CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 75 (2020) 437–440

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Pediatric allergic fungal rhinosinusitis with extensive intracranial extension – Case report and literature review

Haifa Lafi Alenzi [a,*], Ali Al Momen [b], Fadel Molani [c]

[a] Northern Border University, Saudi Arabia
[b] ENT, Rhinology & Skull Base Surgery Consultant at King Fahad Specialist Hospital, Dammam, Saudi Arabia
[c] Department of Medical Imaging at King Fahad Specialist Hospital, Dammam, Saudi Arabia

ARTICLE INFO

Article history:
Received 21 June 2020
Received in revised form 26 August 2020
Accepted 5 September 2020
Available online 12 September 2020

Keywords:
Allergic fungal
Allergy
Sinusitis
Fungal infection
Intracranial extradural extension

ABSTRACT

INTRODUCTION: Over the last two decades, allergic fungal sinusitis (AFRS) has become increasingly common. It's defined as a noninvasive, benign inflammatory fungal disease of the sinuses which develops in young adults and adolescents. Patients often complain of symptoms like nasal obstruction, congestion, purulent or clear rhinorrhea, anosmia, and headache. The cases are also presenting clinically with symptoms like epiphora and eye discharge as a result of nasolacrimal gland obstruction. In this article, we will review a unique case of AFRS, in an adolescent male. The case was diagnosed with intracranial extradural extension.

CASE REPORT: A 15 years old male with AFRS was diagnosed and managed. The case was diagnosed to have allergic fungal sinusitis based on Bent and Khun diagnostic criteria, presented with intracranial extradural extension.

DISCUSSION: In our case, there were no irreversible complications except a recurrent polyp. The case was mainly complaining of long-standing nasal discharge and on-off headache with no orbital complaint and no other neurological signs. This shows a presentation of the fungal sinusitis and the need for aggressive intervention for AFRS both medically and surgically for pediatric patients as well.

CONCLUSION: To conclude, despite AFRS being categorized as a benign, non-invasive disease, its presentation can range from simple nasal obstruction to signs and symptoms of intraorbital and/or intracranial complications; with pediatric cases being very aggressive. Careful clinical evaluation, detailed histopathological examination, navigation assisted endoscopic sinus surgery followed by steroid treatment, and a lifelong follow up to manage the recurrence.

1. Introduction

Over the last two decades, allergic fungal sinusitis (AFRS) has become increasingly common. It's defined as a noninvasive, benign inflammatory fungal disease of the sinuses which develops in young adults and adolescents. It's more common in temperate regions with high humidity. The primary causative organism was thought to be Aspergillus [1]. Generally; fungal sinusitis has been classified into two main groups: invasive form and non-invasive form, these Invasive forms of the fungal disease are usually encountered in immuno-compromised subjects. However, some reports have described these forms occurring in immuno-competent individuals [2,3]. Several organizations have classified rhino-sinusitis into four different groups: acute rhino-sinusitis, chronic rhino-sinusitis with nasal polyps (CSRwNP), chronic rhinosinusitis without nasal polyps (CSRsNP), and allergic fungal rhinosinusitis (AFRS). Thus, categorizing AFRS as a distinct form of chronic rhinosinusitis (CRS) [4,5]. Patients often complain of symptoms like nasal obstruction, congestion, purulent or clear rhinorrhea, anosmia, and headache [6,7]. The cases are also presenting clinically with symptoms like epiphora and eye discharge as a result of nasolacrimal gland obstruction [8]. On examination, allergic mucin, that contains sparse numbers of fungal hyphae are usually seen. In this article, we will review a unique case of AFRS, in an adolescent male. The case was diagnosed with intracranial extradural extension and managed in King Fahad Specialist Hospital (KFSHD), a tertiary care hospital in Al-Dammam, Saudi Arabia. The work has been reported in line with the SCARE criteria [33].

