



Pediatric aggressive giant cell granuloma of nasal cavity



Sung Tae Seo^{a,c}, Ki Ryun Kwon^{a,c}, Ki-Sang Rha^{a,c}, Seon-Hwan Kim^b, Yong Min Kim^{a,c,*}

^a Department of Otorhinolaryngology-Head and Neck Surgery, Chungnam National University School of Medicine, Daejeon, South Korea

^b Department of Neurosurgery, Chungnam National University School of Medicine, Daejeon, South Korea

^c Research Institute for Medical Science, Chungnam National University School of Medicine, Daejeon, South Korea

ARTICLE INFO

Article history:

Received 22 June 2015

Received in revised form 4 September 2015

Accepted 5 September 2015

Available online 18 September 2015

Keywords:

Aggressive giant cell granuloma
Endoscopic craniofacial resection

ABSTRACT

INTRODUCTION: Giant cell granuloma (GCG) is a non-neoplastic osseous proliferative lesion of unknown etiology. Although a benign disease process, GCG can be locally destructive. It is extremely rare to have a pediatric case of GCG occurring in the nasal cavity with intracranial invasion.

PRESENTATION OF CASE: We report a case of an aggressive and recurrent giant cell granuloma with intracranial invasion in a 10 years old female patient which was completely excised with endoscopic craniofacial resection.

DISCUSSION: A literature review on pathogenesis, diagnosis and management is also performed.

CONCLUSION: The most common treatment for giant cell granuloma is surgery, ranging from simple curettage to resection. However, it must be completely excised in cases of aggressive and extensive lesion because of the high recurrence rate after incomplete removal.

© 2015 The Authors. Published by Elsevier Ltd. on behalf of Surgical Associates Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Giant cell granuloma (GCG) is a rare benign proliferative osteolytic lesion that commonly occurs in the maxilla and mandible [1]. GCG usually occur after the second decade of life and has been known for female predominance in spite of existing controversy [2]. GCG was first described by Jaffe in 1953 as a giant cell reparative granuloma [3]. Since GCGs were revealed to be locally aggressive in nature and they can occur without history of trauma, the term “reparative” has not been used recently [4]. There have been few cases of GCG occurring in the nasal cavity with intracranial invasion [5]. We report a 10-year-old girl who had an aggressive GCG occurring in the nasal cavity with intracranial invasion. Her GCG was initially treated with endoscopic excision, but recurred in 6 weeks. The recurrent lesion was eventually completely excised with endoscopic craniofacial resection.

2. Presentation of case

A 10-year-old girl was referred to our institution with one-month history of epistaxis, nasal obstruction, proptosis, and epiphora. The patient denied any history of pain, headache, neurologic symptoms, diplopia or visual disturbance. There was no history of trauma or previous sinonasal operation. On physical

examination, a huge, partially reddish mass was observed through the right nostril and the outer surface of the mass was smooth. The mass completely occupied the right nasal cavity and pushed the nasal septum towards the left side, resulting in the left nasal cavity being completely obstructed. The mass has slightly displaced the right eyeball towards the lateral side and mild exophthalmos was observed. There were no palpable masses or lymphadenopathies in the neck. Routine hematological and biochemical tests including serum calcium, alkaline phosphatase, phosphorous, renal function, and parathyroid hormone were within normal limits.

Paranasal sinus computerized tomography (PNS CT) scanning and magnetic resonance imaging (PNS MRI) revealed a large multiseptated cystic expansile lesion with peripheral enhancement in the right sinonasal cavity. The mass has extensions into the orbit, ethmoid sinus and anterior cranial fossa (Fig. 1A). With these imaging studies, the first impression of the radiologist was an infected mucocele.

Endoscopy with biopsy revealed the outer surface of the mass was smooth and rubbery with the inside being cystic in nature, filled with loose connective tissue and hematoma-like material. Histopathological result of the preoperative biopsy was uncertain and was described as ‘ulcerative and necrotic tissue’ or ‘granulation tissue proliferation’. The pathologist reported that there is the possibility of pyogenic granuloma. From these results, the authors concluded that the lesion was not a neoplastic disease but an inflammatory lesion combined with granulomatous lesion and mucocele. The patient underwent endonasal endoscopic resection without external approach. The mass originated from the lateral wall of the nasal cavity and has eroded the lamina papyracea,

* Corresponding author at: Department of Otolaryngology-Head and Neck Surgery, Chungnam National University Hospital, 282 Munhwa-ro, Jung-gu, Daejeon 301-721, South Korea. Fax: +82 42 253 4059.

