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SM Journal of Pediatric Surgery

Review Article

Paediatric Achalasia: An Experience at a Peripheral Health Care Centre

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Article Information

Received date: Mar 30, 2018
Accepted date: Apr 23, 2018
Published date: Apr 25, 2018

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Keywords Barium esophagogram; Heller's myotomy; Failure to thrive; Severe malnutrition

Abstract

Introduction: achalasia cardia is an oesophageal motility disorder, typically seen in fifth decade of life. The incidence of the same in the paediatric age is grossly underestimated. The basic pathology is a spastic lower oesophageal sphincter, which causes the symptoms and the resultant sequelae.

Objective: the primary objective of this study was to emphasise the importance of clinical suspicion and barium study in diagnosing achalasia.

Methods: this study is a retrospective observational study. Children presenting with symptoms of regurgitation, malnutrition, recurrent respiratory tract infection and dysphagia were subjected to barium study after a clinical suspicion of achalasia cardia. They were subjected to open Heller's myotomy and fundoplication after confirmation of the diagnosis, which provided excellent results.

Results: 7 patients, suspected to have achalasia cardia, were subjected to surgery, after a barium esophagogram; which showed classical signs of achalasia cardia. 1 patient succumbed to respiratory complications. The patients have been on regular follow up ever since and have been symptom free ever since, have good weight gain.

Conclusion: Achalasia cardia in children is not so uncommon, as we have witnessed in this study. 7 cases were diagnosed over a period of 4 years, at a peripheral health care centre. Barium swallow is a simple, easily available and feasible investigation to identify achalasia cardia and it should be done in children with failure to thrive and vomiting. Heller's myotomy with fundoplication (laparoscopic or open) is the treatment of choice in children.

Introduction

Achalasia is one of the best understood oesophageal motility disorders. It was first described in 1674 by Sir Thomas Willis, and has been studied extensively over the centuries. It usually affects the adult population, the mean age of diagnosis being 50 years with no clear cut sexual preponderance, and is rarely seen in the paediatric population. The primary idiopathic variety is by far the most common type of achalasia. Failure of the lower oesophageal sphincter will cause stasis of the contents, eventually leading to the symptoms and sequelae of achalasia. This study focuses on primary achalasia and its management in a peripheral health care centre.

Materials and methods

This is a retrospective chart review study of pediatric achalasia. All cases of achalasia diagnosed in our centre from 2011 to 2015 were chosen. A total of seven cases of pediatric achalasia cardia, were studied. The diagnosis was based on clinical suspicion and barium esophagogram, classical findings such as delayed emptying of the contrast meal, "bird beak" appearance; dilated esophagus on the study confirmed the diagnosis.

The patients were aged between two months and seven years. Their demographics, clinical features, details of the management and outcomes were studied (Figure 1).

Improvement in general condition following Ryle's tube feeding also supported the diagnosis. Manometric studies and endoscopy were not done because of lack of availability. Management included Heller's myotomy with fundoplication (Figures 2 & 3 and Table 1).

Results

Among the patients included in the study, five were males and two were females. The youngest patient was two months old and the oldest one was seven years old. Open Hellersmyotomy with Dor fundoplication was done in four cases, Hellersmyotomy with Toupets fundoplication were done in two cases. One child, who presented with keratomalacia and pneumonia, succumbed to respiratory complications before she could be taken up for surgery. Intra operative oesophageal perforation occurred in a case, which was identified and dealt with successfully with primary repair.

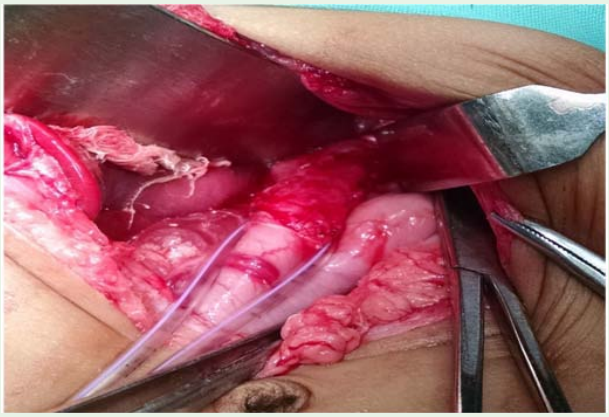

Figure 1: Intra op finding.