2. Case report

AFRS with intracranial extradural extension: 15 years old male presented with nasal discharge and obstruction, occasional headache with a positive history of allergic symptoms. Examina-

* Corresponding author.
E-mail address: haifalafi@hotmail.com (H.L. Alenzi).

https://doi.org/10.1016/j.ijscr.2020.09.049

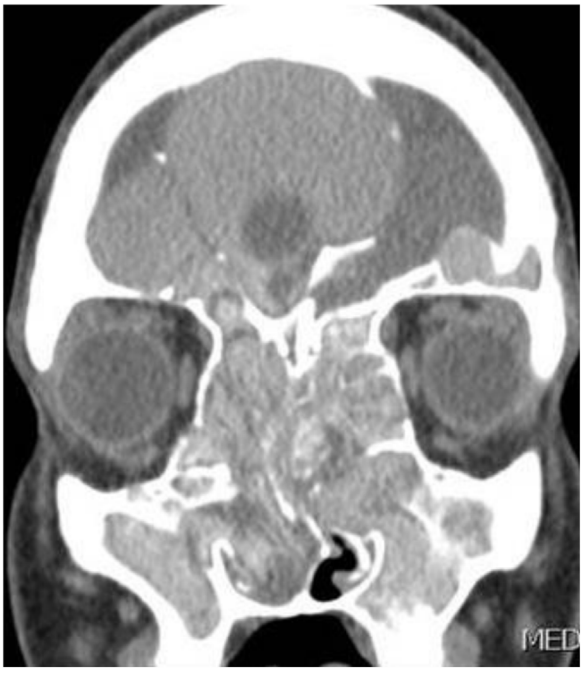

Fig. 1. Coronal CT sinuses show extensive disease with a marked expansion of the sinuses, more pronounced in the right frontal sinus. This results in the dehiscence of the posterior wall and intracranial extension.

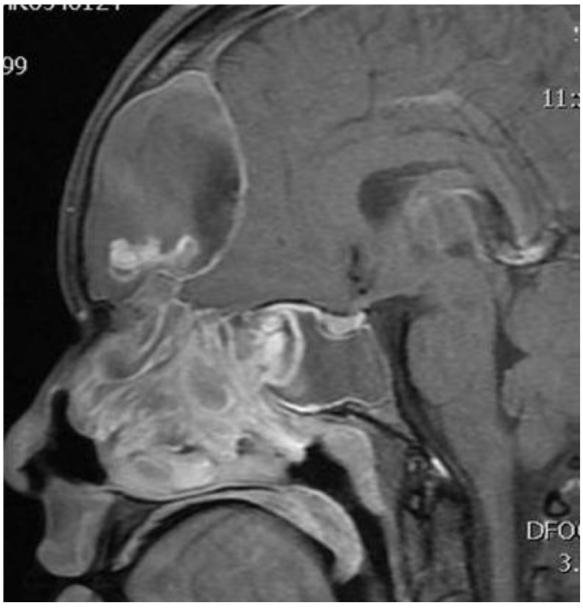

Fig. 2. Sagittal T1 postcontrast MRI shows the right frontal sinus expansion and intracranial extension pushing the frontal lobe posteriorly limited by an intact dura. The sphenoid sinus expanded with an upward displacement of the pituitary fossa.

tion of the nose showed bilateral grade 4 nasal polyps and thick fungal mucin filling both nasal cavities.

CT sinuses showed extensive disease with a marked expansion of the sinuses with dehiscence of the posterior wall and intracranial extension (Fig. 1). MRI showed right frontal sinus expansion and intracranial extension limited by an intact dura (Fig. 2). MR imaging is indicated in all extensive cases of fungal sinusitis with suspected intracranial and or intraorbital involvement, with the differential diagnosis of allergic fungal sinusitis, invasive fungal sinusitis and extensive mucopyoceles.

