Case Report

Stereotactic and Functional Neurosurgery

Stereotact Funct Neurosurg 2007;85:299–302
DOI: 10.1159/000107364

Published online: August 17, 2007

Treatment of a Facial Nerve Neuroma with Fractionated Stereotactic Radiotherapy

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Key Words
Fractionated stereotactic radiotherapy · Facial nerve neuroma · Cerebellopontine angle · Tumor control

Abstract
Background: Facial nerve neuromas are extremely rare and are often mistaken for acoustic neuromas when located near the vestibular nerve. Usually presenting with facial weakness and hearing loss, facial nerve neuromas of the cerebellopontine angle have commonly been managed by surgery. We present the first reported case of a facial nerve neuroma treated with fractionated stereotactic radiotherapy (FSRT). ***Methods:*** The patient was a 40-year-old woman who presented with tinnitus, dizziness and decreased hearing that was associated with a right intracanalicular mass on magnetic resonance imaging (MRI). She underwent a middle fossa craniotomy only to reveal a facial nerve tumor rather than an acoustic neuroma that was not resected due to the high risk of facial paralysis. Following surgery, her facial function worsened and was associated with tumor enlargement on MRI. She was referred for FSRT and received 54 Gy in daily 1.8-Gy fractions with a prescription isodose line of 90%. ***Results:*** Three months after treatment she had no worsening of her pretreatment symptoms, and at the 1-year follow-up, she experienced facial weakness improvement accompanied by an absence of tumor growth on MRI. These clinical and imaging findings persisted at 48 months of follow-up. ***Conclusion:*** In the first report of a facial nerve neuroma treated with FSRT, this treatment resulted in excellent long-term (4-year) tumor control with improvement of pretreatment symptomatology and absence of morbidity. This report demonstrates the potential for using FSRT to treat facial nerve neuromas of the cerebellopontine angle that could otherwise be associated with significant operative morbidity.

Introduction

Defined as Schwann cell tumors arising from the seventh cranial nerve, facial nerve neuromas are exceedingly rare neoplasms, comprising only 0.8% of all intrapetrous mass lesions with an even lower incidence of tumors that present clinically [1, 2]. Since being described by Schmidt in 1930, approximately 500 cases have been reported in the literature [3, 4]. Facial nerve neuromas can present along any portion of the seventh cranial nerve, resulting in a variety of clinical symptoms and surgical challenges [5, 6]. Most commonly, facial nerve neuromas involve the geniculate ganglion and the tympanic or labyrinthine portions of the facial nerve but can mimic acoustic neuromas when involving the cerebellopon-

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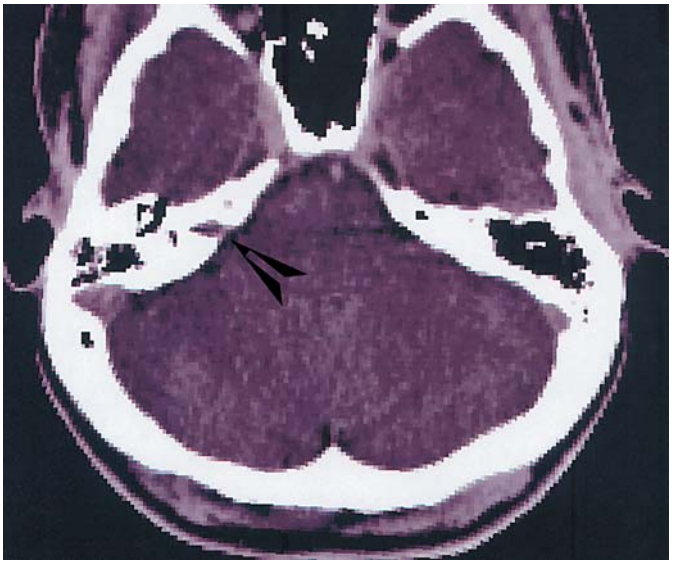

