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CASE REPORT

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Skull base aneurysmal bone cyst presenting with hydrocephalus: progressive residuum obliterated by Gamma Knife stereotactic radiosurgery in a pediatric patient

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Aneurysmal bone cysts (ABCs) are an uncommon entity predominantly encountered in the pediatric population. The skull is rarely involved, but these cysts have been reported to arise in the skull base. Traditional treatment has been with surgery alone; however, there is a gathering body of literature that reports alternative treatments that can achieve long-term disease-free survival. However, these therapies are predominantly directed at peripheral skeletal lesions. To the authors' knowledge, this report is the first to describe long-term follow-up of the efficacy of Gamma Knife stereotactic radiosurgery for treatment of ABC residuum in the skull base that resulted in long-term patient stability and likely ABC obliteration.

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KEYWORDS aneurysmal bone cyst; hydrocephalus; skull base; Gamma Knife stereotactic radiosurgery

Aneurysmal bone cysts (ABCs) are uncommon lesions predominantly presenting in children and young adults, with a median age of 13 years at presentation.[1] ABCs are nonneoplastic expansile osteolytic lesions of unknown etiology and are generally solitary. There is a slight female preponderance, with a predilection for the metaphysis of long bones such as the femur; however, ABC formation sites may occur anywhere in the skeleton. Patients typically present with pain and swelling, and ABC can predispose patients to pathological fracture. Traditional treatment has been with surgery alone; however, there is a gathering body of literature describing alternative treatments that can achieve long-term disease-free survival. It is critical to confirm a histological diagnosis of an ABC in addition to identification of a radiologically hemorrhagic "fluid-fluid level" within cystic cavities. Although this radiological fluid-fluid level finding is a classical sign of ABC, it can also be seen in malignant lesions such as telangiectatic osteosarcoma, chondroblastoma, malignant fibrous histiocytoma, and any necrotic bone tumor, among others.[2] ABC can occur secondary to other bone tumors, including chondroblastoma, giant cell tumor, chondromyxoid fibroma, nonossifying fibroma, and fibrous dysplasia.[3]

Here we present the case of a pediatric patient ini-

ABBREVIATIONS ABC = aneurysmal bone cyst; GKSRS = Gamma Knife stereotactic radiosurgery.

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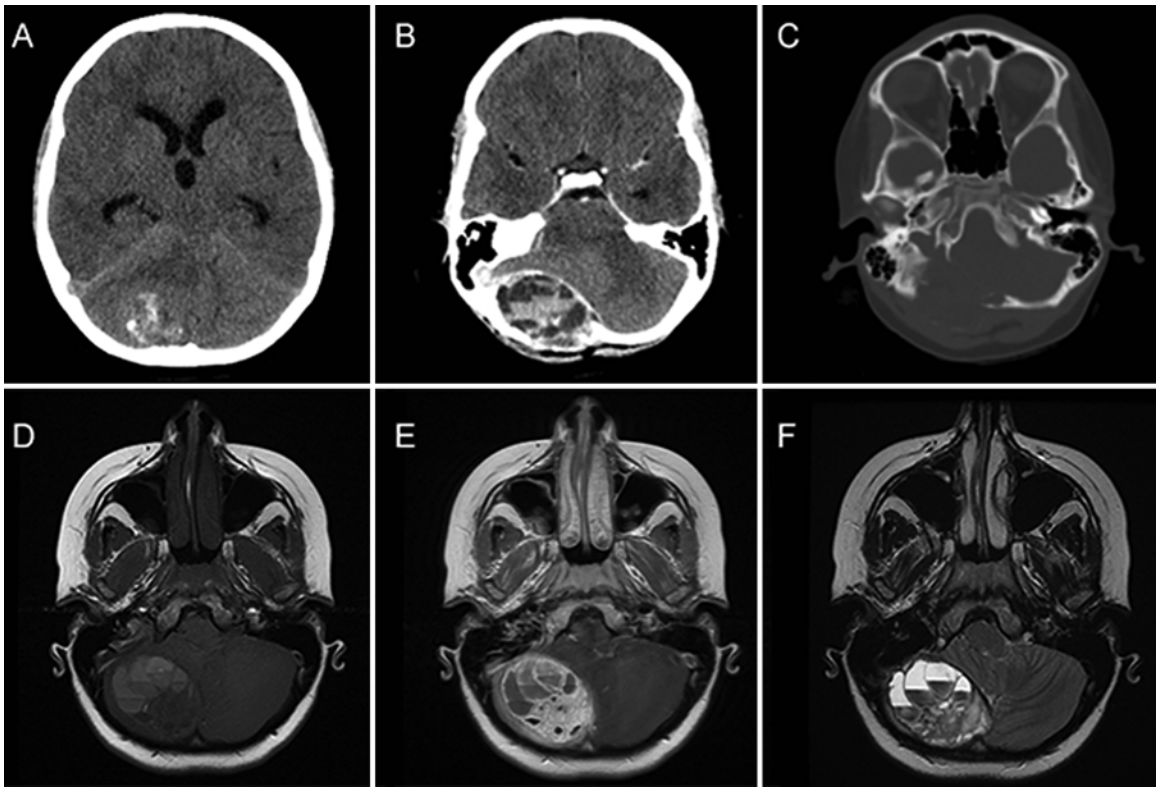