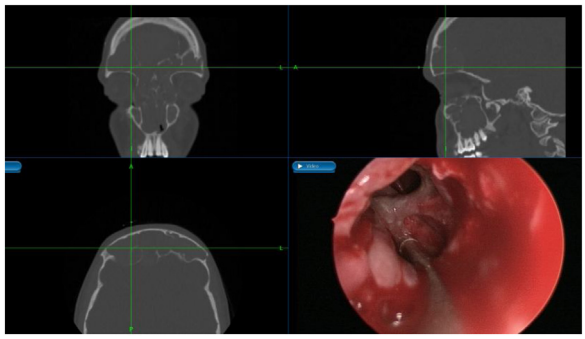

Fig. 3. Intra-operative Navigation assisted endoscopic wide frontal sinusotomy And fungal mucin removal.

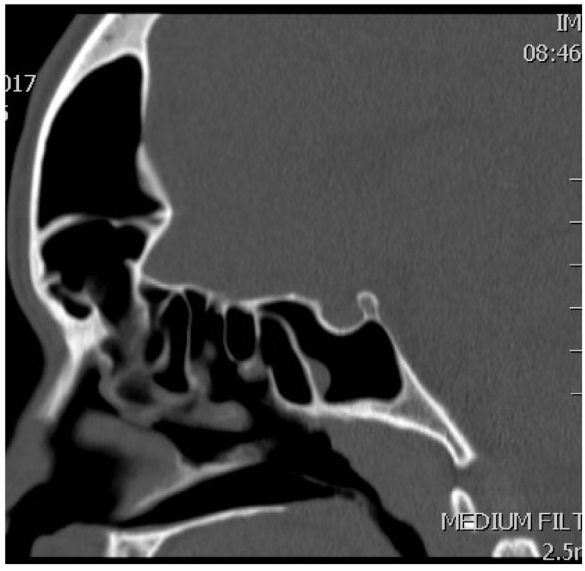

Fig. 4. Post-operative follow-up CT scan with normally aerated sinuses.

The patient underwent navigation assisted functional endoscopic sinus surgery (FESS) (Fig. 3).

The polyps and fungal mucin were entirely removed from the nasal cavities, bilateral wide maxillary sinus antostomies, and thick mucin was aspirated. The anterior and posterior ethmoid sinuses were full of polyps, mucin, and fungal mud which was aspirated. The frontal recesses and sinuses were full of polyps, mucin, and fungal mud causing erosion of frontal floors and displacing the orbits, all were cleaned, drained with the help of navigation. The pulsating intact dura was observed and cleaned from the thick mucin, and fungal mud with no CSF leak observed.

The patient was managed by a short course of systemic oral steroid, topical steroid sprays, and continuous nasal saline irrigations. The patient continued to have regular endoscopic follow up with no signs of recurrence after four years, as demonstrated by the clear postoperative CT scan sinuses (Fig. 4).

3. Discussion

There are many studies in the literature regarding AFRS in adults. However even though AFRS presents as well but to a lesser extent in the pediatric age group, there only a few data in the literature regarding its nature, clinical course, and recurrence in children [11]. Children and adolescents with AFS usually appear to differ from adults in some aspects of their clinical presentation. The adolescent boy, in our evaluation, had a clear facial dysmorphism. The obvious facial alteration, usually in the form of proptosis, has specif-

