

Cavitary Pulmonary Aspergilloma after Norwood-I-Procedure - A Rare Complication after Delayed Sternal Closure in a Patient with HLHS

Robert Wagner^{1*}, Marcel Vollroth², Martin Kostelka², Christian Paech¹, Reinhard Berner³, Ingo Daehnert¹ and Michael Weidenbach¹

¹Department of Pediatric Cardiology, University of Leipzig, Heart Center, Leipzig, Germany.

²Department of Cardiac Surgery, University of Leipzig, Heart Center, Leipzig, Germany.

³Department of Pediatrics, University Hospital, Dresden, Germany.

Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/CA/2017/37814

Editor(s):

(1) Thiago Andrade de Macedo, Hypertension Unit, Cardiology Division Heart Institute (InCor), Brazil.

Reviewers:

(1) Paul Schoenhagen, Imaging Institute, USA.

(2) Damyanti Agrawal, Institute of Medical Sciences, Banaras Hindu University, India.

Complete Peer review History: <http://www.sciencedomain.org/review-history/22286>

Case Study

Received 29th October 2017
Accepted 17th November 2017
Published 14th December 2017

ABSTRACT

The presented case is the first to report on a child with hypoplastic left heart syndrome (HLHS) with invasive aspergillosis (IA) progressing to cavitary pulmonary aspergillosis after Norwood stage I procedure.

Keywords: Invasive pulmonary aspergillosis; hypoplastic left heart; Norwood procedure; delayed sternal closure.

1. INTRODUCTION

We present a case of a newborn with hypoplastic left heart syndrome (HLHS) with invasive

pulmonary aspergillosis that illustrates harms related to inevitable invasive treatment and changes of successful antifungal long-term treatment in such high-risk patients.

*Corresponding author: E-mail: robert.wagner@medizin.uni-leipzig.de;

2. PRESENTATION OF CASE

The child was born at term after prenatal diagnose of HLHS with mitral and aortic atresia. The foramen ovale was severely restrictive with additionally atypical interatrial venous connections. Despite facial dysmorphism standard karyogram and Array-CGH was normal. Even chromosomal deletions on 22q11.2 (CATCH 22) and 10p14 (Di-George-2) or mutations in BRAF and KRAS genes (CFC- and Noonan- syndromes) were ruled out. The child underwent Norwood stage I procedure with a 5 mm RV-PA conduit on day 5 of life. For cardiac insufficiency, postoperative transthoracic veno-arterial ECMO therapy was necessary for 3 days. Delayed sternal closure was on day 6 post-surgery. Prophylactic antibiotics were given according to standard care procedures. Due to rising inflammation markers and clinical signs of infection therapy was escalated to broad spectrum antibiotics. Tracheal aspirate on day 11 after surgery grew aspergillus fumigatus. In addition, galactomannan antigen was positive. Treatment with voriconazole was initiated. 20

days after surgery a hyperlucent lesion on chest x-ray evolved that was further characterized by computer tomography as an aspergilloma of the left lower lobe (Figs. 1 and 2).

Follow-up imaging showed decreasing size of the cavity under long-term oral therapy with voriconazole. Immunodeficiency was ruled out by lymphocyte population count flow cytometry, analysis of immunoglobulin class, IgG subclass, phagocytosis, oxidative burst and neutrophil extracellular trap (NET) testing. The child successfully underwent bidirectional Glenn anastomosis at the age of 6 months. The antifungal therapy was stopped one month thereafter. MRI scanning revealed only scar tissue (Fig. 3).

Three years after discontinuation the child remains in stable clinical condition without residuals on chest x-ray and will soon be evaluated for an implantation of an extracardiac conduit implantation to complete total cavopulmonary anastomosis (TCPC, Fontan circulation).



