Interdisciplinary Neurosurgery: Advanced Techniques and Case Management 7 (2017) 50–52

Contents lists available at ScienceDirect

Interdisciplinary Neurosurgery: Advanced Techniques and Case Management

journal homepage: www.inat-journal.com

Case Report & Case Series

Calvarial aneurysmal bone cyst associated with fibrous dysplasia: Case report and literature review☆

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ARTICLE INFO

Article history:
Received 10 June 2016
Revised 2 November 2016
Accepted 13 November 2016

Keywords:
Bone cyst
Calvarium
McCune-Albright syndrome
In situ cranioplasty
Cosmesis

ABSTRACT

Aneurysmal bone cysts are benign, expansile bone lesions primary or secondary to other pathology. The majority are seen in long bones and the spine, rarely occurring within the cranial vault. Here we describe the case of a 17-year-old gentleman with McCune-Albright syndrome who developed a right parietal aneurysmal bone cyst in the setting of fibrous dysplasia. The patient was treated with lesion excision and in situ cranioplasty using methyl methacrylate and molded titanium mesh, ultimately rendering excellent cosmetic outcome. Our case report highlights the efficacy of in situ titanium cranioplasties in contouring to the native skull and enabling optimization of cosmesis, specifically through the use of titanium mesh plates with the bone flap as a template.

1. Introduction

Aneurysmal bone cysts are benign lesions comprising less than 2% of bone tumors [1]. Fewer than 5% of these lesions occur in the calvarium, and very few have been described [2–6]. Herein we present the novel case of a patient with McCune-Albright syndrome and known fibrous dysplasia of the parietal bone that secondarily developed a large aneurysmal bone cyst. The lesion was treated with surgical excision and in situ cranioplasty with methyl methacrylate and molded titanium mesh. We review the pathogenesis of aneurysmal bone cysts as well as management and treatment strategies. Of note, we highlight the robust use of this unique in situ cranioplasty technique in achieving excellent skull cosmesis.

2. Case report

2.1. Patient description

A 17-year-old gentleman with McCune-Albright syndrome was found to have a right-sided skull abnormality. CT scan revealed a right parietal bone lesion without brain parenchyma compression (Fig. 1). The patient was asymptomatic and the lesion managed through observation. Three-and-a-half years later, he reported increasing difficulty with spatial awareness and concentration capacity over several months. Imaging revealed cystic expansion of the lesion to 7 cm × 6 cm × 6 cm with considerable compression of the underlying brain parenchyma and a resultant 6 mm midline shift (Fig. 2).

2.2. Surgical procedure and postoperative outcome

Following a U-shaped right temporal-frontal-parietal-occipital skin incision and subsequent scalp flap retraction, an exophytic bone cyst was visualized with soft tissue erosion through cortical surface of the bone. A drill was used to excise the lesion, and the dura was visible and pulsating. Next, titanium mesh was molded to the presumed shape of the right parietal region and screwed into the bone, and the epidural space dissected. Abnormal dura was then opened in a circumferential manner and excised, and samples of abnormal bone and soft cystic tissue sent for pathologic examination. The dural defect was grafted with two pieces of SurgAssist and augmented with fibrin glue.

Molded titanium mesh was reattached to the surrounding craniotomy bone flap and methyl methacrylate was laid into the inner side of the mesh and flattened to approximate the width or the normal bone. Once hardened, in situ cranioplasty was performed. This technique has been described as enabling additional mesh cranioplasty strength after cranial defect repair [2]. The wound was irrigated and vancomycin powder placed over the cranioplasty. The galea was then closed with interrupted 3-0 Vicryl and running 2-0 Vicryl, and skin with running 4-0 Novofil suture. A Hemovac drain was placed in the subgaleal space, and a head dressing applied.

