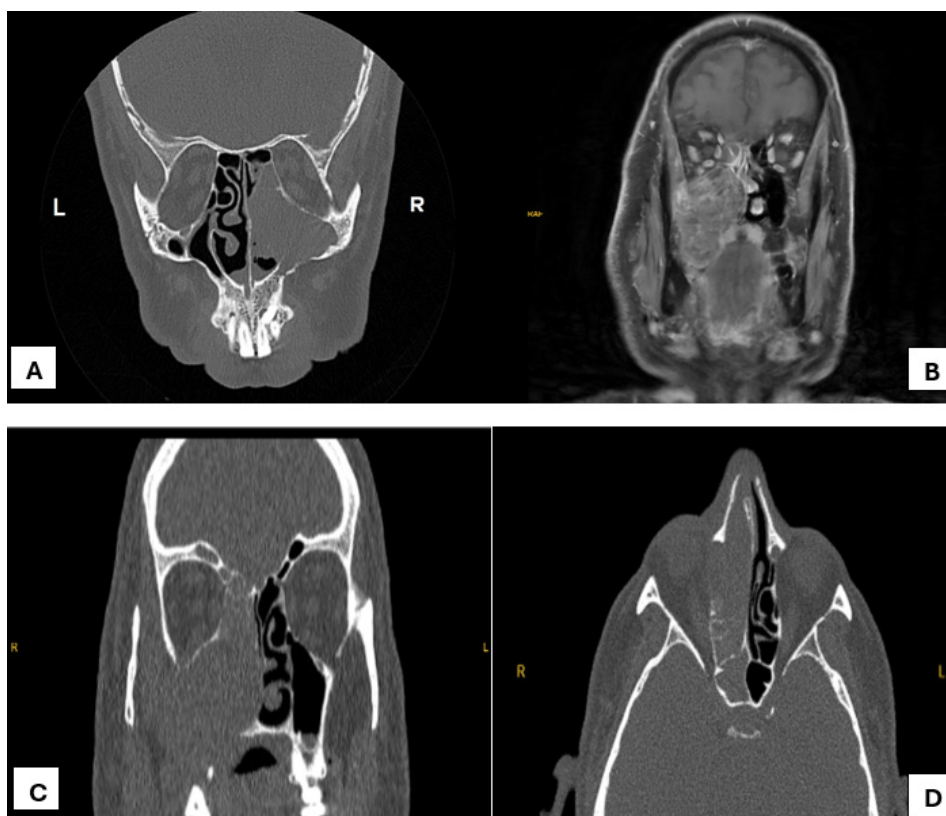


Figure 2 A: showing External center paranasal CT scan image taken 2 months before applying to our clinic. B showing the MRI taken in our clinic, a heterogeneous enhancing mass lesion measuring 73x53 mm in size, with irregular borders, was observed, filling the maxillary sinus on the right side, extending into the nasal cavity and resting on the septum, invading the inferior turbinate in the anterior, and extending to the choana in the posterior. The lesion invades the hard palate inferiorly. Superiorly, it extends to the orbital floor.

4. Discussion

Sinonasal malignancies represent a heterogeneous group of tumors that pose substantial diagnostic and therapeutic challenges because of their rarity, nonspecific presenting symptoms, complex regional anatomy, and tendency for advanced local invasion at the time of diagnosis. The two cases presented in this series illustrate different ends of the sinonasal malignancy spectrum: an extranodal hematolymphoid malignancy diagnosed as plasmablastic lymphoma and an aggressive epithelial malignancy characterized by SMARCB1 loss. Despite their distinct histopathological origins, both cases shared several clinically important features, including delayed recognition, extensive local involvement, and the need for close correlation between clinical, radiological, pathological, and immunohistochemical findings.

Plasmablastic lymphoma (PBL) is a highly aggressive B-cell non-Hodgkin lymphoma that is frequently associated with immunosuppression, particularly human immunodeficiency virus (HIV) infection [8]. Although rare, PBL can also occur in HIV-negative patients, typically at an older age, and may demonstrate extensive anatomical involvement as in the present case including the nasal cavity, hard palate, and orbit.

Plasmablastic myeloma (PBM) constitutes an important differential diagnosis. However, the primary localization in the nasal region, the absence of bone marrow involvement (at initial evaluation), and EBER positivity support a diagnosis in favor of PBL. In addition, MYC translocations in PBL are more commonly associated with immunoglobulin genes, distinguishing it from PBM. The high Ki-67 proliferation index (80–85%) reflects the aggressive biological behavior and poor prognosis of the disease [5].