ically been addressed in case reports [9,10] and small series [12,13] and has been mentioned in larger reviews [14,15]. Generally; nasal polyposis is considered a rare presentation in children and adolescents unless associated with an underlying pathology like cystic fibrosis or other ciliary dysmotility syndromes [16,17]. Our case was a male with 15 years old. A review of children with AFS at the University of Texas Southwestern revealed a distinct male predominance, with a 2.1:1 M/F ratio and the average age at diagnosis of 13 years [18]. A Male/Female ratio of 1.5:1 with a mean age at diagnosis of 13.6 years was also reported in another series of children with AFRS [19]. Another pediatric series consisted of 10 patients and had a male-to-female ratio of 1.5 to 1, and a mean age of 13.6 years [23] Interestingly, when two series of patients who come from a single institution were reviewed over time, an early study with a male-to-female ratio of 1.5 had an age range of 13–51 years (average age, 27.5 y), one, and a later review had a female-to-male ratio of 1.4 with an age range of 13–69 years (no average age is given) [24]. Children report a slow onset of nasal airway obstruction and production of large, dark-coloured nasal debris [20,21]. In another prospective study of 200 cases divided into two groups. Sixty-eight cases in group 1 (less than 15 years) and 132 cases in group 2 (more than 15 years). The study concluded the following: nasal obstruction was the most common presentation in both groups. The children had a higher incidence of having unilateral disease compared with adults, and finally, group 1 had a higher incidence of facial deformities, proptosis, intraorbital/intracranial extension along with a higher rate of recurrence. This also suggested a more aggressive nature of AFRS in children when compared to adults as the previous study [22]. The largest study that addressed the issue of laterality of fungal disease was an interinstitutional review of 45 patients having an average age of 25 years (age range, 8–68 y). The authors concluded that bilateral disease was more common than unilateral disease but reported almost equal rates (51% [bilateral] vs 49% [unilateral]). However, they did point out that, overall, the disease asymmetrically involved the sinuses in 78% of the cases [25]. Another study by John E. McClay et al. [26] found children to have unilateral disease on presentation much more often than adults (70% vs 37%). Overall, the fungi recovered from the sinuses in children and adults were similar. Both adults and children had mostly Bipolaris species, followed by Curvularia species. Most large reviews have indicated that Bipolaris and Curvilaria species are the most common fungi recovered [27,28]. Several other studies in the literature have reported alteration in the facial skeleton in 42% of pediatric patients which includes Proptosis, telecanthus, or malar flattening, compared to 10% for adults. This could be explained by the more pliable bony structure of children [29,30]. Also, a case of AFRS in the sphenoid sinus of an immuno-competent pediatric patient can present with visual loss. In this case, the visual loss was irreversible even after aggressive endoscopic debridement of sinuses and the use of corticosteroids. This underscores the potentially serious nature of sphenoid sinusitis and highlights its possibility to induce visual loss [31]. Ikram et al. reported a retrospective study of 26 patients, all of whom had AFRS with an intracranial spread. Twenty-four underwent minimally invasive endoscopic sinus surgery, only two patients however required additional external procedures as pathology was inaccessible endoscopically. None required a craniotomy. Postoperatively, all patients received oral steroid therapy for one month (0.5 mg/kg) and remained disease-free for a period of up to 5 years [32]. In our case, there were no irreversible complications except a recurrent polyp. The case was mainly complaining of long-standing nasal discharge and on-off headache with no orbital complaint and no other neurological signs. Both ophthalmology and neurosurgery teams were consulted for any intraoperative interference. Intraoperatively, the neurosurgery team was available in case needed small bone erosion was found in the suprasellar, clivus parasellar region which was repaired with Hadad flap successfully. Therefore, there was no need for neurosurgery team intervention. After endoscopic sinus surgery with debridement, He improved, followed by medical therapy. It should also be noted that some rare, extensive cases might require an intracranial approach for clearing the intracranial extension, also for possible defect repair that cannot be managed with endoscopic approach alone [19]. This shows a presentation of the fungal sinusitis and the need for aggressive intervention for AFRS both medically and surgically for pediatric patients as well (Table 1).

Table 1
Classification of fungal sinusitis.

Non invasive
Asymptomatic fungus isolation
Saprophytic fungal infection (crust)
Fungal ball (mycetoma)
Allergic fungal sinusitis
Coexistence of invasive and noninvasive
Allergic fungal sinusitis with coexistent granuloma
Invasive
Granulomatous invasive fungal sinusitis
Chronic invasive fungal sinusitis
Acute fulminant fungal sinusitis

4. Conclusion

AFRS can present clinically in different ways. Despite being categorized as a benign, non-invasive disease, its presentation can range from simple nasal obstruction to signs and symptoms of intra-orbital and/or intracranial complications; with pediatric cases being very aggressive, Careful clinical evaluation, detailed histopathological examination to rule out mixed types and malignancies, and a lifelong follow up should be done to manage the recurrence.

