INTRODUCTION

Chordomas are rare, relatively slow-growing tumors of notochordal origin that are aggressive, locally invasive, and radioresistant with a high rate of treatment failure.1,2 Although local recurrence continues to be the predominant form of treatment failure and best predictor of mortality, recurrence along the surgical pathway is becoming more recognized, especially as stereotactic radiation becomes more common.2,3 Tumor seeding along the pathway of surgical access is an infrequent mechanism for treatment failure, with only isolated cases documented in the literature.2

CASE PRESENTATION

A 35-year-old female presented to the Emergency Department with a 1-day history of acute onset upper and lower extremity weakness. Physical exam revealed a left hemiparesis as well as a left intranasal mass. Pertinent medical history included a clival chordoma treated at an outside hospital with multiple partial resections, most recently via a transseptal approach over 2 years prior, along with postoperative Gamma Knife radiotherapy. Outside records suggested only incomplete or subtotal resections for debulking purposes instead of intent to cure.

Rhinoscopy revealed a gelatinous, submucosal soft tissue mass completely occluding the left nasal passage and causing mild contralateral septal deviation. Magneto Resonance Imaging (MRI) demonstrated acute left posterior internal capsule and right lateral thalamic infarctions, as well as a 4 cmellar mass encasing the bilateral internal carotids seen in figure 1. There was an additional 2.8 x 2.5 cm T1 hypointense, T2 hyperintense heterogeneously enhancing mass in the left nasal cavity with mass effect and septal deviation. The two masses were confirmed by radiology to be separate entities without an obvious connection (Figures 1, 2).

Biopsy of the left nasal mass was performed and histopathologic examination confirmed the presence of chordoma with characteristic physaliferous cells with pale bubbly cytoplasm in a myxoid stroma (Figure 3).

The patient was diagnosed with a nasal chordoma from seeding of the surgical pathway. Due to multiple neurologic comorbidities, she was managed nonoperatively throughout her hospital course. At last available follow-up, the patient was discharged to a long-term acute care facility due to ventilator dependence.

DISCUSSION

Chordomas are histologically low-grade neoplasms with an indolent and slow-growing nature, but can be significantly invasive locally.2,4 They often do not present clinically until late stages of disease and have a high rate of recurrence after treatment.2,4 Local recurrence is the most common form of treatment failure, occurring in 21-29% of patients, and is considered the most important predictor of mortality.3,4 Distant metastasis is uncommon except in very late-stage diagnosis, and tends to have a lesser effect on patient survival than does the local growth of tumor.2,4

Tumor recurrence from seeding along the pathway of prior surgical access is becoming more recognized as a mechanism of treatment failure. In a review of several case series, the combined incidence of tumor recurrence from seeding of the surgical pathway was reported in 1.9% of patients after surgery +/- radiotherapy.3

Clival chordomas, which represent about one third of cases, are especially difficult to manage because the surgical access, resectability, and delivery of radiotherapy are limited due to proximity to vital anatomical structures.3 The current accepted treatment of intracranial chordomas at most large cancer centers is maximal tumor resection while preserving neurologic function followed by postoperative proton-beam therapy.3,4

REFERENCES