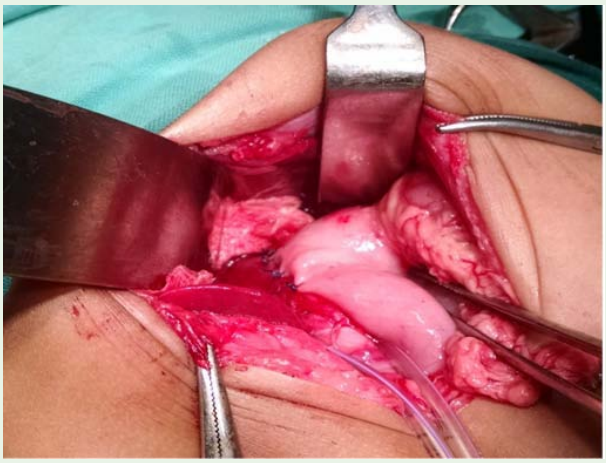

Figure 2: Fundoplication done after Heller's myotomy.

Following surgery, the patients are being followed up regularly on outpatient clinics with the maximum follow up of four years post-surgery. All children operated upon, have had complete resolution of their symptoms and have gained weight.

Discussion

Achalasia literally means "failure to relax" [1]. It was first described by Sir Thomas Willis in 1674 [2], who treated the condition by dilating the lower oesophageal sphincter with a cork tipped

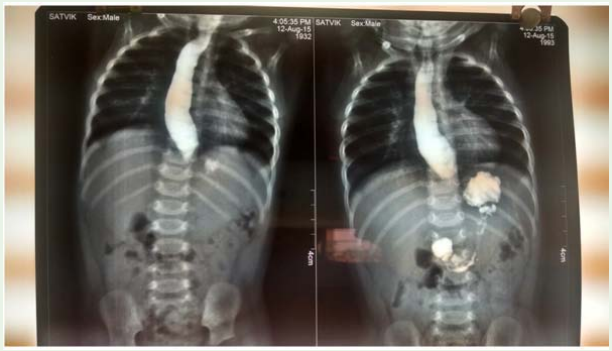

Figure 3: Pre-operative barium swallow.

apparatus. It is currently one of the oesophageal motility disorders that are well understood. Achalasia is a rare motility disorder of the oesophagus characterised by loss of enteric neurons leading to absence of peristalsis and impaired relaxation of the lower oesophageal sphincter [2]. It is a rare disorder affecting 1 per 100,000 general population and just 4-5% of this population is constituted by children and amongst the affected children, less than 1% cases in infants [3]. Achalasia usually presents between fourth to sixth decade of life and is rare in the first two decades.3R King reported the first case of infantile achalasia, in 1953 [3]. The incidence of achalasia cardia in the British population is higher than that of Asian and African population. Among the Asian population, higher incidences have been reported among Indians and Chinese [3].

Achalasia can be primary/ idiopathic or secondary. Primary achalasia is the most common variant. It can be associated with certain syndromes such as Allgrove syndrome (alacrima, achalasia, ACTH deficiency), Down's syndrome, familial visceral neuropathy [2].

Pathogenesis

The pathophysiology is characterized by the failure of the lower esophageal sphincter relaxation, secondary to an imbalance in the excitatory and inhibitory nerve supply that occurs due to neuronal degeneration [4]. Chronic ganglionitis is suggested by the presence of cytotoxic lymphocytes that express activation markers [5] and evidence of compliment activation in the ganglia [6]. In accordance, serum samples of patients with achalasia [7] antibodies against myenteric neurons have been detected, especially in those with HLA DQA1*0103 and DQB1*0603 alleles [8]. HLA proteins are crucial for antigen recognition; thus an aberrant immune response to so far unknown antigens is postulated.

Table 1: Heller's myotomy with fundoplication.

