INTRODUCTION

Glomus jugulare tumors are rare, slow-growing, hypervascular tumors that arise within the jugular foramen of the temporal bone. They are included in a group of tumors called paragangliomas, which include carotid body, glomus vagale, and glomus tympanicum tumors. These tumors have their embryologic origin in neural crest cells. The Fisch classification of glomus tumors is based on extension of the tumor to surrounding anatomic structures and is closely related to mortality and morbidity.

Type A tumor - Tumor limited to the middle ear cleft (glomus tympanicum)

Type B tumor - Tumor limited to the tympanomastoid area with no infralabyrinthine compartment involvement

Type C tumor - Tumor involving the infralabyrinthine compartment of the temporal bone and extending into the petrous apex

Type C1 tumor - Tumor with limited involvement of the vertical portion of the carotid canal

Type C2 tumor - Tumor invading the vertical portion of the carotid canal

Type C3 tumor - Tumor invasion of the horizontal portion of the carotid canal

Type D1 tumor - Tumor with an intracranial extension less than 2 cm in diameter

Type D2 tumor - Tumor with an intracranial extension greater than 2 cm in diameter. The optimal treatment of these tumors has evolved over time. Initially, complete surgical excision of these tumors was advocated. Due to morbidity and mortality concerns, radiation therapy had become more common. Most recently, combined modality therapy has become more widely accepted.

METHODS AND MATERIALS

A 61 year old Caucasian female presented with dizziness, ataxia, right sided facial weakness, pulsatile tinnitus, pain, and decreased hearing on the right for several months. MRI revealed a 3.5 x 1.5 x 2.6 cm enhancing lesion of the right temporal bone. The patient underwent preoperative embolization followed immediately by a subtotal excision of this tumor through translabyrinthine/retro-sigmoid combined approach. Pathology revealed paraganglioma with 10% of the tumor cells positive for Ki67 stain. The patient did well postoperatively and underwent postoperative proton beam radiation fractionated to a total of 45 Gy. Results: A comprehensive review of the literature revealed only one other case in the medical literature involving subtotal resection of a large glomus tumor followed by proton beam radiation. Conclusions: 1) Proton beam therapy may be beneficial in the postoperative treatment of patients with subtotally resected large skull base glomus tumors; 2) prospective studies of this approach are indicated; and 3) comparison of similar patients undergoing conventional photon therapy would be useful.

RESULTS

The patient underwent preoperative embolization followed the same day by a subtotal excision of this tumor through a combined translabyrinthine/retro-sigmoid approach which also involved transposition of the facial nerve and neck exploration with control of major vessels. The facial nerve was completely dehiscent and embedded in tumor throughout the vertical segment. Surprisingly, the patient’s minimal lower division facial weakness was unchanged postoperatively and after proton radiation. Pathology revealed paraganglioma with 10% of the tumor cells positive for Ki67 stain (Fig. 2). The patient did well postoperatively and underwent postoperative fractionated proton beam radiation 2 Gy per day for 35 fractions to a total of 70 Gy. The patient continues to do well after 6 month follow up. A comprehensive review of the literature revealed only one other case in the medical literature involving subtotal resection of a large glomus tumor followed by proton beam radiation (5). That case involved a 9 year old female with a 3.5 x 3.5 x 3.7 cm tumor. That patient underwent preoperative embolization, followed the next day by a combined translabyrinthine/retro-sigmoid resection which like the present case also involved transposition of the facial nerve and neck exploration with control of major vessels. That case differed in that the mass was resected in its entirety except for a small portion adherent to the carotid artery (5). That patient underwent proton therapy fractionated to a total of 45 Gy.

DISCUSSION

Rivas, et al. (6) assessed trends in the management of Glomus Jugulare over a 40-year period and identified disease related influences, including evaluation of surgical outcomes after total and subtotal resections.

CONCLUSIONS

1) Fractionated proton beam therapy may be beneficial in the postoperative treatment of patients with subtotally resected large skull base glomus tumors;
2) prospective studies of this approach are indicated; and
3) comparison of similar patients undergoing conventional photon therapy would be useful.

REFERENCES