SMARCB1-deficient sinonasal carcinoma is an aggressive malignancy that often presents at an advanced stage and poses significant diagnostic and therapeutic challenges. A multidisciplinary approach integrating clinical, radiological, and pathological findings is essential to optimize patient outcomes. While the erosion of the maxillary sinus wall was not obvious in the external center images of the patient 2 months ago, widespread invasion was seen in the imaging performed in our clinic (Figure 2). This shows us how aggressive the tumor is and how fast it progresses. This case shows us how aggressive a mass with no obvious erosion, which initially looks like a polyp, can be.

Both cases also underline the aggressive nature of sinonasal tumors and the importance of early multidisciplinary assessment. Because these lesions arise in anatomically confined spaces and may remain clinically silent until they invade adjacent structures, diagnosis is often delayed. Radiological findings such as bone erosion, orbital involvement, palatal destruction, masticator space extension, and rapid interval progression should raise suspicion for malignancy. Management should involve otorhinolaryngology, radiology, pathology, medical oncology, radiation oncology, and, when necessary, reconstructive surgical teams. Treatment is generally individualized according to tumor

type, extent of disease, resectability, and systemic status. Intensive chemotherapy regimens such as EPOCH may be used for plasmablastic lymphoma, whereas SMARCB1-deficient sinonasal carcinoma often requires multimodal treatment including surgery, radiotherapy, and/or systemic therapy.

In summary, these cases emphasize that rare sinonasal malignancies may mimic benign inflammatory disease and may present with advanced local invasion by the time of diagnosis. Persistent sinonasal symptoms, discordance between pathology and imaging, or radiological signs of aggressive behavior should prompt repeat tissue sampling and comprehensive immunohistochemical analysis. Recognition of entities such as plasmablastic lymphoma and SMARCB1-deficient sinonasal carcinoma is essential because early diagnosis and prompt multidisciplinary treatment may influence prognosis in these highly aggressive tumors.

5. Conclusion

First case holds importance in that a second lymphoma developed in the paranasal sinus 30 years after remission had been achieved following treatment for a prior lymphoma. It also highlights that PBL can occur in HIV-negative individuals. Furthermore, it demonstrates that PBL originating from the nasal cavity in HIV-negative patients may follow an aggressive clinical course. In the diagnosis of PBL, not only morphological features but also a broad immunohistochemical panel and EBER status are critical. In terms of management, the use of intensive chemotherapy regimens such as EPOCH and a multidisciplinary approach are of vital importance for prognosis. This case is noteworthy because of the potential for late-onset secondary lymphoid malignancies even decades after successful treatment of a prior lymphoma. It also underscores that plasmablastic lymphoma should be considered in the differential diagnosis of aggressive sinonasal masses, even in HIV-negative patients, as early recognition and prompt multidisciplinary management are critical for improving outcomes.

Sinonasal carcinoma with SMARCB1 loss is a rare but highly aggressive malignancy. It is typically diagnosed at an advanced stage and demonstrates a strong tendency for invasion into surrounding anatomical structures. Common sites of spread include the orbit, pterygopalatine fossa, skull base, and oral cavity. The most notable aspect of this case is that the initial biopsy was reported as an inflammatory polyp. In sinonasal malignancies, superficial or inadequate sampling may lead to false-negative results. In the presence of aggressive radiological findings—such as bone destruction, involvement of the masticator space, and orbital invasion—further evaluation and repeat biopsy are warranted despite a benign pathological report. SMARCB1 loss is demonstrated immunohistochemically by the absence of INI1 expression and serves as a key diagnostic marker. The treatment of these tumors is generally multimodal, involving combinations of surgical resection, radiotherapy, and chemotherapy. However, due to extensive local invasion, complete surgical resection is often challenging, and overall prognosis remains poor [9].

In conclusion, this case series highlights two rare and aggressive sinonasal malignancies with distinct histopathological characteristics but similar diagnostic challenges. Both plasmablastic lymphoma and SMARCB1-deficient sinonasal carcinoma may present with nonspecific sinonasal symptoms and may clinically or histopathologically mimic benign inflammatory lesions. These cases emphasize that persistent nasal obstruction, rapidly progressive disease, bone destruction, orbital extension, palatal involvement, or discordance between radiological and pathological findings should prompt repeat biopsy and comprehensive immunohistochemical evaluation. Early recognition, close clinicopathological correlation, and multidisciplinary management are essential for accurate diagnosis and timely treatment of these uncommon but highly aggressive sinonasal tumors.

6. Ethical Statement

Ethical approval was not required for this article because it is a case report describing clinical observations from routine practice. Our institution does not require formal ethical committee approval for reporting individual cases or case series. All procedures were conducted in accordance with the ethical standards of the institutional and national research committee and with the principles of the Declaration of Helsinki.

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