Fig. 1. Axial CT with contrast demonstrating the right facial nerve neuroma (dark arrow) located in the cerebellopontine angle during treatment planning for FSRT.

tine angle or internal auditory canal [7–9]. Although the definitive treatment of facial nerve neuromas is surgical resection, the timing of surgery is controversial, due to the inevitability of a postoperative House-Brackman grade III facial palsy [10, 11]. However, there is an increased risk of hearing loss if prolonged observation is instituted [12, 13]. Recently, fractionated stereotactic radiotherapy (FSRT) has been demonstrated as an efficacious treatment modality for acoustic neuromas of the cerebellopontine angle or internal auditory canal with minimal morbidity [14–16]. We present the first report of a cerebellopontine angle facial nerve neuroma treated with FSRT.

Clinical Materials and Methods

Patient History

Our patient was a 40-year-old woman with no significant medical history who began to experience tinnitus, dizziness and decreased hearing in her right ear 3 years prior to presentation. The magnetic resonance imaging (MRI) scan revealed an enhancing right intracanalicular lesion located in the internal auditory canal, and an audiogram confirmed a 20- to 30-dB decrease in hearing. Due to the imaging findings and the audiogram results, the presumptive diagnosis of a right acoustic neuroma was made. Given her young age and the size and location of the tumor, surgical resection of the lesion via a middle fossa approach was recommended in order to provide the best chance of hearing preservation.

A middle fossa craniotomy was performed 1 month later. The temporal bone was drilled to expose the internal auditory canal, after which the dural sheath covering the facial nerve was opened. Upon opening the sheath, the complex of cranial nerves 7 and 8 were immediately visible, and following microsurgical dissection it became apparent that the facial nerve and not the acoustic nerve was enlarged and contained the neuroma. The superior and inferior vestibular nerves and the cochlear nerve were identified as normal. Due to the risk of causing a permanent facial paralysis with attempted tumor removal, the decision was made to abort the surgical procedure.

Following surgery, the patient did well until the 2nd postoperative day, when she developed a House-Brackmann grade V right facial nerve palsy, which improved to a grade II to III 1 week later. Subsequently, she was followed with annual MRI scans to monitor the size of her lesion. Her right-sided hearing remained stable for 16 months until it worsened and was associated with decreased right eye lacrimation, decreased right eye closure and the new onset of a right facial twitch. Four months later, she experienced paralysis of her right face, and a subsequent MRI revealed growth of the tumor. On examination, she was deaf in the right ear, and the right facial nerve palsy had become a House-Brackmann grade IV, manifesting as an asymmetric droop to the right mouth, inability to close the right eyelid and inability to puff out her cheeks due to loss of air from the right side of her mouth. There was no lack of sensation on either side of the face.

At this point, she was considered for microsurgery, single-dose stereotactic radiosurgery or FSRT. After an informed consent, the patient decided to proceed with FSRT with the understanding that it would be unlikely that she would regain her right-sided hearing after treatment but that treatment would likely arrest the progression of her facial palsy without causing further damage of the facial nerve.

FSRT Treatment

All treatments were overseen by the neurosurgeon, radiation oncologist and medical physicist. Following a standardized testing simulation, the patient had FSRT treatment planning for her right facial nerve neuroma. After placement of a Gill-Thomas-Cosman relocatable headframe for immobilization (Integra Radionics, Burlington, Mass., USA), a CT scan with contrast was obtained (fig. 1) and planning was performed using the Radionics X-Knife 4.0 three-dimensional planning system (Integra Radionics). The size of the tumor was 1.0 × 1.0 × 0.6 cm, which was easily encompassed with a single 1.4-cm collimator. The patient received daily fractions of 1.8 Gy until a total target dose of 54 Gy was administered. The radiation was prescribed to the 90% isodose line using 5 rotational arcs with 300° of rotation, and an average dose of 0.016 Gy was delivered to the brainstem per daily fraction. Thirty treatments were stereotactically delivered over 6 weeks using a dedicated Varian 6/100 linear accelerator (Varian Medical Systems, Inc., Palo Alto, Calif., USA). The patient tolerated the procedure well, with no immediate or delayed posttreatment complications.