E-mail address: entkym@cnu.ac.kr (Y.M. Kim).

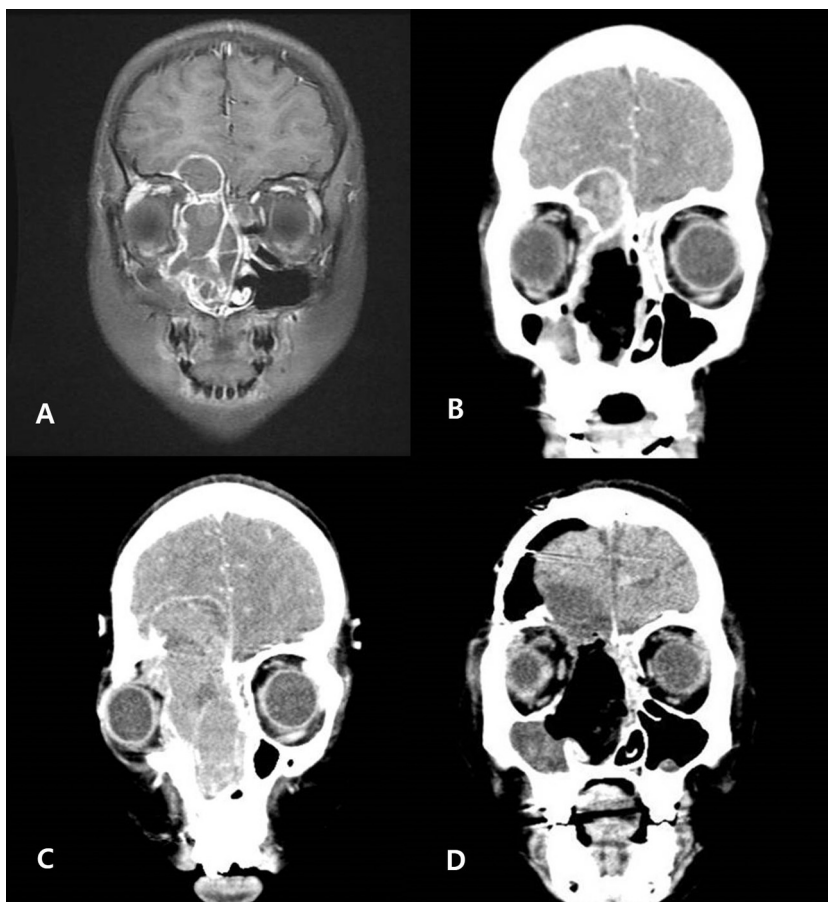


Fig. 1. Images of clinical course. Preoperative T1-weighted MRI (A) postoperative Coronal view of PNS CT after incomplete resection (B) on postoperative 6 weeks showing recurrent GCG (C) on postoperative 7 days after completely removal (D).

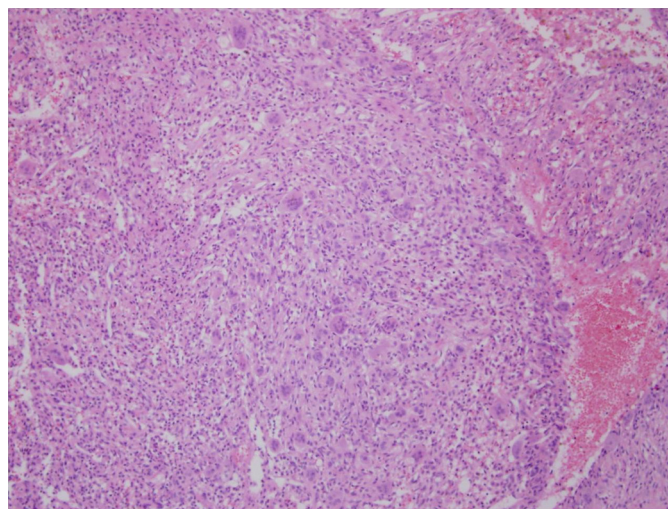


Fig. 2. Histopathologic findings of GCG. (Hematoxylin & eosin, original magnification $\times 100$) Numerous multinucleated giant cells in the dense fibrous connective tissue.