SL NO.	Reference	n	Age (yr)	Procedure	Complications	Treatment	Follow up (m)
1	Pastor et al[34]	40	10.7	6 OHM 3 LHM, 11 LHM + NF 21 Dilatn	1 perforation 2 Perforations	Sutured	75
2	Corda et al[41]	20	12	20 LHM	4 conversion OHM, 5 dysphagia	1 lap LOA, 1 redo LHM, 1 redo OHM	60
3	Esposito et al[43]	31	8.4	31 LHM+DF	3 perforations, 5 dysphagia	2 sutured, 1 redo HM, 2 dilated, 1 redo OHM	9-156
4	Tannuri et al[44]	15	12	15 LHM+DF	2 dysphagia	1 botox injection	32.5
5	Current study	7		2 OHM+TF, 4OHM+DF	1 perforation	Sutured	Mar-48

Note: OHM: Open Heller Myotomy; LHM: Laparoscopic Heller's Myotomy; DF: Dor Fundoplication; TP: Toupet Fundoplication.

Potential viral triggers for the neuronal degeneration are Herpes Simplex Virus 1 (HSV-1), measles, and human papillomavirus as proven by the identification of HSV-1 DNA in oesophageal tissue. Evidence suggests that isolated oesophageal T cells are oligoclonal in nature in achalasia and specifically proliferate to release cytokines on exposure of HSV-1 antigens [9]. HSV-1 is a neurotropic virus with a predilection for squamous epithelium; hence this hypothesis would fit with the selective loss of enteric neurons in the oesophagus. However, HSV-1 DNA was as also identified in the oesophagus of individuals in the control group thereby suggesting that HSV-1 triggers a persistent immune activation with resultant loss of enteric neurons in genetically susceptible individuals [10]. However, other studies have not found HSV-1 or other viruses such as measles or human papillomavirus in oesophageal resection specimens from patients with achalasia [11].

Clinical features

Children usually present with progressive dysphagia, vomiting and weight loss. Younger children and atypical presentation in infants include nocturnal cough, hoarseness, recurrent pneumonia, aspiration and feeding difficulties [12]. Achalasia in children is often misdiagnosed as Gastro-Oesophageal Reflux Disease (GERD). Children frequently present with failure to thrive,asthma, eosinophilicoesophagitis, eating disorders, resulting in a delay in diagnosis for as long as 6-10 years [12]. Up to 50% of children undergo treatment with antacids or prokinetics before the diagnosis of achalasia is identified [13].

Investigations

Due to the significant delay in diagnosis of achalasia in children, a barium oesophagogram study alone is diagnostic4. Barium swallow classically demonstrates a dilated proximal oesophagus with "bird's-beak" like tapering of the distal oesophagus. It may also be helpful in cases where oesophageal manometryprovides equivocal findings. An oesophagogram is also useful to assess for late- or end-stage achalasia changes (tortuosity, angulation, and megaesophagus) that have implications for treatment [14].

A Timed Barium Oesophagogram is done to study the accurate emptying of oesophagus and is an essential investigation in the evaluation of motor dysphagia, often caused by achalasia [15]. Barium studies are inexpensive and readily available. The oesophageal emptying was initially demonstrated by measuring barium column height at 1 and 5 minutes after ingestion of a large barium bolus in an upright position; an approach that has come to be known as the " Timed Barium Oesophagogram (TBE) " [16]. Achalasia is suggested by the partial emptying or persistent barium at the 5 minutes film on a timed barium oesophagogram [15]. Subsequent studies suggested usefulness of TBE for the objective evaluation of achalasia patients, post treatment, as it helps identify patients who are more likely to have relapse of symptoms despite an initial symptomatic improvement [17]. In our patients, the film taken at the end of 5 minutes showed incomplete emptying, and lengthy columns of barium in the oesophagus persisted even in the films taken after 15 minutes of ingestion, thereby suggesting achalasia cardia.

The role of endoscopy in the work up of achalasia in a child is mainly to rule out oesophagitis and to diagnose Chagas disease. It is used to rule out the secondary causes of achalasia. Findings on endoscopy, in achalasia may range from a seemingly normal examination to a tortuous dilated sigmoid oesophagus with pooled secretions and retained food particles. Thus, in those patients with a non-dilated oesophagus endoscopy might not be sensitive and oesophageal motility test is indicated if there is clinical suspicion for achalasia. Endoscopy also plays a role in patients with recurrence of symptoms following therapy, to evaluate for return of puckered EGJ *versus* reflux-induced stricturing from GERD [18].