FIG. 1. Axial preoperative imaging. **A:** Unenhanced head CT demonstrating a mass with internal hyperdensity in the posterior fossa compressing the cerebellum, effacing the fourth ventricle, and causing obstructive hydrocephalus. **B:** Enhanced CT confirms a solid cystic lesion with fluid-fluid levels. **C:** A bone window on unenhanced CT confirms that the lesion is centered in the occipital bone. **D:** An unenhanced T1-weighted image demonstrates fluid-fluid levels suggesting a lesion with multiple cysts containing blood products of varying ages. **E:** A contrast-enhanced T1-weighted image confirms avid enhancing soft tissue surrounding the cysts. **F:** A T2-weighted image again confirms a lesion with fluid-fluid levels centered in the occipital bone.

tially treated with subtotal surgical resection, which was followed by Gamma Knife stereotactic radiosurgery (GKSRS) to a progressive ABC residuum. Eight years after initial treatment the patient has excellent quality of life with no neurological deficits.

Case Report

History and Examination

An 11-year-old girl presented with a 4-week history of headache, which was worse in the morning and had recently began to wake her at night. Clinical examination revealed incoordination and nystagmus on the extreme right gaze but no other focal neurological symptoms. Fundoscopy demonstrated blurred disc margins but no frank edema. CT followed by MRI revealed a large extraaxial tumor centered in the occipital bone causing marked cerebellar compression, effacement of the fourth ventricle, and obstructive hydrocephalus (Fig. 1). The lesion demonstrated fluid-fluid levels with avidly enhancing solid cystic components. Preceding urgent surgery the patient developed episodes of bradycardia and apnea.

Operation

The patient was placed prone in the Mayfield position. A right paramedian incision was performed and then a right suboccipital craniectomy. Large superficial veins were encountered and controlled. The tumor was easily localized because it was invading the bone with a good plane between the tumor and the muscle. The tumor, along with a cuff of surrounding normal bone, was resected from the occipital bone. The tumor was peeled off the dura without difficulty, with an easily dissectable plane throughout that allowed preservation of the dura and allowed the underlying cerebellum to be left undisturbed. The bone was resected as far laterally as the mastoid air cells, and the right transverse and sigmoid sinuses were identified and preserved. Despite careful dissection, significant hemorrhage was encountered at the most anterior and lateral aspect of the tumor; however, control was gained with absorbable hemostat material (oxidized regenerated cellulose) and injectable liquid hemostatic matrix (gelatin thrombin). The right vertebral artery was identified and completely preserved. The dura was intact at the end of the procedure, but attempted CSF diversion with extraventricular drain placement was abandoned as the right frontal ventricle could not be cannulated (presumably due to the resolution of hydrocephalus). Layered closure was performed with absorbable polyglactin 910 sutures. A small tumor residuum was suspected given the tumor location and its complex association with temporal-occipital neurovascular structures, which were completely preserved.

Pathological Findings

The mass extended above and below the skull surface and consisted of cystic areas filled with blood and extensive firm pale rubbery tissue with focal calcification. The lesion was well circumscribed with blood-filled spaces separated by connective tissue septa. The stroma consisted of short spindle-shaped cells of varying cellularity and occasional osteoclast-type giant cells. The spindle cells were of bland appearance. There were scattered mitotic figures but no atypical form. There was direct formation of bone and a chondroid-type matrix by lesional cells. The new bone was woven, with irregularly arranged collagen fibers, rimmed by osteoblasts, and deposited in a linear pattern. In many areas a characteristic "blue bone matrix" was found. There were adjacent reactive changes in the cortical bone. The observed appearance was consistent with an ABC with a marked solid component.

