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Case Report

Relying too much on upfront radiosurgery: Indolent course misinterpreted as effectiveness of radiosurgery in a case of skull base chondrosarcoma

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Received : 08 December 2019
Accepted : 23 April 2020
Published : 16 May 2020

DOI
10.25259/SNI_590_2019

Quick Response Code:

ABSTRACT

Background: Skull base lesions are still considered surgically challenging and primary gamma knife radio surgery (GKRS) is gaining popularity. However the effectiveness of GKRS may be overrated especially in lesions with indolent course.

Case Description: We report a case of chondrosarcoma, mimicking a trigeminal schwannoma treated with upfront radio surgery. Relatively lower dose was administered in view of proximity to the brainstem. The patient was asymptomatic and the size of the lesion remained static for over a decade. This was misinterpreted as effectiveness of GKRS. The lesion grew after a decade necessitating surgery.

Conclusion: With popularity of upfront GKRS, suboptimal but maximal safe radiation dose is usually prescribed for lesions close to critical structures like brainstem. In these cases the long indolent natural course of the pathology, as in the case of chondrosarcoma may be misconstrued as success of radiosurgery. An extended follow up beyond this static period is necessary before concluding its effectiveness.

Keywords: Chondrosarcoma, Gamma knife radiosurgery, Indolent course, Skull base

INTRODUCTION

Skull base chondrosarcomas are slow-growing tumors close to the brainstem and they often present with cranial nerve deficits.[2] Radiosurgery following surgical decompression is currently practiced. However, the long-term behavior of chondrosarcoma following radiosurgery is less discussed. We report a case of chondrosarcoma mimicking trigeminal schwannoma on radiology that was initially treated with upfront gamma knife radiosurgery (GKRS). The tumor remained static for a decade after radiosurgery and then changed its morphology, size, and radiological character necessitating excision.

We have discussed the natural course of chondrosarcoma and 5th nerve schwannoma close to the brainstem. The importance of surgical debulking before radiosurgery has been highlighted. The disadvantages of relying on suboptimal radiation, due to their proximity to brainstem, especially without a tissue diagnosis of such lesions, have been emphasized.

CASE REPORT

A 50-year-old otherwise healthy lady presented with complains of mild headache and diplopia for 8 weeks duration. On examination, she had the right lateral rectus palsy with diminished right corneal reflex. Radiology showed lesion extending from the cerebellopontine angle to the middle cranial fossa straddling across the petrous apex with its erosion [Figure 1a-d]. There were mild ventriculomegaly and early papilledema for which a ventriculoperitoneal shunt was placed. Her headache improved and diplopia disappeared, though the sixth nerve paresis was apparent on examination.

The patient was offered surgery. However, she opted for upfront GKRS with presumptive diagnosis of trigeminal schwannoma. As the lesion was close to the brainstem, total dose of 25 Gy was administered. The patient was regularly followed up and showed no symptoms for a decade. Sequential MRI showed no increase in size or change in character of lesion [Figure 1e-h]. However, in the past 6 months, she presented with features of pseudobulbar palsy (severe ataxia, difficulty in swallowing, change in voice, and spasticity) and the radiology showed significant increase in size of the lesion with brainstem compression and heterogeneous contrast enhancement [Figure 2a-d].

She was operated through the right temporal craniotomy and interdural approach (by senior author PS). The tumor was grayish, fleshy with mild vascularity with a plane from the fifth nerve fascicles. Through the expanded Meckel's cave, the component from posterior fossa was excised. Histopathology confirmed chondrosarcoma Grade II [Figure 3]. Tumor cells were positive for S-100 and vimentin [Figure 4]. The patient improved clinically and doing well at 8-month follow-up.

DISCUSSION

Radiosurgery is gaining popularity in treating skull base neoplasm such as schwannomas and chondrosarcomas.[1,4] These lesions may present with features of cranial nerve involvement due to mechanical compression.