☆ Case Report

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http://dx.doi.org/10.1016/j.inat.2016.11.011

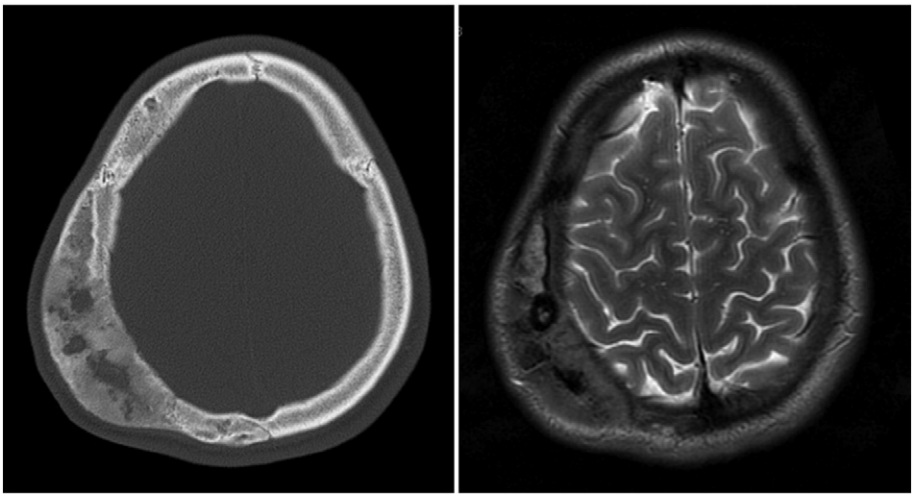

Fig. 1. A right parietal bone lesion is seen on axial CT scan (left panel) and T2-weighted axial MRI (right panel), without underlying brain compression.

Histopathologic evaluation revealed aneurysmal bone cyst with traces of fibrous dysplasia. Post-operative imaging showed resolution of the lesion (Fig. 3). Cosmesis was deemed excellent by both the patient and family members.

3. Discussion

Calvarial aneurysmal bone cysts may present as a cosmetic defect or with neurologic symptoms. Primary aneurysmal bone cysts arise from genetic rearrangements in the oncogene *TRE17*/*USP6* [3], and may result from subsequent upregulation of matrix metalloproteinases via NF-κB [4,5]. The pathogenesis of secondary aneurysmal bone cysts is not well established, though an insult resulting in increased venous pressure has long been posited as a potential mechanism [6]. A minority of cases of fibrous dysplasia are due to McCune-Albright syndrome, caused by activating mutations in *GNAS*, polyostotic fibrous dysplasia co-occurs with café-au-lait spots and endocrinopathies including precocious puberty [7–11].

To ensure holistic treatment, a variety of medical modalities should be considered in the management of aneurysmal bone cysts. Although for cranial lesions the mainstay of treatment is surgical excision, pre-operative or stand-alone embolization has been used with success as well [12]. Radiotherapy has been used in the management of both primary and recurrent lesions, including a recurrent temporal lesion [13,14]. In aneurysmal bone cysts of the spine and pelvis deemed unresectable, bisphosphonates and denosumab have been used with some efficacy [15,16]. Finally, sclerotherapy has also been an effective management strategy [17].

It is important to note that large aneurysmal bone cysts treated with cranioplasty may require complex skin excision, as was the case with our patient who benefitted greatly from in situ cranioplasty with methyl methacrylate and molded titanium mesh. Major objectives of cranioplasty include wound healing, protection of cranial vault structures, as well as achieving cosmetic satisfaction. In situ cranioplasty for aneurysmal bone cysts avoids advanced preparation of custom prostheses often required in other cranioplasty techniques. Furthermore, it curtails the technical problems of bone graft cranioplasties such as harvest site disfigurement when autologous bone is taken from a donor site. As compared to the split calvarial approach, in situ cranioplasty avoids added morbidity to harvest autologous bone from donor skull. Lastly, it has been documented that in situ cranioplasty costs are reasonable and approximately 2.5 times less expensive than osteo-inductive materials such as hydroxyapatite [18]. As shown by our case report, use of this in situ cranioplasty technique can also result in superb cosmesis.