Declaration of Competing Interest

The authors report no declarations of interest.

Funding

There is no financial support of sponsorship involved in this study.

Ethical approval

Ethical approval by IRB for case reports is not needed in this institution.

Consent

Written consent was obtained from parent's patients for the publication of this case report accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Haifa Al Enzi: writing the original manuscript draft, review and editing of the manuscript.

Fadel Molani: data collection and analysis.

Ali Al Momen: study concept, data analysis, and final approval of manuscript.

Registration of research studies

Not applicable.

Guarantor

Dr. Ali Al Momen.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Data availability

The data used to support the findings of this study are included in the article. Also, they are available from the corresponding author upon request.

References

[1] B.F. Marple, Allergic fungal rhinosinusitis: current theories and management strategies, Laryngoscope 111 (6) (2009) 1006–1019.
[2] S.R. Sridhara, G. Paragache, N.K. Panda, A. Chakrabarti, Mucormycosis in immunocompetent individuals: an increasing trend, J. Otolaryngol. 34 (6) (2005) 402–406.
[3] J.B. Taxy, S. El-Zayaty, A. Langerman, Acute fungal sinusitis: natural history and the role of frozen section, Am. J. Clin. Pathol. 132 (1) (2009) 86–93.
[4] D.L. Hamilos, Chronic sinusitis, J. Allergy Clin. Immunol. 106 (2) (2000) 213–227.
[5] C. Chang, M.E. Gershwin, G.R. Thompson, Fungal disease of the nose and sinuses: an updated overview, Curr. Allergy Asthma Rep. 13 (2) (2013) 152–161.
[6] A.L. Katzenstein, S.R. Sale, P.A. Greenberger, Pathologic findings in allergic aspergillus sinusitis. A newly recognized form of sinusitis, Am. J. Surg. Pathol. 7 (5) (1983) 439–443.
[7] J.E. McClay, B. Marple, L. Kapadia, M.J. Biavati, B. Nussenbaum, et al., Clinical presentation of allergic fungal sinusitis in children, Laryngoscope 112 (3) (2002) 565–569.
[8] C. Kim, A. Kacker, R.-I. Chee, G.J. Lelli, Allergic fungal sinusitis causing nasolacrimal duct obstruction, Orbit 32 (2) (2013) 143–145.
[9] T.F. Pingree, G.R. Holt, R.A. Otto, M.G. Rinaldi, Bipolaris-caused fungal sinusitis, Otolaryngol. Head Neck Surg. 106 (1992) 302–305.
[10] C.N. Young, J.G. Swart, D. Ackerman, D. Davidge-Pitts, Nasal obstruction and bone erosion caused by Drechslera Hawaiiensis, J. Laryngol. Otol. 92 (1978) 137–143.
[11] S.K. Patro, R.K. Verma, N.K. Panda, A. Chakrabarti, Understanding paediatric allergic fungal sinusitis: is it more aggressive? Int. J. Pediatr. Otorhinolaryngol. 79 (11) (2015) 1876–1880.
[12] W.J. Chang, D.T. Tse, K.L. Bressler, R.R. Casiano, R.H. Rosa, T.E. Johnson, Diagnosis and management of allergic fungal sinusitis with orbital involvement, Ophthal. Plast. Reconstr. Surg. 16 (2000) 72–74.
[13] C. Torres, J.Y. Ro, A.K. El-Naggar, S.J. Sim, R.S. Weber, A.G. Ayala, Allergic fungal sinusitis: a clinicopathologic study of 16 cases, Hum. Pathol. 27 (1996) 793–799.
[14] R.D. DeShazo, R.E. Swain, Diagnostic criteria for allergic fungal sinusitis, J. Allergy Clin. Immunol. 96 (1995) 24–35.