Thus, normal findings on EGD or oesophagogram in patients suspected of having achalasia should prompt oesophageal motility testing. However classic endoscopic and/or oesophagogram findings, an oesophageal motility test would be considered supportive to confirm the diagnosis [18]. The manometric finding of incomplete LES relaxation with no evidence of a mechanical obstruction and aperistalsis solidifies the diagnosis of achalasia in the appropriate setting [19]. Other findings, such as an elevated baseline oesophageal body pressure, increased basal LES pressure and simultaneous non propagating contractions, can also support the diagnosis of achalasia but are not required for making the diagnosis [20].

The American College of Gastroenterology recommendations for diagnosis of achalasia14, for adults are:

All patients with suspected achalasia who do not have evidence of a mechanical obstruction on endoscopy or esophagram should undergo esophageal motility testing before a diagnosis of achalasia can be confirmed (strong recommendation, low-quality evidence).

The diagnosis of achalasia is supported by oesophagogram findings including dilation of the oesophagus, a narrow esophago-gastric junction with "bird-beak " appearance, aperistalsis and poor emptying of barium (strong recommendation, moderate-quality evidence).

Barium oesophagogram is recommended to assess oesophageal emptying and oesophago gastric junction morphology in those with equivocal motility testing (strong recommendation, low-quality evidence).

Endoscopic assessment of the gastro-oesophageal junction and gastric cardia is recommended in all patients with achalasia to rule out pseudo achalasia (strong recommendation, moderate-quality evidence).

Treatment

Treatment options include medical management, endoscopic procedures and surgical procedures.

Medical management

Nifedipine: a calcium channel blocker is primarily used in adults. Its role in paediatric achalasia is not well documented. Maksimak et al [21] reported 4 children having symptomatic relief following intake of nifedipine before meals likely due to a decrease in resting LES pressure. Nifedipine is to be used as a bridge to relieve symptoms until other modalities such as pneumatic dilatation, Botox injection or myotomy can be performed, and is not a definitive therapy, neither in adults nor in children [22].

Table 2: Efficacy and safety in pediatric age group.

SL No.	Age	Sex	Weight	Complaints	Treatment	complications	Recurrence
1	4 and half months	Male	5 kg	Vomiting, failure to thrive	OHM + DP	None	None
2	2 months	Male	2 kg	malnutrition	OHM + DP	None	None
3	3 and half years	FeMale	12 kg	Recurrent chest infection	OHM+TP	None	None
4	7 years	Male	15 kg	Failure to thrive	OHM +TP	None	None
5	1 year	FeMale	4.5 kg	Vomiting, keratomalacia	-	Death due to pneumonia before surgery	
6	8 months	Male	4 kg	Vomiting, failure to thrive	OHM+DP	None	None
7	5 years	Male	10 kg	Vomiting, dysphagia	OHM+DP	None	None

Note: OHM: Open Heller's Myotomy; TP: Toupet's procedure; DP: Dors Fundoplication Procedure; Kg: kilogram.

Endoscopic 4 quadrant botulinum injections: it has been proven effective in adults, providing permanent symptomatic relief in 10% to 40 % of adults. However a subsequent surgery is required in majority [23]. Optimal dosing and injection frequency of botulinum toxin to relieve achalasia symptoms in children has not been well defined [4].

Pneumatic dilatation: Multiple dilatations are often required to achieve successful symptomatic relief although an initial response can be used to predict the success or failure of the subsequent dilatations. Hamzaet al [23] reported a 90% success rate in children treated with multiple pneumatic dilations. The advantages of balloon dilatation include shorter hospital stay, decreased cost and quick recovery [24]. Complications of pneumatic dilatation include sub sternal pain, prolonged epigastric pain, oesophageal perforation, aspiration pneumonia and GERD [24]. Multiple studies suggest that pneumatic dilatation is safe and effective modality of initial treatment of achalasia in older, hence preventing a surgical intervention. However there are no long term follow up studies to document the success rates of pneumatic dilatation for achalasia in children [25]. Recurrence of symptoms in children despite multiple dilatations warrants surgical myotomy.

