**Abstract**

**Objectives**  To present a case of squamous cell carcinoma (SCC) of the nasal cavity arising in a patient with granulomatosis with polyangiitis (GPA), formerly known as Wegener’s granulomatosis and to summarize literature recommendations for management.

**Study design**  Retrospective case report.

**Methods**  The medical records of a patient with longstanding GPA treated at a tertiary academic medical center were reviewed. The MEDLINE database was searched for “squamous cell carcinoma” with either “granulomatosis with polyangiitis” or “Wegener.”

**Results**  A 35-year-old male who had been diagnosed 15 years previously with GPA and treated at various intervals with cyclophosphamide, corticosteroids, mycophenolate mofetil, methotrexate, etanercept, and rituximab for sinusal, pulmonary, and renal involvement presented for follow-up at the Head and Neck surgery clinic complaining of right-sided orbital and cheek pain and nasal obstruction. These symptoms had developed since his last endoscopic sinus surgery 10 months prior. Repeat endoscopic sinus surgery was performed, revealing a friable, exophytic mass involving the right lateral nasal wall and septum. Pathological analysis demonstrated well-differentiated SCC. A comprehensive literature review yielded only three additional cases of SCC arising in patients with GPA.

**Conclusions**  The sinonasal symptoms associated with GPA are consistent with those seen in patients with chronic rhinosinusitis, but the presence of unilateral symptoms should raise suspicion for a neoplastic process, however rare. Cyclophosphamide and corticosteroids have previously been implicated in the pathophysiology of this malignancy, but equally plausible is the oncogenic role of chronic inflammation from GPA. Oncologic management closely parallels that in patients without GPA.

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**Case Presentation**

A 35-year-old male with GPA presented to the head and neck clinic for evaluation of sinonasal symptoms. He had been diagnosed 15 years previously with GPA, and had been treated at various intervals with cyclophosphamide, corticosteroids, mycophenolate mofetil, methotrexate, etanercept, and rituximab for sinusal, pulmonary, and renal involvement. He had most recently undergone bilateral revision endoscopic sinus surgery 10 months prior for chronic rhinosinusitis, and had previously undergone over 10 additional endoscopic sinus surgeries.

At the present visit, he complained of left-sided orbital and cheek pain and left-sided nasal obstruction. These symptoms had developed since his last endoscopic sinus surgery. Office endoscopy revealed mucosal edema and polypoid changes bilaterally. Repeat endoscopic sinus surgery was performed. Intraoperatively, a friable, exophytic mass involving the left lateral nasal wall and septum was noted, with extension into the lamina papyracea. Pathological analysis of specimen taken during surgery revealed well-differentiated SCC, stage T4N0M0.

Thereafter, the patient underwent a combined open and endoscopic maxillary lexcotomy for tumor resection. A medial maxillectomy was first performed endoscopically to resect the inferior extent of tumor. Then, a Lynch incision was used to approach the superior aspect of the tumor abutting the orbit. The lamina papyracea; medial periorbital; and medial orbital tissue, including the medial rectus were resected in bocce, and the medial orbit was reconstructed with an absorbable plate.

**Discussion**

Granulomatosis with polyangiitis (GPA), previously known as Wegener’s granulomatosis, is a systemic vasculitis affecting the upper airways, the lower airways, and the renal system. Otolaryngologic manifestations include septal perforation, saddle nose deformity, sinusitis, and subglotic stenosis. Treatment, which consists of induction and maintenance phases, includes long-term use of corticosteroids in addition to cyclophosphamide and/or immunosuppressive medications.

The association between GPA and the development of malignant tumors has been known for decades, but causality remains unclear. While chronic immunosuppression, including the use of cyclophosphamide, may predispose patients with GPA to develop malignancies, the oncogenic effect of systemic inflammation associated with GPA cannot be understated. Common malignancies encountered in patients with GPA include bladder carcinoma, renal carcinoma, gastrointestinal carcinoma, leukemia, and non-melanoma skin cancers.

Three previous cases of squamous cell carcinoma (SCC) arising in the nasal cavity and paranasal sinuses of patients with GPA have been reported. Here, we report the case of a patient who underwent long-term immunosuppressive treatment for GPA, ultimately presenting with SCC involving the left nasal cavity and paranasal sinuses with extension into the orbit. Optimal treatment, as with other cases of locally-advanced malignancy of the nasal cavity and paranasal sinuses, comprised surgical resection followed by postoperative adjuvant chemoradiotherapy. In the current presentation, the patient was diagnosed with recurrent and/or residual disease two months following completion of cancer therapy, and is currently undergoing further treatment.

**Conclusion**

The management of sinonasal manifestations of GPA may pose unique challenges, given the often unrelenting course of the disease, but standard therapies for chronic rhinosinusitis (CRS), including irrigation and topical therapies, as well as endoscopic sinus surgery, are routinely employed. As demonstrated in this case, however, the risk of malignancy in the paranasal sinuses should not be taken lightly: any severe worsening of CRS symptoms or new orbital findings in a patient with GPA should raise the suspicion of new sinonasal malignancy. Rapid diagnosis may aid in ensuring optimal medical and surgical management of any complications of this complex disease.

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**References**