Posttreatment Course

Three months after treatment, the patient returned for serial follow-up with neurosurgery and radiation oncology, where she was noted to have no worsening of her pretreatment symptoms. MRI 6 months after treatment revealed the tumor to be stable in

size, with an area of central necrosis within the tumor. At the 1-year follow-up, the patient began noticing a small amount of improvement in her right-sided facial droop and improved control of her right eyelid. MRI scans 2 and 3 years after treatment revealed continued stability in tumor size, which was associated with gradually improving right-sided facial weakness. One year later, the patient reported that she was doing 'exceptionally well', with continuing improvement of her facial weakness and no clinical evidence of disease progression. At 48 months after treatment, the patient continues to demonstrate improvement of her facial weakness (House-Brackmann grade II to III) and stable tumor size on MRI.

Discussion

Most commonly presenting with facial weakness and hearing loss, facial nerve neuromas can occur at any age and have no particular gender predilection [4, 7, 17]. When occurring in the cerebellopontine angle or internal auditory canal, facial nerve neuromas can clinically mimic acoustic neuromas and are often not distinguished until operative intervention, occasionally distinguishable noninvasively by subtle findings on gadolinium-enhanced MRI [8, 18, 19]. Although the definitive surgical approach for these tumors involves a middle cranial fossa or retrosigmoid approach, the morbidity involved in removing the tumor from the involved nerve remains significant, even following facial nerve reconstruction using the sural or greater auricular nerve as a graft [10, 17, 20–22].

Over the past 2 decades, the predominant treatment modality for small (maximum diameter <3 cm) acoustic neuromas has shifted away from microsurgery to stereotactic radiosurgery, with FSRT recently emerging as a promising treatment modality for this disease [15, 16, 23–25]. Given the similarity in location between small acoustic neuromas and cerebellopontine angle facial nerve neuromas of similar size, it would be logical to surmise that there should be a role for stereotactic radiosurgery or FSRT for facial nerve neuromas of the cerebellopontine angle. This concept has already been applied with regard to stereotactic radiosurgery, whereby 2 recent reports demonstrated tumor control of facial nerve neuromas after they had been treated with focused radiation following subtotal resection [26, 27].

The present report details the use of FSRT for a cerebellopontine angle facial nerve neuroma. The choice of FSRT over stereotactic radiosurgery for this patient was derived primarily from the existing literature of both modalities with regard to treatment of acoustic neuromas, which demonstrated FSRT to have less permanent trigeminal nerve morbidity than stereotactic radiosurgery with comparable efficacy [14–16, 25, 28–31]. The results in our patient demonstrated excellent 4-year long-term tumor control associated with improvement in facial nerve function and no treatment-related morbidity. FSRT should be seriously considered in appropriately selected facial nerve neuroma patients as an alternative to microsurgery, due to its efficacy and reduced risk of cranial nerve morbidity compared to microsurgery of the skull base [25, 28, 29].

Conclusion

In the first report of facial nerve neuroma treated with stereotactic radiotherapy, FSRT resulted in excellent long-term tumor control (4 years) with improvement in pretreatment symptoms and no morbidity. This report demonstrates the potential for stereotactic radiotherapy in treating facial nerve neuromas of the cerebellopontine angle that could otherwise be associated with significant operative morbidity. The low risk of cranial nerve morbidity generally associated with FSRT of the skull base in comparison with microsurgery should make FSRT a serious consideration for situations where facial nerve neuromas can be managed either partially or completely in a noninvasive manner.

Acknowledgments

We would like to thank Drs. Bruce Gerbi, Samuel Levine and Eric Nussbaum for invaluable assistance.

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