Surgical management

Despite having multiple treatment options, the most successful and definitive treatment option available is surgery. Surgical options include:

Heller's myotomy with or without fundoplication. A longitudinal myotomy, about 5 cm above the OG junction and extending about 2 cms into the cardia of the stomach is done in Heller's myotomy. Laparoscopic Heller's myotomy is the treatment of choice for achalasia in children and adults [26,27]. The laparoscopic approach provides the advantages of better visualisation of the operative field, shorter hospital stay, minimal pain, superior cosmetic results and an early return to normal routine [27].

The 2 main complications associated with surgery are oesophageal perforation and recurrence of dysphagia. Recurrence of dysphagia varied from 0% to 26%, with or without an anti-reflux procedure in various studies [28]. Studies report a fall in recurrence of dysphagia with growing experience of the operating surgeon [29]. In our study the recurrence of dysphagia following surgery has not been reported to date.

POEM: Per Oral Endoscopic Myotomy, follows the principle of natural ortifice trans luminal endoscopic surgery (NOTES). A mucosal incision is taken and dissection is done to create a sub mucosal tunnel in the distal oesophageal wall, to approach the oesophago- gastric junction. A 2-3 cm longitudinal incision in the inner circular muscle approximately 4 cm from the LES, will produce similar results to Heller myotomy [30]. 24 hours, following the procedure, an oesophagogram is performed and patient is started on soft diet [30]. However its efficacy and safety in pediatric age group is yet to be determined Table 2.

Conclusion

Achalasia cardia in children is not so uncommon, as we have witnessed in this study. 7 cases were diagnosed over a period of 4 years, at a peripheral health care centre. Barium swallow is a simple, easily available and feasible investigation to identify achalasia cardia and it should be done in children with failure to thrive and vomiting. Heller's myotomy with fundoplication (laparoscopic or open) is the treatment of choice in children.

References

1. Malangoni MA, Rosen MJ. Hernias. In: Townsend, Beauchamp, Evers, Mattox, editors. Sabiston Textbook of Surgery. Philadelphia: Saunders Elsevier; 2012; 2: 1025.
2. Boeckxstaens GE, Zaninotto G, Richter JE. Achalasia. Lancet. 2014; 383: 83-93.
3. Rai GS, Sarawagi R, Sharma S, Apte A. An infant with nasal regurgitation since birth and failure to thrive. J Clin Diagn Res. 2015; 9: TD03- TD04.
4. Franklin AL, Petrosyan M, Kane TD, Childhood achalasia: A comprehensive review of disease, diagnosis and therapeutic management. World J Gastrointest Endosc. 2014; 6: 105-111.
5. Clark SB, Rice TW, Tubbs RR. The nature of the myenteric in filterate in achalasia: an immunohistochemical analysis. Am J Surg Pathol. 2000; 24: 1153-1158.
6. Storch WB, Eckardt VF, Junginger T. Compliment components and terminal complement complex in esophageal smooth muscle of patients with achalasia. Cell MolBiol(Noisy-le-grand). 2002; 48: 247-52.
7. Storch WB, Eckardt VF, Wienbeck M, Eberl T, Auer PG, Hecker A, et al. autoantibodies to Auerbach's plexus in achalasia. Cell Mol Biol (Noisy-le-grand). 1995; 41: 1033-1038.
8. Ruiz-de-León A, Mendoza J, Sevilla-Mantilla C, Fernández AM, Pérez-de-la-Serna J, Gónzalez VA, et al. Myenteric antiplexus antibodies and class II HLA in achalasia. Dig Dis Sci. 2002; 47: 15-19.

9. Castagliuolo I, Brun P, Costantini M, Rizzetto C, Palù G, Costantino M, et al. Esophageal achalasia: is herpes simplex virus really innocent? J Gastrintest Surg. 2004; 8: 24-30.