Gamma Knife Stereotactic Radiosurgery

The patient's initial postoperative course was unremarkable, with no focal neurological deficits. However, the patient complained of diplopia 4 months after the initial surgery and had developed rigid right-sided torticollis. Fundoscopy revealed bilateral papilledema, and a progressive tumor residuum was identified on early follow-up 4 months after resection (Fig. 2). Clear progressive bone erosion was demonstrated, with definite changes on MRI showing further cyst formation (again demonstrating fluid-fluid levels) and growth of enhancing soft tissue invading the petrous temporal bone, occipital condyle, and clivus. Following multidisciplinary team discussions, GKSRS was administered with curative intent on an urgent basis at 5 months after the initial surgery. Although an ABC is not a vascular malformation per se, cerebral angiography was undertaken to identify any tumoral blush to optimize the GKSRS targeting (Fig. 2). All cystic and pathologically enhancing soft tissue was included in the treatment volume (Fig. 2I). Angiography and stereotactic radiosurgery (Gamma Knife Leksell model 4C) were performed, with the patient under total general anesthesia for 7 hours. A total of 20 Gy at the 50% isocontour was administered. Based on substantial established experience with pediatric arteriovenous malformation treatment, the dose was prescribed after consideration of the patient's age and the treatment volume.[4] The total treated volume was 14.74 cm^3, which was felt to be within acceptable limits based on the site of irradiation being predominantly in the bone and also based on our experience of treating vascular malformations of this volume in pediatric cases.

Postoperative Course

After GKSRS, the patient complained of poor short-term memory, fatigue, and lack of concentration; however, no structural abnormalities were demonstrated on follow-up imaging. Torticollis and diplopia had completely resolved. Long-term radiological follow-up of the patient at 19 years of age, 7 years after surgery and GKSRS, demonstrated stablity in the appearance of the treatment site (Fig. 3). The patient had no neurological deficits and was living independently with good quality of life. No CSF flow diversion was required following the initial surgery. No neurovascular abnormalities were demonstrated at the long-term follow-up despite GKSRS treatment in close proximity to the sigmoid sinus, jugular bulb, and carotid canal. All cranial nerves were functional at the most recent clinical follow-up 8 years after the initial treatment.

Discussion

Surgical resection of a large skull ABC has been described as early as 1977, in a case in which, interestingly, the ABC originated in a location similar to that in our patient.[5] This historic case report documented 3 surgical sessions to achieve a presumed cure; however, the authors did not report any long-term results. Therapy of ABC has evolved, but although en bloc resection is felt to be curative and have the lowest rates of recurrence, the associated morbidity is often high. Currently curettage with or without bone grafting is felt to be the best treatment; however, radiotherapy may have an increasingly important role.[6] Radionuclide ablation has been reported as a therapeutic option, but its use has been reported only in a single-center series.[15] Curettage and local treatment include intralesional resection with a high-speed burr, argon beam coagulation, phenol wash, or cryosurgery.[6] Furthermore, some authors have utilized polymethylmethacrylate cement to augment bone reconstruction for large cavities, particularly in pediatric patients.[8] Arterial embolization is an attractive option in the peripheral skeleton given that a tumor blush can be identified, as was visible in this study, where it was useful for GKSRS localization (Fig. 2G and H). However, in a few patients skin necrosis has occurred with this method, which is a highly undesirable side effect.[9] In addition, the use of arterial embolization in the cranium is potentially hazardous given the intrinsic risk of intracranial reflux from the profuse vascular collateral anastomoses in the skull base. Sclerotherapy with percutaneous doxycycline has also been reported as an alternative minimally invasive method,[6] although this technique has negative aspects similar to those previously described for arterial embolization.

To our knowledge this is the first report of the efficacy of GKSRS for the treatment of ABC residuum in the skull. Adjuvant radiation therapy used in ABC cases has been reported in the literature, but efficacy is difficult to evaluate as this is a rare disease reported only in small series. In the largest reported series of 12 patients receiving external beam radiotherapy as an adjuvant or primary treatment to the peripheral skeleton or spine, the authors reported that 11 patients had no evidence of disease on follow-up.[10] Similarly, in an older North American study of 11 patients with recurrent ABC after surgery, radiotherapy achieved control in all but 1 patient.[11] The most recent European study, with reported treatment experience of more than 30 years, still only documented 10 cases of ABC treated with radiotherapy, with 7 patients obtaining local control and the remainder lost to follow-up.[7] Follow-up durations in many of these studies span a wide range from as short as 12 months to as long as 36 years.[7] Certainly, radiological stability of a nonneoplastic lesion for more than 5 years is considered to predict a long-term benign course and likely indicates curative treatment, such as in our current case.

