

Background

- Chronic lymphocytic leukemia (CLL) and small lymphocytic lymphoma (SLL) are the most common leukemias in adults and primarily found in the bone marrow and lymph nodes, respectively.¹
- Common symptoms include weight loss, fevers, night sweats, muscle wasting, hepatosplenomegaly, and lymphadenopathy. As the disease progresses, patients may present with signs of anemia and thrombocytopenia, as well as recurrent infections.²
- The disease manifests as a monoclonal proliferation of incompetent mature B cells and has a characteristic cell immunophenotype: CD5, CD19, CD20, and CD23.
- The pathogenesis of chronic rhinosinusitis with nasal polyps (CRSwNP) is complex, and any factor affecting sinus ciliary function, mucus composition, immune response, and epithelial cells could promote disease.^{3,4}

Case Presentation

- A 60-year-old female recently diagnosed with CLL/SLL presented with three months of rhinosinusitis symptoms.
- Multiple medication trials including Ala-Hist PE, Nasonex, Afrin, Cefdenir, and Cleocin were unsuccessful.
- CT of paranasal sinuses (Figure 1) was obtained to assess the severity of sinus disease which revealed complete opacification of the bilateral ethmoid chambers with mucosal thickening in the maxillary sinuses and right sphenoid sinus.
- Nasal endoscopy revealed bilateral nasal polyps, turbinate hypertrophy, and closed sinus ostia.
- The patient underwent bilateral maxillary and sphenoid and left frontal balloon sinuplasty which provided moderate, temporary improvement.
- Medical therapy was attempted again with Alahist PE, prednisone, antibiotics, montelukast, ipratropium bromide, Trelegy, and Afrin.
- CRSwNP persisted for six months before sinus surgery was performed. The patient underwent total anterior and posterior ethmoidectomy and maxillary antrostomy with biopsy of bilateral nasal polyp tissue.
- Results of the biopsy are shown in Table 1. Tissue results were consistent with CLL/SLL indicating the patient's nasal polyps were indeed manifestations of underlying CLL/SLL.
- Within two months of the biopsy, the patient initiated chemotherapy with Rituximab. Presently, her symptoms are improved with the exception of mild headache.

Case Presentation (cont'd)

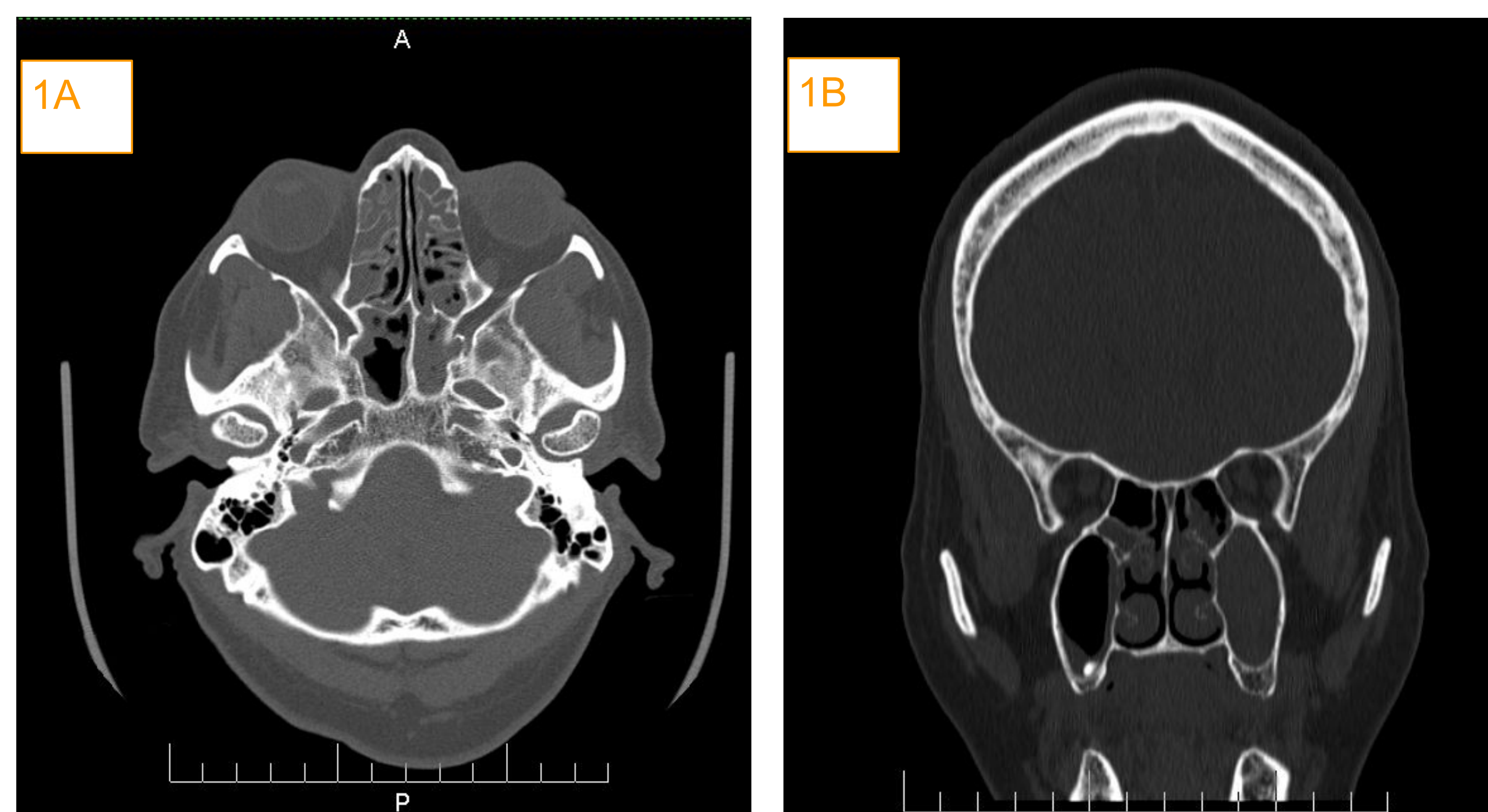


Figure 1. Computed Tomography of the Paranasal Sinuses Obtained Prior to Endoscopic Sinus Surgery. 1A is an axial CT image revealing near complete ethmoid and left greater than right sphenoid sinus opacification with mucosal thickening. 1B is a coronal CT image revealing complete left maxillary sinus opacification and turbinate hypertrophy. Polyp burden unable to be appreciated on pre-op CT imaging.