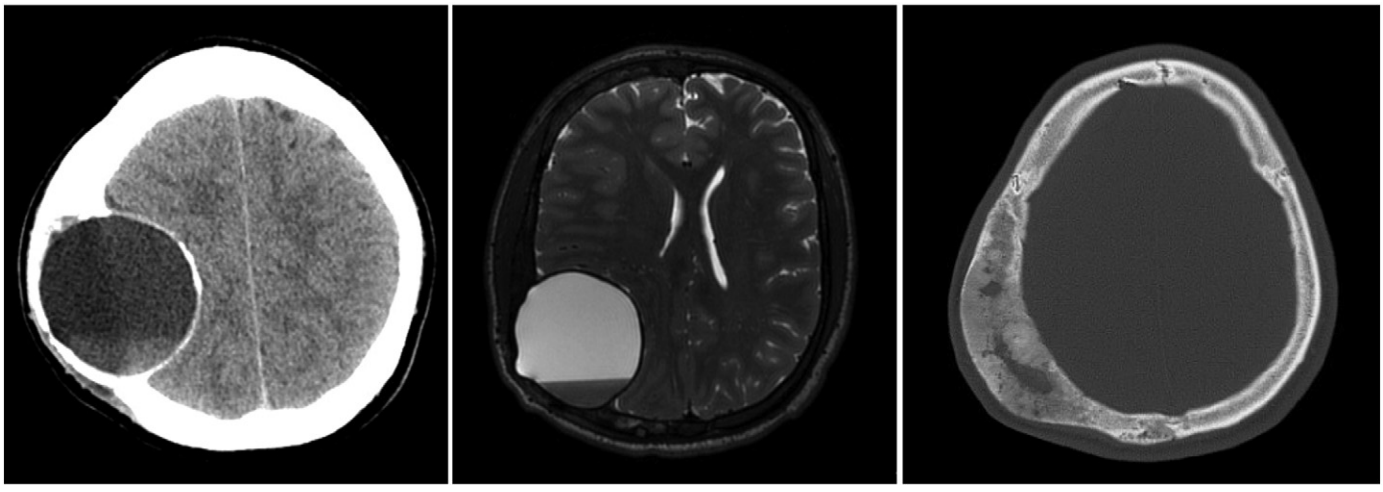

Fig. 2. A large aneurysmal bone cyst is demonstrated on axial non-contrast CT scan (left panel) and T2-weighted axial MRI (middle panel) with compression of the parietal lobe and significant midline shift. CT bone window shows aneurysmal bone cyst is located within area of fibrous dysplasia (right panel).

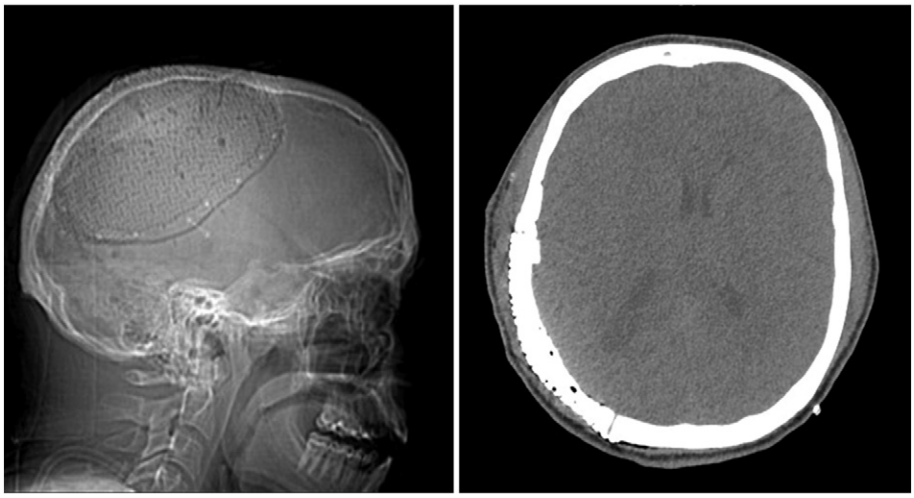

Fig. 3. The in situ cranioplasty is demonstrated on lateral x-ray (left panel) and axial non-contrast CT scan (right panel).