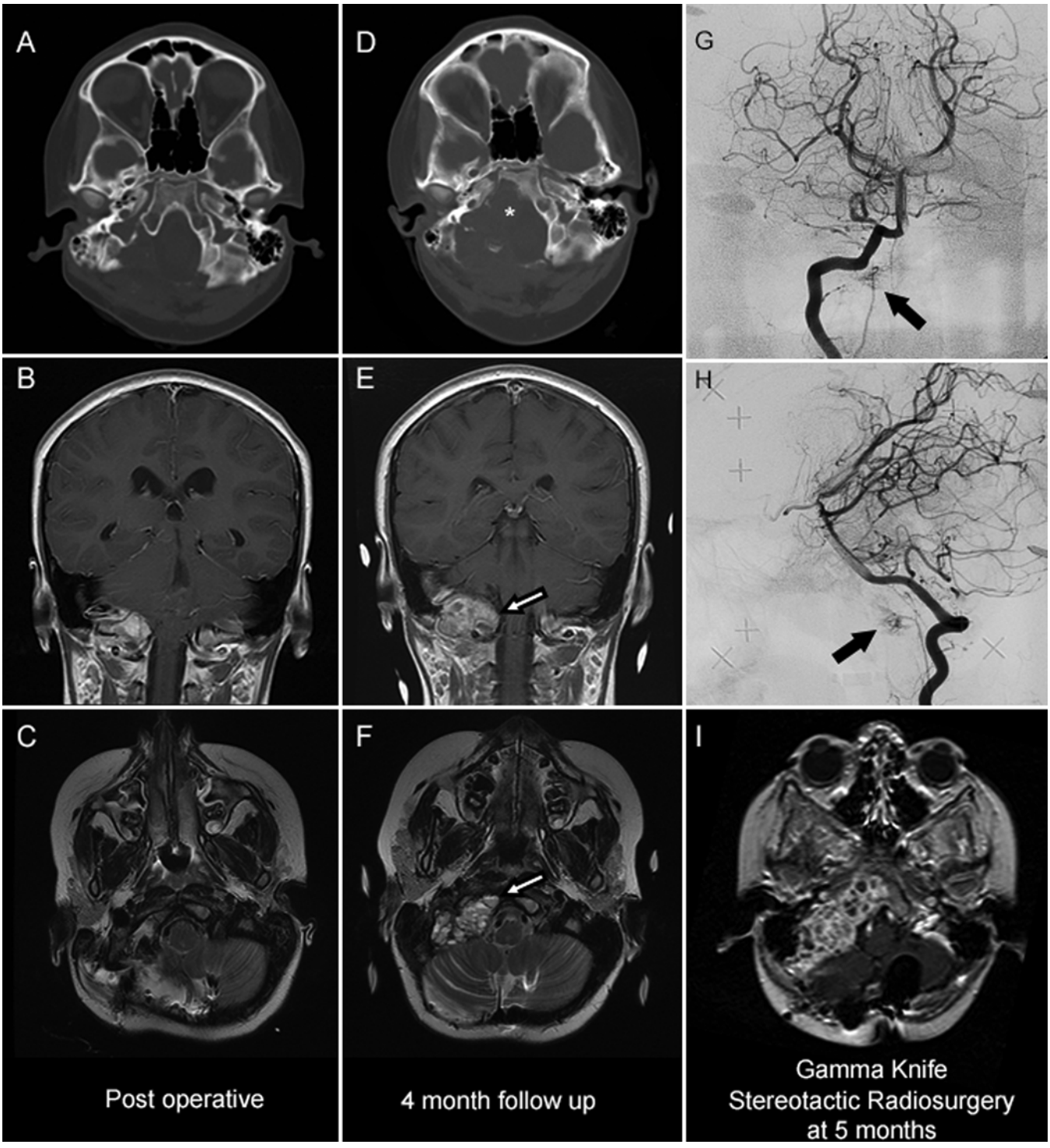

FIG. 2. Postoperative imaging (A–C). Bone window in unenhanced CT (**A**), enhanced T1-weighted MRI (**B**), and T2-weighted MRI (**C**) demonstrating residual abnormality in the petrous temporal bone. Early 4-month follow-up after presentation with new diplopia and torticollis (D–F). **D:** Bone window CT demonstrating progressive erosion of the right occipital condyle and clivus (*asterisk*). **E:** Expanding soft-tissue abnormality in the right occipital condyle (*white arrow*). **F:** The progressive residuum demonstrates new cystic areas with fluid-fluid levels (*white arrow*) similar to those in the presentation lesion. Angiography for GKSRS planning performed at 5 months postsurgery (G–I). **G:** Anteroposterior view of the tumoral