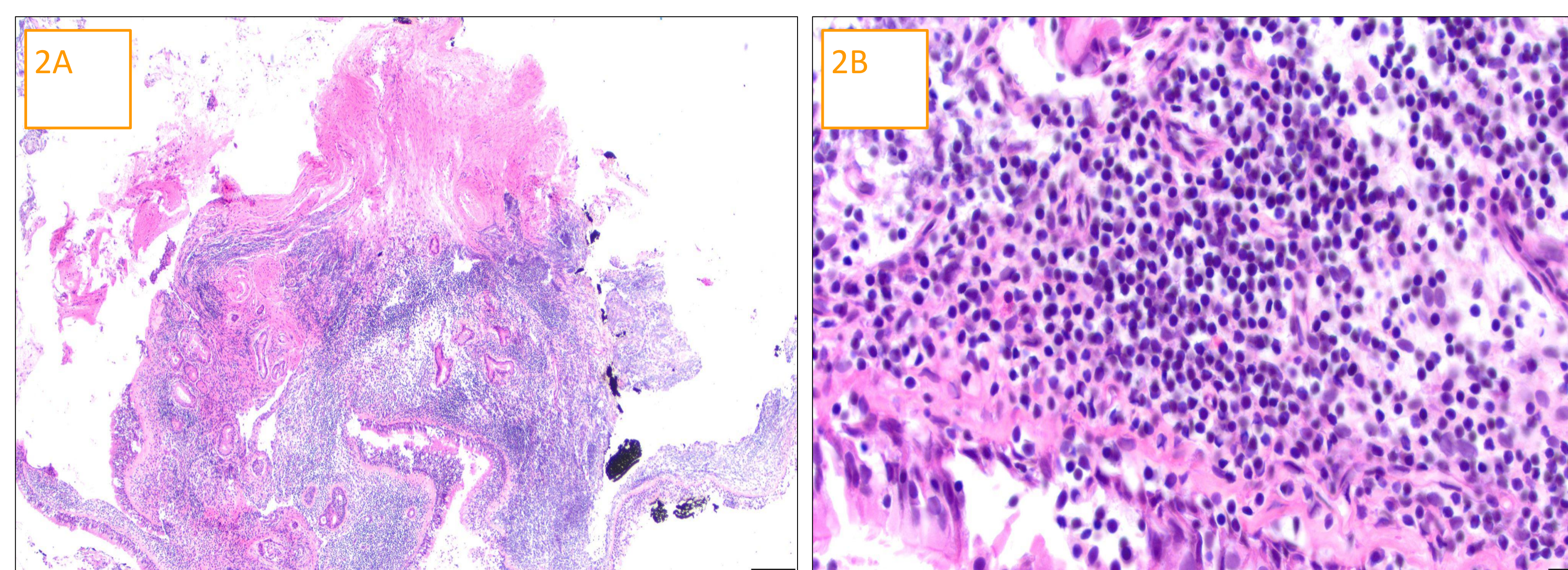


Figure 2. A and B. Histological images of the left (A) and right (B) nasal polyp specimens showing a lymphoid infiltrate composed predominantly of small lymphocytes. A few somewhat nodular-appearing areas were noted.

Test Performed	Result	Test Performed	Result
CD20	Positive	CD21	Negative
PAX-5	Positive	CD23	Positive
CD3	Negative	IgD	Positive
CD10 (CALLA)	Negative	CD43	Positive
CD5	Positive	MUM-1	Partial
BCL-1 (Cyclin D1)	Negative	CD30 (Ber-H2)	Negative
Ki-67 (MIB-1)	Partial	Kappa by ISH	Positive
BCL-2	Positive	Lambda by ISH	Negative
BCL-6	Weak partial	LEF-1	Variable

Table 1. Panel of immunostains performed on nasal polyp specimens. This patient's tissue samples were positive for CD5, CD20, CD23, CD43, PAX-5, BCL-2, IgD and trisomy 12.

Discussion

- The current treatment for CRSwNP typically involves a combination of medical therapies and sinus surgery for those with severe cases.³
- We believe this patient's CRSwNP was refractory to medical and surgical treatment due to the unique pathogenesis of sinus disease in CLL/SLL and its effects on sinonasal mucosa.
- Nasal polyps contain a higher number of B cells compared to healthy nasal tissue and CLL/SLL weakens the host immune system. The crowding of B cells and decreased immune function may both affect epithelial function as well.^{3,5}
- The patient's improvement on Rituximab, an anti-CD20 antibody which depletes CLL cells throughout the body, could be explained by the reduction of B cells present in sinonasal mucosa and subsequent return of sinonasal function.
- To our knowledge, this is the only reported case of CLL/SLL involving the sinonasal mucosa.
- Without endoscopic sinus surgery and polyp biopsy, the patient's underlying malignancy would have continued under watchful waiting. We hope this case will bring awareness to the rare yet possible role of underlying malignancy in recurrent sinonasal disease, and improve the quality of care for these unique patients.

Acknowledgments

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References

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