Fig. 1. Consecutive chest x-rays

From left to right: Initial chest x-ray on day 5 of life, day 11 of life after ECMO therapy was ended and chest closure. On day 21 a faint hyperlucent area can be seen in the left lower lung field that becomes obvious on day 25

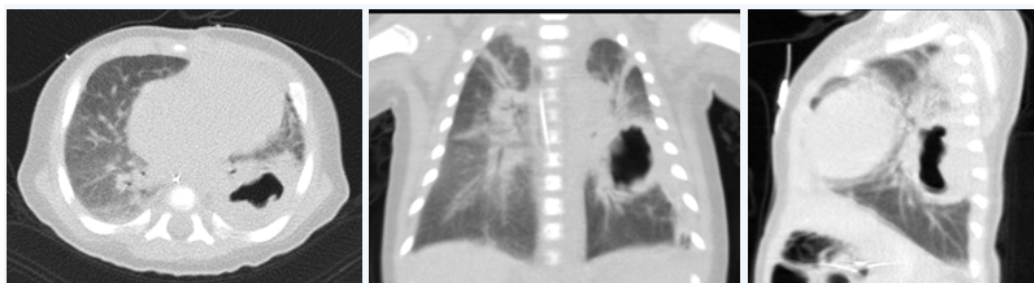


Fig. 2. Chest CT scan with reconstructions on day 35 of life

The size of the cavity is about 2.5 x 1.5 x 3 cm

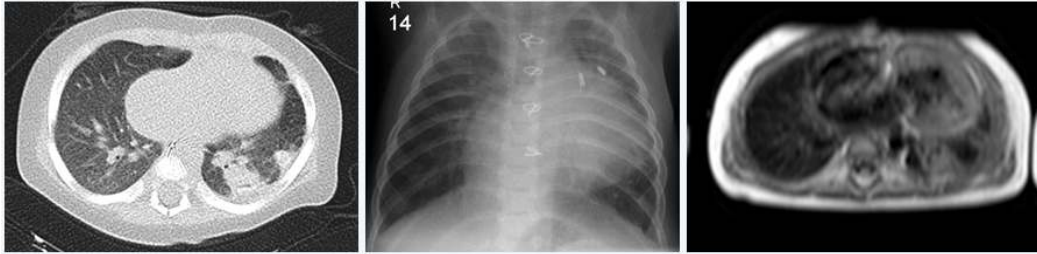


Fig. 3. Chest CT scan at the age of 3.5 months (before Glenn procedure), chest X-ray and chest MRI at the age of 8 months after Glenn procedure (from left to right)

3. DISCUSSION

Nosocomial infections are an increasing problem, including fungal infections. There is still very limited data on how to provide right antibiotic prophylaxis for pediatric patients undergoing cardiac surgery [1–3]. Several risk factors for bacterial surgical site infections with systemic involvement were identified such as complexity of the procedure, younger age at surgery and delayed sternal closure [3]. Fungal surgical site infections with systemic involvement have rarely occurred so far [4]. Thus, antifungal prophylaxis in the context of pediatric cardiac surgery has been even more ambivalently debated. Some authors highlighted its significant value 'in the face of many risk factors' [5]. Risk factors for invasive fungal infections were identified such as prematurity (<32 weeks of gestational age), low birth weight (<1500 g), presence of immune deficiencies, prolonged antibiotic therapy (>5 days), prolonged endotracheal intubation (>3 days), prolonged central vascular catheterization (>7 days), prolonged catecholamine support and delayed sternal closure [5,6].

Although antifungal treatment was initiated in time in the presented case, pneumonia evolved to pulmonary aspergilloma which is an orphan disease state in immunocompetent infants [7]. Retrospectively, several factors other than immunodeficiency put our patient at risk for such an infection that is believed to have high impact on morbidity and mortality. It remains unclear, which specific impact each of these risk factors had, such as neonatal complex cardiac surgery in deep hypothermia on cardiopulmonary bypass, catecholamine use, transthoracic ECMO therapy with open chest for 6 days and broad spectrum antibiotic therapy. Usually, single aspergilloma is treated surgically. In patients undergoing Fontan pathway an optimal pulmonary status is a prerequisite. Since total or subtotal resection of a lung lobe was to put the child at a higher risk in

the long run we opted for long-term antifungal therapy [8,9] that was stopped after 6 months of therapy after successful Glenn procedure. Chest x-ray on follow-up did not show any residuals. While the further course was uneventful the child will soon be evaluated for completion of total cavopulmonary circulation.