[15] M.W. Yung, J. Gould, G.J.G. Upton, Nasal polyposis in children with cystic fibrosis: a long-term follow-up study, Ann. Otol. Rhinol. Laryngol. 111 (12) (2002) 1081–1086.
[16] Lands L.C. Do BAJ, C. Saint-Martin, M.A. Mascarella, J.J. Manoukian, et al., Effect of the F508del genotype on outcomes of endoscopic sinus surgery in children with cystic fibrosis, Int. J. Pediatr. Otorhinolaryngol. 78 (7) (2014) 1133–1137.
[17] S.K. Patro, R.K. Verma, N.K. Panda, A. Chakrabarti, Understanding paediatric allergic fungal sinusitis: is it more aggressive? Int. J. Pediatr. Otorhinolaryngol. 79 (11) (2015) 1876–1880.
[18] B.D. Thorp, K.A. McKinney, A.S. Rose, C.S. Ebert, Allergic fungal sinusitis in children, Otolaryngol. Clin. North Am. 45 (3) (2012) 631–642.
[19] Noorah A.L. Mulhem, et al., The different clinical presentations of allergic fungal rhinosinusitis, Otolaryngol. Open Access J. 3 (2) (2018), http://dx.doi.org/10.23880/OOAJ-16000172, ISSN: 2476-2490.
[20] J.E. McClay, B. Marple, L. Kapadia, M.J. Biavati, B. Nussenbaum, et al., Clinical presentation of allergic fungal sinusitis in children, Laryngoscope 112 (3) (2002) 565–569.
[21] M.W. Yung, J. Gould, G.J.G. Upton, Nasal polyposis in children with cystic fibrosis: a long-term follow-up study, Ann. Otol. Rhinol. Laryngol. 111 (12) (2002) 1081–1086.
[22] A.K. Gupta, S. Bansal, A.K. Gupta, N. Mathur, Is fungal infestation of paranasal sinuses more aggressive in pediatric population? Int. J. Pediatr. Otorhinolaryngol. 70 (4) (2006) 603–608.
[23] S.B. Kupferberg, J.P. Bent, Allergic fungal sinusitis in the pediatric population, Arch. Otolaryngol. Head Neck Surg. 122 (1996) 1381–1389.
[24] S.B. Kupferberg, J.P. Bent, F.A. Kuhn, Prognosis for allergic sinusitis, Otolaryngol. Head Neck Surg. 117 (1997) 35–41.
[25] S.K. Mukherji, R.E. Figuero, L.E. Ginsberg, et al., Allergic fungal sinusitis: CT findings, Radiology 207 (1998) 417–422.
[26] John E. McClay, et al., Clinical presentation of allergic fungal sinusitis in children, Laryngoscope 112 (March) (2002).
[27] R.D. DeShazo, R.E. Swain, Diagnostic criteria for allergic fungal sinusitis, J. Allergy Clin. Immunol. 96 (1995) 24–35.
[28] M.S. Schubert, D.W. Goetz, Evaluation and treatment of allergic fungal sinusitis, I: demographics and diagnosis, J. Allergy Clin. Immunol. 102 (1998) 387–394.
[29] B.D. Thorp, K.A. McKinney, A.S. Rose, C.S. Ebert, Allergic fungal sinusitis in children, Otolaryngol. Clin. North Am. 45 (3) (2012) 631–642.
[30] J.M. Campbell, M. Graham, H.C. Gray, C. Bower, M.S. Blaiss, et al., Allergic fungal sinusitis in children, Ann. Allergy Asthma Immunol. 96 (2) (2006) 286–290.
[31] B.W. Herrmann, F.V. White, J.W. Forsen, Visual loss in a child due to allergic fungal sinusitis of the sphenoid, Otolaryngol. Head Neck Surg. 135 (2) (2006) 328–329.
[32] M. Ikram, S. Akhtar, S. Ghaffar, S.A. Enam, Management of allergic fungal sinusitis with intracranial spread, Eur. Arch. Otorhinolaryngol. 265 (2) (2008) 179–184.
[33] R.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, For the SCARE Group, The SCARE 2018 statement: updating consensus surgical CAse REport (SCARE) guidelines, Int. J. Surg. 60 (2018) 132–136.