blush (*black arrow*). **H:** Lateral view of angiogram again demonstrating tumoral blush (*black arrow*). **I:** Further progression of the avidly enhancing solid cystic lesion into the clivus at the time of GKSRS.

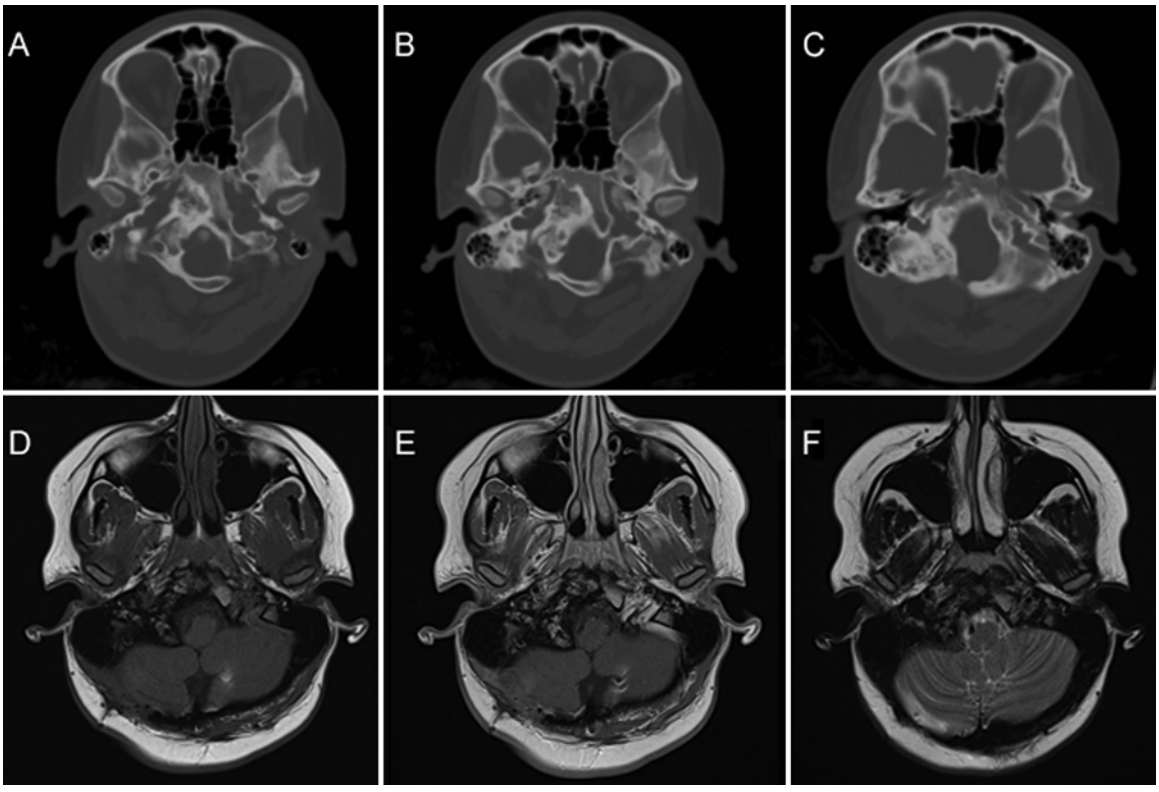

FIG. 3. Follow-up when the patient was 19 years old, at 7 years after surgery and GKSRS. **A–C:** Unenhanced CT bone window serial slices demonstrating new bone formation with no aggressive features in the area of progressive residual ABC treated by GKSRS. **D and E:** T1-weighted precontrast (D) and postcontrast (E) MR images demonstrating normal bone marrow enhancement in the dysplastic temporal bone and occipital condyle. **F:** Normal low T2 signal in cortical bone with no evidence of recurrent fluid-fluid levels.