10. Boeckxstaens GE. Achalasia: virus-induced euthanasia of neurons? Am J Gastroenterol. 2008; 103: 1610-1612.

11. Birgisson S, Galinski MS, Goldblum JR, Rice TW, Richter JE. Achalasia is not associated with measles or known herpes and human papilloma viruses. Dig Dis Sci. 1997; 42: 300-306.

12. Hallal C, Kieling CO, Nunes DL, Ferreira CT, Peterson G, Barros SG, et al. Diagnosis, misdiagnosis, and associated diseases of achalasia in children and adolescents: a twelve-year single center experience. Pediatr Surg Int. 2012; 28: 1211-1217.

13. Lee CW, Kays DW, Chen MK, Islam S. Outcomes of treatment of childhood achalasia. J Pediatr Surg 2010; 45:1173-1177.

14. ACG guidelines.

15. Ghoshal UC, Gupta M, Verma A, Neyaz Z, Mohindra S, Misra A, et al. High-resolution manometry is comparable to timed barium esophagogram for assessing response to pneumatic dilation in patients with achalasia. 2015; 34:144-151.

16. de Oliveira JM, Birgisson S, Doinoff C et al. timed barium swallow: a simple technique for evaluating esophageal emptying in patients with achalasia. AJR Am J Roentgenol 1997; 169: 473-479.

17. Vaezi MF, Baker ME, Richter JE. Assessment of esophageal emptying post-pneumatic dilation: use of the timed barium oesophagogram. Am J Gastroenterol. 1999; 94: 1802-1807.

18. Ates F, Vaezi MF. The Pathogenesis and Management of Achalasia: Current status and Future directions. Gut Liver. 2015; 9: 449-463.

19. Pandolfino JE, Kahrilas PJ. AGA technical review on the clinical use of esophageal manometry. Gastroenterology. 2005; 128: 209-224.

20. Spechler SJ, Castell DO. Classification of oesophageal motility abnormalities. Gut. 2001; 49: 145-151.

21. Makismak M, Perlmutter DH, Winter HS. The use of nifedipine in treatment of achalasia in children. J Peditr Gastroenterol Nutr. 1986; 5: 883-886.

22. Chuah SK, Hsu PI, Wu KL, Wu DC, Tai WC, Changchien CS. 2011 update on esophageal achalasia. World J Gastroenterol. 2012; 18: 1573-1578.

23. Pastor AC, Mills J, Marcon MA, Himidan S, Kim PC. A single centre 26-year experience with treatment of esophageal achalasia: is there an optimal method? J Pediatr Surg. 2009; 44: 1349-1354

24. Babu JT, Cohen S, Watkins JB. Successful treatment of achalasia in childhood by pneumatic dilatation. Pediatr Surg Int. 2001; 17: 505-507.

25. Hussain SZ, Thomas R, Tolia V. A review of achalasia in 33 children. Dig Dis Sci. 2002; 47: 2538-2543.

26. Rosemurgy AS, Morton CA, Rosas M, Albrink M, Ross SB. A single institution's experience with more than 500 laparoscopic Heller's myotomies for achalasia. J Am Coll Surg. 2010; 210: 645-647.

27. Salvador R, Costantini M, Cavallin F, Zanatta L, Finotti E, Longo C, et al. Laparoscopic Heller myotomy can be used as a primary therapy for oesophageal achalasia regardless of age. J Gastro intest Surg. 2014; 18: 106-111.

28. Esposito C, Riccipetitoni G, Chiarenza SF, Roberti A, Vella C, Alcchio F, et al. Long-term results of laparoscopic treatment of esophageal achalasia in children: a multicentric survey. J Laparoendosc Adv Surg Tech A. 2013; 23: 955-959.

29. Rothenberg SS, Patrick DA, Bealer JF, Chang JH. Evaluation of minimally invasive approaches to achalasia in children. J Pediatr Surg. 2001; 36: 808-810.

30. Espocita C, CucchiaraS, Borrelli O, Roblot-Margret B, Desruelle P, Montupet P. Laparoscopic esophagomyotomy for the treatment of achalasia in children. A preliminary report of eight cases. Surg Endosc. 2000; 14: 110-113.