4. CONCLUSION

To the authors' knowledge this case represents IA progressing to cavitary pulmonary aspergillosis after Norwood stage I procedure for palliation of HLHS. The authors opted for long-term antifungal therapy rather than surgical treatment options for the treatment of the aspergilloma. Nevertheless, the child's further clinical course after completion of TCPC will indicate the long-term effectiveness of treatment with this approach ('time will tell').

CONSENT

The patient's parents have given their informed consent for the case report to be published.

ETHICAL APPROVAL

As the report is a case presentation formal ethics approval is not applicable. The report has been conducted in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Costello JM, Graham DA, Morrow DF, Morrow J, Potter-Bynoe G, Sandora TJ et

- al. Risk factors for surgical site infection after cardiac surgery in children. *Ann Thorac Surg.* 2010;89(6):1833-41: discussion 1841-2.
DOI: 10.1016/j.athoracsur.2009.08.081
PID 20494036.
2. Murray MT, Corda R, Turcotte R, Bacha E, Saiman L, Krishnamurthy G. Implementing a standardized perioperative antibiotic prophylaxis protocol for neonates undergoing cardiac surgery. *Ann Thorac Surg.* 2014;98(3):927-33.
DOI: 10.1016/j.athoracsur.2014.04.090
PID 25038006.
 3. Bath S, Lines J, Loeffler AM, Malhotra A, Turner RB. Impact of standardization of antimicrobial prophylaxis duration in pediatric cardiac surgery. *J Thorac Cardiovasc Surg.* 2016;152(4):1115-20.
DOI: 10.1016/j.jtcvs.2016.04.091
PID 27245416.
 4. Kansy A, Jacobs JP, Pastuszko A, Mirkowicz-Matek M, Manowska M, Jezierska E et al. Major infection after pediatric cardiac surgery: External validation of risk estimation model. *Ann Thorac Surg.* 2012;94(6):2091-5.
DOI: 10.1016/j.athoracsur.2012.07.079.
PID 23040826.
 5. Jaworski R, Haponiuk I, Irga-Jaworska N, Chojnicki M, Steffens M, Paczkowski K, Zielinski J. Fungal infections in children in the early postoperative period after cardiac surgery for congenital heart disease: a single-centre experience. *Interact Cardiovasc Thorac Surg.* 2016;23(3):431-7.
DOI: 10.1093/icvts/ivw156
PID 27222000
 6. Groll AH, Jaeger G, Allendorf A, Herrmann G, Schloesser R, Loewenich V von. Invasive pulmonary aspergillosis in a critically ill neonate: case report and review of invasive aspergillosis during the first 3 months of life. *Clin Infect Dis.* 1998; 27(3):437-52.
PID 9770138.
 7. Leroy P, Smismans A, Seute T. Invasive pulmonary and central nervous system aspergillosis after near-drowning of a child: Case report and review of the literature. *Pediatrics.* 2006;118(2):e509-13.
DOI: 10.1542/peds.2005-2901
PID 16864641.
 8. Walsh TJ, Lutsar I, Driscoll T, Dupont B, Roden M, Ghahramani P, et al. Voriconazole in the treatment of aspergillosis, scedosporiosis and other invasive fungal infections in children. *Pediatr Infect Dis J.* 2002;21(3):240-8.
PID 12005089.
 9. Scott LJ, Simpson D. Voriconazole: A review of its use in the management of invasive fungal infections. *Drugs.* 2007; 67(2):269-98.
PID 17284090.

© 2017 Wagner et al.; This is an Open Access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Peer-review history:

The peer review history for this paper can be accessed here:
<http://sciedomain.org/review-history/22286>