Case Report

Delayed pseudoprogession of a vestibular schwannoma postradiosurgery

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Abstract

Radiosurgery (RS) can offer excellent local control in the management of both benign and malignant tumors measuring less than 3 cm in size. A known late complication of radiosurgery is radiation necrosis which generally occurs within 6-18 months following treatment and has an increased risk of occurrence with higher radiation doses. The lower dose used to treat vestibular schwannomas (VS) makes this complication less frequent. Tumors that do not respond to radiosurgery and continue to grow may require surgical intervention. We report a case of a young male who received radiosurgery (18 Gy in 3 fractions) in February 2016 for a recurrent VS following initial debulking surgery in 2008. Follow-up imaging revealed an interval decrease in size by May 2017; however, by April 2018, there was a significant interval increase in the cisternal components of the tumor. By September 2018, the lesion had increased by >50% (to a size of 29 mm) compared to May 2017. The patient agreed to undergo repeat surgical debulking. Upon review of the preoperative MRI, the cisternal component of the tumor had substantially decreased in size. Although uncommon, this reflects delayed, pseudoprogession which, in our case, was self-limiting. This raises a question regarding when to proceed with surgical intervention of growing VS following radiosurgery given the potential for delayed resolution of radiation necrosis and demonstrates a gap in our current literature involving surgery of VS following radiosurgery.

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Background

Vestibular schwannomas (VS) are benign tumors arising in the internal auditory canal that represent approximately 6% of all intracranial tumors with an annual incidence of 1 in every 100,000 [1]. VS can be managed by several strategies including observation for small tumors, surgery for larger symptomatic tumors, or radiosurgery for small to mid-sized tumors demonstrating growth.

Following radiosurgery, some lesions undergo pseudoprogression, or a transient increase in size followed by stability or regression. Here, we present a case of pseudoprogression occurring much later than is typically reported in the literature.

Case presentation

In 2008, a 29-year-old healthy male presented with headaches, nausea, and ataxia. Magnetic resonance imaging (MRI) of the brain revealed a 3.6 cm VS in the left internal auditory canal (Fig. 1). The patient underwent surgical debulking that was complicated by a complete facial palsy. Postoperative imaging demonstrated a small residual tumor (Fig. 2). The patient remained clinically stable until 2015. However, follow-up imaging in 2014 and 2015 revealed an increase in the size of the residual tumor to 18 × 8 mm (Fig. 3). As a result of the apparent
serial growth of the residual component on imaging, his case was presented at multidisciplinary rounds and the consensus was to offer radiosurgery. He underwent robotic radiosurgery on CyberKnife in March 2016, at a dose of 18 Gy in 3 Fractions (Fig. 4).

Over the subsequent 2 years, he remained clinically stable with no new neurologic deficits. The VS had decreased in size by May 2017. An MRI in April 2018 demonstrated an interval increase in the size of the tumor up to 21 mm in maximum diameter. Close surveillance imaging in September 2018 revealed a further increase in the size of the residual tumor up to a maximum diameter of 29 mm in size (Fig. 5).

He was evaluated by his neurosurgeon and consented to repeat surgical debulking of the tumor. An MRI for surgical planning was ordered and revealed a spontaneous regression of the tumor down to 26 mm in maximum diameter from 31 mm. The tumor remained stable between January and August 2019 at approximately 23 mm in maximum diameter, and the patient was placed back on routine surveillance imaging (Fig. 6).

Discussion

Radiosurgery as a primary treatment modality for VS has been a topic of much debate over the last 2 decades. Multiple studies have been conducted to identify the efficacy and long-term outcomes with radiosurgery, surgery, or observation as management options. A recent review of radiotherapy for VS indicates stereotactic radiosurgery offers similar local control (>90%) when compared to surgery [2].

One of the challenges associated with radiosurgery for VS is the interpretation of follow-up MRI. Radiologic stability may not be the best surrogate of treatment success since radiation-related imaging changes can make the accurate measurement of post-treatment tumor volume difficult. Pseudoprogression...
can occur postradiosurgery and is defined as a transient post-treatment tumor enlargement due to necrotic/inflammatory changes. In a series by Patel et al., with a median follow-up of 72 months, 21.7% of patients had enlarged imaging abnormalities at last follow-up but only 1.8% of patients required intervention for a progressive process suggestive of tumor growth [3].

In a study by Hayhurst et al., of 75 patients treated with radiosurgery for VS at the University of Toronto, 65% of patients demonstrated tumor stability or regression and 23% had tumor pseudoprogression [4]. Of the patients that experienced pseudoprogression, the increase in tumor size occurred between 6 and 12 months after radiosurgery. Régis et al., have previously reported their experience of radiosurgery for VS noting that a significant percentage of their patients demonstrated transient tumor growth without any predictive value for long-term tumor control [5]. Based on years of experience and case reports, some have advocated that transient growth induced by radiosurgery, followed by stabilization or decrease, can occur as late as 3-4 years after treatment [6].

We recently analyzed 120 patients treated with robotic radiosurgery using CyberKnife (18 Gy in 3 fractions) at our institution with a minimum 4 years’ follow-up [7]. Pseudoprogression occurred in 6% of patients. The average volume increase was 11% and the median time of pseudoprogression was 5 months. The median time to subsequent regression to pretreatment size was 14.7 months (range 10-23 months). No patient with pseudoprogression developed local late recurrence.

In the present case, the patient developed radiologic enlargement of the cystic component of the VS between 25 and 31 months postradiosurgery. This falls outside the “typical” timeframe in which pseudo-progression would be anticipated. This phenomenon could foreseeably result in unnecessary surgical intervention. This case highlights the need for further research into differentiation between what is truly tumor progression and what is delayed radiation change, particularly in circumstances where radiographic changes fall outside of our anticipated timeline for pseudo-progression. Longer radiologic follow up and close clinical surveillance is needed in these cases to help distinguish late radiosurgery responders from true treatment failures.

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