Sources of support

None relevant for disclosure.

Conflicts of interest/disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

References

[1] W.M. Mendenhall, et al., Aneurysmal bone cyst, Am. J. Clin. Oncol. 29 (3) (2006) 311–315.
[2] D. Lau, M.W. McDermott, A method for combining thin and thick malleable titanium mesh in the repair of cranial defects, Cureus 7 (5) (2015) e267.
[3] A.M. Oliveira, et al., USP6 and CDH11 oncogenes identify the neoplastic cell in primary aneurysmal bone cysts and are absent in so-called secondary aneurysmal bone cysts, Am. J. Pathol. 165 (5) (2004) 1773–1780.
[4] L.M. Pringle, et al., Atypical mechanism of NF-kappaB activation by TRE17/ubiquitin-specific protease 6 (USP6) oncogene and its requirement in tumorigenesis, Oncogene 31 (30) (2012) 3525–3535.
[5] Y. Ye, et al., TRE17/USP6 oncogene translocated in aneurysmal bone cyst induces matrix metalloproteinase production via activation of NF-kappaB, Oncogene 29 (25) (2010) 3619–3629.
[6] L. Lichtenstein, Aneurysmal bone cyst. A pathological entity commonly mistaken for giant-cell tumor and occasionally for hemangioma and osteogenic sarcoma, Cancer 3 (2) (1950) 279–289.
[7] C.E. Dumitrescu, M.T. Collins, McCune-Albright syndrome, Orphanet. J. Rare Dis. 3 (2008) 12.
[8] D. McCune, Osteitis fibrosa cystica; the case of a nine year old girl who also exhibits precocious puberty, multiple pigmentation of the skin and hyperthyroidism, Am. J. Dis. Child 52 (743) (1936) 4.
[9] F. Albright, et al., Syndrome characterized by osteitis fibrosa disseminata, areas of pigmentation and endocrine dysfunction, with precocious puberty in females: report of five cases, N. Engl. J. Med. 216 (17) (1937) 727–746.
[10] L.S. Weinstein, et al., Activating mutations of the stimulatory G protein in the McCune-Albright syndrome, N. Engl. J. Med. 325 (24) (1991) 1688–1695.
[11] A.M. Boyce, M.T. Collins, Fibrous Dysplasia/McCune-Albright Syndrome, in: R.A. Pagon, et al., (Eds.), GeneReviews(R), University of Washington, Seattle: Seattle (WA), 1993.
[12] B.Y. Sheikh, Cranial aneurysmal bone cyst "with special emphasis on endovascular management", Acta Neurochir. 141 (6) (1999) 601–610 (discussion 610-1).
[13] S.J. Feigenberg, et al., Megavoltage radiotherapy for aneurysmal bone cysts, Int. J. Radiat. Oncol. Biol. Phys. 49 (5) (2001) 1243–1247.
[14] P. Kumar, et al., Role of radiotherapy in a recurrent aneurysmal bone cyst of the temporal bone: case report, Neurosurgery 58 (3) (2006) E584 (discussion E584).
[15] F. Cornelis, et al., Bisphosphonate therapy for unresectable symptomatic benign bone tumors: a long-term prospective study of tolerance and efficacy, Bone 58 (2014) 11–16.
[16] T. Lange, et al., Denosumab: a potential new and innovative treatment option for aneurysmal bone cysts, Eur. Spine J. 22 (6) (2013) 1417–1422.
[17] J. Dubois, et al., Sclerotherapy in aneurysmal bone cysts in children: a review of 17 cases, Pediatr. Radiol. 33 (6) (2003) 365–372.
[18] J. Hohne, et al., Feasibility of the custom-made titanium cranioplasty CRANIOTOP, Surg. Neurol. Int. 4 (2013) 88.