medial wall of maxilla, and anterior skull base. The mass was excised piecemeal using forceps and microdebrider. Strong adhesion in the frontal sinus and intracranial portions were noted. Complete removal of the intracranial lesion and frontal sinus lesion was not achieved because of massive bleeding and poor operation fields. Promptly, an emergent consultation with the neurosurgeon was conducted to completely remove. His opinion was that further resection may not be necessary because of the possibility of

spontaneous involution. Eventually, the operation was concluded even with remnant lesions (Fig. 1B). Histopathologic diagnosis was giant cell granuloma and sections showed fibroblastic proliferation with rich osteoclast-like polynuclear giant cells interspersed with spindle-shaped stroma (Fig. 2).

One month after the operation, the patient did not complain of any headache, nasal obstruction and exophthalmos and her postoperative course seemed to be uneventful. She visited the

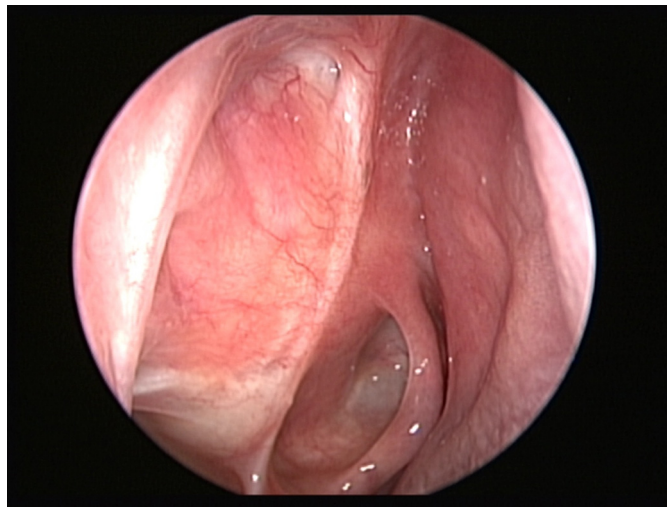


Fig. 3. Photograph of nasoendoscopic finding of the right nasal cavity in the office taken 2 years after surgery.

outpatient clinic 6 weeks after the first operation complaining of nasal obstruction, headache, and proptosis on the right side. Physical examination and PNS CT & MRI revealed the recurrent GCG has completely obliterated the right nasal cavity and appeared more aggressive in nature compared to the previous preoperative status (Fig. 1C). The patient underwent surgical resection via combined endonasal endoscopic resection with craniotomy by bilateral coronal incision. The lesion was visibly invasive of the surrounding bone and a wide resection of all affected bone was conducted. There was no evidence of dura or periorbital invasion and the mass was dissected completely using an elevator without difficulty. Resulting bony defects of the skull base were reconstructed using a pericranial flap. The postoperative course was uneventful, and she recovered quickly in the intensive care unit (Fig. 1D). After a 2 year of follow-up, the patient was free of recurrence (Fig. 3).

3. Discussion

GCG can be clinically classified as non-aggressive or aggressive type [6]. In general, aggressive GCGs affect children at an earlier age, were larger at the time of diagnosis, and recurred more frequently than non-aggressive type. Especially, GCG affecting the cranial bone is extremely rare and has a locally aggressive and destructive behavior [1]. There has been few case of GCG occurring in the nasal cavity with intracranial invasion [5,7]. To the best of our knowledge, our case may be first in a pediatric patient.

Clinical symptoms of sinonasal GCG are nonspecific and dependent on the involved site and the extent of local destruction [7,8]. There has been no specific radiological description for GCG due to its rarity of presentation [1]. With CT scanning, the lesion sometimes shows enhanced cystic formations, small calcifications, and bony remodeling. The lesion can be confused with infected mucocele or multiseptated cyst. With MRI, GCG demonstrates a low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images.