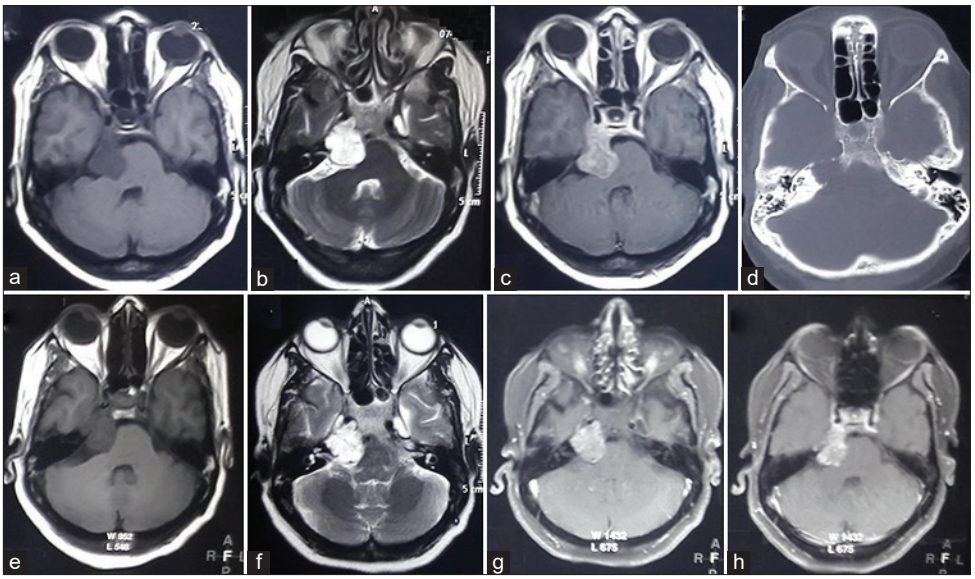

Figure 1: Upper row: MRI showing tumor straddling across the petrous apex from middle fossa to the posterior fossa. (a) Tumor is hypointense on T1, (b) hyperintense on T2, and (c) showing contrast enhancement. (d) Note the bony erosion of the petrous apex without any calcification. Lower row: (e-h) serial MR images in the follow-up showing tumor with similar size on volumetric analysis.

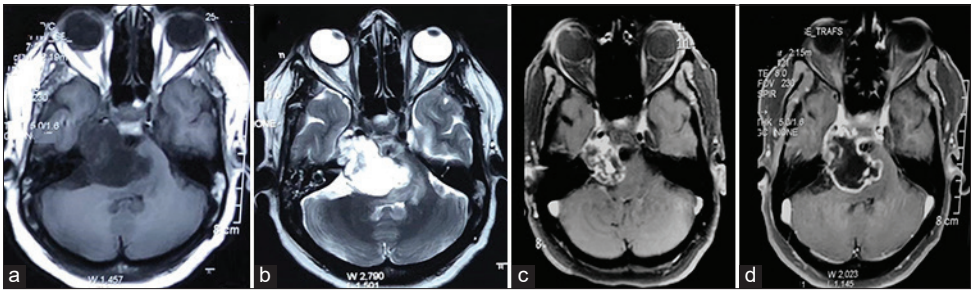

Figure 2: (a) MR images showing enlargement of tumor on T1, (b) T2, (c and d) sequences with areas of cystic changes and irregular contrast enhancement.

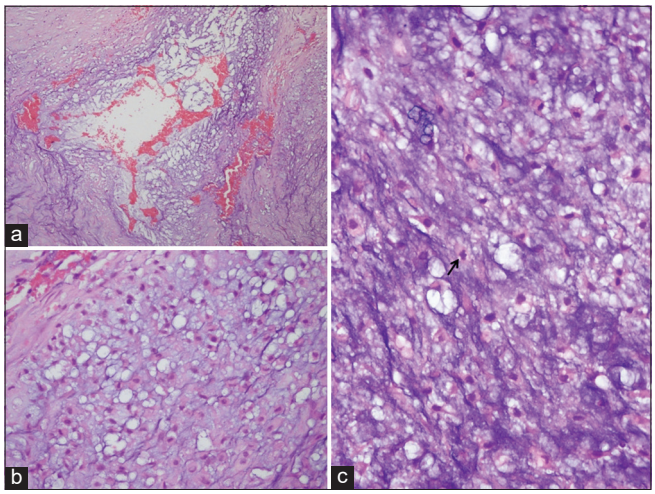

Figure 3: (a) Low magnification demonstrating tumor arranged in lobules with a prominent chondromyxoid matrix (H&E ×100); (b) high magnification demonstrating closely packed lacunae containing mononucleate and few binucleate cells with pleomorphic nuclei (H&E ×400); (c) malignant chondrocytes with hyperchromatic nuclei with a prominent chondromyxoid matrix. Occasional mitoses are noted (arrow) (H&E ×400).

