Background. Malignant triton tumor is an uncommon, aggressive subtype of malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation. The sinonasal region is rarely involved as the primary site.

Study Design. Case report including a detailed histopathologic and radiologic analysis along with a literature review.

Methods. We report the case of a 42-year-old man who presented to a tertiary university hospital with nasal airway obstruction, proptosis from a large fleshly mass in his left sinonasal tract with intracranial extension. Histopathologic assessment along with radiologic details are reviewed. A literature review to describe the background, incidence, disease course, and treatment options are presented.

Results. The extensive malignant triton tumor was treated with surgical excision via a lateral rhinotomy and craniofacial approach to the anterior fossa with postoperative radiation therapy. Histopathology demonstrated classic findings of rhabdomyoblastic scattered among elongated spindle cells. The literature demonstrates this disease to be rare in the sinonasal region with only 11 cases. The disease occurs predominantly in patients with neurofibromatosis type 1, with treatment being mainly complete resection followed by radiotherapy.

Conclusion. Sinonasal malignant triton tumor is a rare disease. Clinicians should include that as a differential diagnosis for a sinonasal mass and be aware of its clinical features and treatment.

Introduction

Malignant triton tumor (MTT) is a subtype of malignant peripheral nerve sheath tumor (MPNST) with rhabdomyoblastic differentiation. This histologic variant was first recognized in the cervical mass of a patient with neurofibromatosis type 1 (NF1) by Masson in 1932. The term triton, introduced in 1973 by Woodruff, refers to the triton salamanders that grow supernumerary limbs with both bone and muscle after implantation of severed sciatic nerve in their back in Locatelli’s experiment.

MTT is an uncommon and highly aggressive soft tissue sarcoma with less than 100 cases in the literature. One third of those arose in the head and neck with no more than 40 reported cases. Only 11 cases of MTTs are located in the sinonasal tract. We report the case of a 42-year-old man with a left sinonasal MTT with intracranial extension. The characteristic histopathologic features of MTT along with its radiologic presentation in the sinonasal tract are reviewed. A literature review to describe the background, incidence, disease course, and treatment options are presented.