Histologically, the major components of GCGs are multinucleated giant cells related to focal areas of hemorrhage, abundant spindle-shaped fibroblastic cells, low mitotic activity, cyst formation and reactive bone formation [7,9]. The aggressive type had a higher relative size index of giant cells, and recurrent GCG had a higher relative size index of giant cells and fractional surface area occupied by giant cells. However, they described histologic differences between the two groups (non-aggressive vs. aggressive) were unclear [6]. It is also difficult to decide plan of invasive operation

range depending on only uncertain histopathological result of the preoperative biopsy and radiological finding in pediatric case. In addition, it may be difficult to differentiate it from the other giant cell lesions with similar histological appearance and clinical course. Aneurismal bone cyst, giant cell tumor (osteoclastoma) and Brown tumor characterized by hyperparathyroidism are the lesions that must be included in the differential diagnosis.

The traditional treatment of choice for GCG has been surgical excision. The extent of tissue removal ranges from simple curettage to en bloc resection. Recurrence rates following surgery have been reported at 11–72% and were higher in patients with aggressive lesions [2,6].

Non-surgical treatments include supplementary modalities such as corticosteroid intralesional injection, calcitonin, interferon alpha, and radiation. These procedures should be considered as secondary tools for lesions that cannot be completely resected because of higher possibility of additional surgery [9].

We learned a significant lesson about diagnosis & treatment through very interesting experience in the aggressive change after the first surgery. We have two main questions under treatment for GCG of this case. First, how far in advance it can be judged an aggressive type of GCG? Second, what extent it will determine the surgical range in pediatric case of GCG? In conclusion, only complete resection is the appropriate treatment for GCG because, accurate diagnosis is difficult and recurrent GCG develops a more aggressive behavior in a short duration.

4. Conclusion

We report a pediatric case of aggressive GCG that originated from the nasal cavity with intracranial invasion. The extent of surgical treatment includes a wide range of options. However, we have to consider complete resection of GCG as a means of the high priority, despite younger age.

Conflict of interest

The authors declare that they have no conflict of interests.

Funding

None.

Ethical approval

No ethical approval was required.

Consent

Written informed consent was obtained from the patients for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Authors contribution

Yong Min Kim and Seon-Hwan Kim operated the patients.

Sung Tae Seo and Ki Ryun Kwon prepared the first draft of the manuscript.

Ki-Sang Rha and Yong Min Kim revised the manuscript for important intellectual content and technical details.

Sung Tae Seo and Yong Min Kim provided useful suggestions on content and editing issues. All authors have read and approved the final manuscript.

References

- [1] A. Conley, B.H. Cho, G.W. Tye, et al., Giant cell reparative granuloma of the pediatric cranium: case report and review of the literature, *Childs Nerv. Syst.* 30 (2014) 521–526.
- [2] J. de Lange, H.P. van den Akker, H. Klip, Incidence and disease-free survival after surgical therapy of central giant cell granulomas of the jaw in The Netherlands: 1990–1995, *Head Neck* 26 (2004) 792–795.
- [3] H.L. Jaffe, Giant-cell reparative granuloma traumatic bone cyst, and fibrous (fibro-osteous) dysplasia of the jawbones, *Oral Surg. Oral Med. Oral Pathol.* 6 (1953) 159–175.
- [4] J.E. O'Connell, G.J. Kearns, Aggressive giant cell granuloma of the jaws treated with interferon alpha: a report of two cases, *Ir. J. Med. Sci.* 182 (2013) 163–170.
- [5] H. Ishinaga, K. Otsu, G. Mouri, K. Takeuchi, Aggressive giant cell reparative granuloma of the nasal cavity, *Case Rep. Otolaryngol.* 2013 (2013) 690194.
- [6] R. Chuong, L.B. Kaban, H. Kozakewich, A. Perez-Atayde, Central giant cell lesions of the jaws: a clinicopathologic study, *J. Oral Maxillofac. Surg.* 44 (1986) 708–713.
- [7] H.N. Arda, M.F. Karakus, M. Ozcan, N. Arda, T. Gun, Giant cell reparative granuloma originating from the ethmoid sinus, *Int. J. Pediatr. Otorhinolaryngol.* 67 (2003) 83–87.
- [8] C.B. Koay, H.B. Whittet, R.M. Ryan, C.E. Lewis, Giant cell reparative granuloma of the concha bullosa, *J. Laryngol. Otol.* 109 (1995) 555–558.
- [9] S. Hirschl, A. Katz, Giant cell reparative granuloma outside the jaw bone: diagnostic criteria and review of the literature with the first case described in the temporal bone, *Hum. Pathol.* 5 (1974) 171–181.

Open Access

This article is published Open Access at sciedirect.com. It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.