

A 46-year-old male presented to an outside emergency department with five days of progressively worsening diplopia and nasal congestion. Ophthalmology evaluation noted right papilledema and follow up computed tomography (CT) and magnetic resonance imaging (MRI) revealed an expansile mass of the right nasal cavity involving the bilateral ethmoid sinuses and bilateral sphenoid sinuses with orbital apex involvement and planum sphenoidale erosion. No intracranial extension was noted. Endoscopic biopsy revealed alveolar rhabdomyosarcoma (RMS). The patient was presented at a multidisciplinary tumor board as a T2a RMS of the ethmoid sinus. Tumor board recommendation was for neoadjuvant induction chemotherapy followed by definitive resection followed by adjuvant chemotherapy and radiation. The patient had a significant clinical and radiographic response to induction chemotherapy. He underwent definitive endoscopic resection including an orbital/optic nerve decompression and extended pterygopalatine fossa dissection to obtain negative frozen section margins along the sphenoid and ethmoid skull base, periorbita, pterygopalatine fossa (PPF), and optic nerve sheath. Final pathology did unexpectedly reveal microscopic disease along the optic nerve sheath. The patient is currently one year out from completion of concurrent adjuvant chemotherapy and proton beam radiation therapy with intact vision and without radiologic or clinic evidence of disease recurrence.