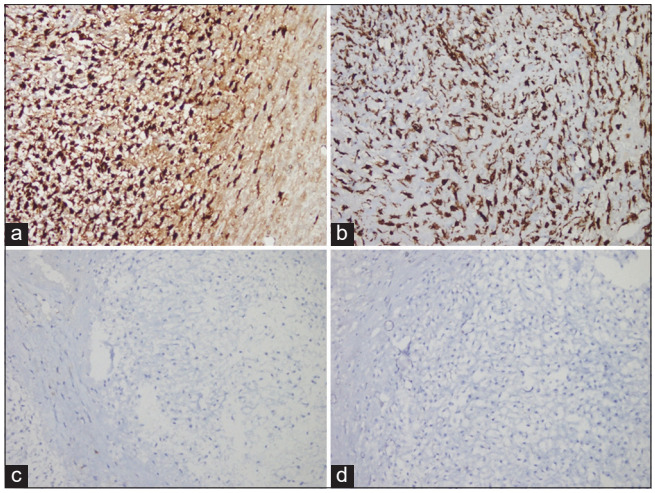

Figure 4: (a) Diffuse and strong nuclear immune reactivity for S-100; (b) tumor cells were positive for vimentin while were negative for EMA (c) and pancytokeratin (d), (a-d) immunoperoxidase (×200).

Because of their common anatomical location around the petrous apex, they may have overlapping clinical features. At times, it is difficult to make accurate diagnosis on the basis of clinical feature and radiology. Upfront GKRS is a viable treatment option for moderate size lesions without significant mass effect. Efficacy of radiosurgery is well documented for skull base chondrosarcomas. The 10-year progression-free survival following radiosurgery is over 70%.[1] The recommended dose for chondrosarcoma is over 30 Gy. However, at times, it is difficult to administer this dose, especially when the tumor is located in proximity to critical structures.[3] In such cases, relatively lower doses have been administered. Koga *et al*. have diligently described the long-term outcome of chondrosarcoma following radiosurgery. One of their patients treated with suboptimal dose, had local relapse 100 months after GKRS.[3] Hence, chondrosarcomas may remain silent both clinically and radiologically for a prolonged period and it would be wrong to consider the static disease in 5–10 years following GKRS as an effective treatment.

In the present case, one of our differentials was trigeminal schwannoma. The marginal prescription dose for these tumors is 12–15 Gy.[4] The 5-year tumor control rate for these tumors is over 80%.[5] Most of the tumor recurrences have been documented within 5 years of therapy and static tumors beyond this time frame may be considered as effectiveness of GKRS.[4] In the present case, as the tumor remains silent clinically and radiologically for over a decade, we presumed it as an effective treatment. The final diagnosis of chondrosarcoma was made only after the surgical excision 10 years after radiosurgery. In the present case, the lesion was treated with 25 Gy with a presumptive diagnosis of trigeminal schwannoma, which may be considered as adequate dose treatment. However, the dose is suboptimal for chondrosarcoma. Even with presumptive diagnosis of chondrosarcoma with proximity to the brainstem, it would not have been possible to administer dose beyond 25 Gy. In retrospect, the indolent course of chondrosarcoma was mistaken as effectiveness of gamma knife to keep the disease static.

This case report exemplifies the variable course of chondrosarcoma. The tumor may remain radiologically static following radiosurgery but may not inactive. The indolent phase of such tumors may be long enough and should not be mistaken as effectiveness of radiosurgery. Adjacent critical neural structures like brainstem may restrain the dose of gamma knife. A relatively high marginal dose of 15 Gy is needed for long-term tumor control.[3] Although skull base lesions may be difficult to access surgically, their decompression is essential not only to establish the diagnosis but also for giving adequate radiation dose. Stereotactic radiosurgery without diagnosis and debulking may not be effective, especially in lesions close to the brainstem with long indolent courses.

CONCLUSION

Rather than resorting to the suboptimal dose for skull base lesions close to the brainstem, surgical debulking should be considered. This not only establishes the diagnosis but also reduces the tumor volume for an effective radiation dose. The indolent course of the lesion must not be misinterpreted as static disease and effectiveness of GKRS.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Malik P, Sahoo SK, Gupta K, Salunke P. Relying too much on upfront radiosurgery: Indolent course misinterpreted as effectiveness of radiosurgery in a case of skull base chondrosarcoma. Surg Neurol Int 2020;11:112.