In our own experience of GKSRS in over 5000 patients treated for both benign and malignant intracranial disease, an elevated risk of secondary tumor has not been found, though the bone was largely not included in the treatment volumes.[12] However, in the present case, similarly to other reported cases, secondary tumor development in GKSRS-treated bone is unlikely.

External beam radiation therapy can be delivered in various forms, including movable linear accelerators (vendors include X-Knife, CyberKnife, and Linac), Gamma Knife (which utilizes 200 converging beams of cobalt-60 gamma radiation), and particle beam therapy (including proton and helium ion beams). GKSRS is an attractive therapeutic option in the cranium and considered the gold standard for intracranial radiosurgery due to its precision in targeting the lesion to be treated and reducing the collateral tissue dose.[13] No reliable randomized comparative study has allowed assessment of the efficacy and safety of different types of linear accelerator delivered treatments compared to GKSRS for malignant skull or brain lesions.[14]

Conclusions

In summary, we believe that this is the first long-term follow-up report of the efficacy of GKSRS for the treatment of ABC residuum that demonstrates long-term stability and likely obliteration, further highlighting the utility of GKSRS in treating complex cranial lesions.

References

1. Leithner A, Windhager R, Lang S, et al. Aneurysmal bone cyst. A population based epidemiologic study and literature review. *Clin Orthop Relat Res*. 1999;(363):176–179.
2. O'Donnell P, Saifuddin A. The prevalence and diagnostic significance of fluid-fluid levels in focal lesions of bone. *Skeletal Radiol*. 2004;33(6):330–336.
3. Bonakdarpour A, Levy WM, Aegerter E. Primary and secondary aneurysmal bone cyst: a radiological study of 75 cases. *Radiology*. 1978;126(1):75–83.
4. Dinca EB, de Lacy P, Yianni J, et al. Gamma knife surgery for pediatric arteriovenous malformations: a 25-year retrospective study. *J Neurosurg Pediatr*. 2012;10(5):445–450.
5. Chalapati Rao KV, Rao BS, Reddy CP, et al. Aneurysmal bone cyst of the skull. Case report. *J Neurosurg*. 1977;47(4):633–636.
6. Park HY, Yang SK, Sheppard WL, et al. Current management of aneurysmal bone cysts. *Curr Rev Musculoskelet Med*. 2016;9(4):435–444.
7. Elsayad K, Kriz J, Seegenschmiedt H, et al. Radiotherapy for aneurysmal bone cysts: a rare indication. *Strahlenther Onkol*. 2017;193(4):332–340.
8. Wallace MT, Henshaw RM. Results of cement versus bone graft reconstruction after intralesional curettage of bone

tumors in the skeletally immature patient. *J Pediatr Orthop*. 2014;34(1):92–100.
9. Rossi G, Rimondi E, Bartalena T, et al. Selective arterial embolization of 36 aneurysmal bone cysts of the skeleton with N-2-butyl cyanoacrylate. *Skeletal Radiol*. 2010;39(2):161–167.
10. Zhu S, Hitchcock KE, Mendenhall WM. Radiation therapy for aneurysmal bone cysts. *Am J Clin Oncol*. 2017;40(6):621–624.
11. Marcove RC, Sheth DS, Takemoto S, Healey JH. The treatment of aneurysmal bone cyst. *Clin Orthop Relat Res*. 1995;(311):157–163.
12. Rowe J, Grainger A, Walton L, et al. Risk of malignancy after gamma knife stereotactic radiosurgery. *Neurosurgery*. 2007;60(1):60–66.
13. Levivier M, Gevaert T, Negretti L. Gamma Knife, CyberKnife, TomoTherapy: gadgets or useful tools? *Curr Opin Neurol*. 2011;24(6):616–625.
14. Canadian Agency for Drugs and Technologies in Health (CADTH). TomoTherapy, Gamma Knife, and CyberKnife therapies for patients with tumours of the lung, central nervous system, or intra-abdomen: a systematic review of clinical effectiveness and cost-effectiveness. *CADTH Technol Overv*. 2010;1(3):e0119.
15. Bush CH, Drane WE. Treatment of an aneurysmal bone cyst of the spine by radionuclide ablation. *AJNR Am J Neuroradiol*. 2000;21(3):592–594.

Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Tse, Radatz, Sinha, Zaki. Acquisition of data: Tse. Analysis and interpretation of data: Tse. Drafting the article: Tse, Jiang. Critically revising the article: Jiang. Reviewed submitted version of manuscript: Tse. Approved the final version of the manuscript on behalf of all authors: Tse. Administrative/technical/material support: Tse.

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