

Congenital Segmental Emphysema: An Evolving Lesion

Authors

S. Paramalingam¹, E. Parkinson¹, M. Sellars², S. Diaz-Cano³, K. H. Nicolaidis¹, M. Davenport⁵

Affiliations

Affiliation addresses are listed at the end of the article

Key words

- congenital lung anomalies
- cystic adenomatoid malformation
- bronchial atresia

Abstract

Introduction: Congenital segmental emphysema (CSE) is a newly-recognised sub-type of congenital parenchymal lung anomaly. It is characterised by antenatal detection and post-natal evolution from an initially solid segmental appearance to a hyperlucent and hyperinflated segment.

Methods: A retrospective review of a single-centre tertiary referral database between Jan 1994 and Dec 2007 was performed.

Main Results: 130 infants had antenatally detected lung anomalies, and of these 12 (9.2%) infants (initially labelled as congenital cystic adenomatoid malformation (CCAM)), showed features better defined as CSE. The lesions were described antenatally as non-progressive microcystic (n=6), hyperechogenic (n=2) or both (n=2). Early post-natal CT scans showed areas

of solid segmental parenchyma, initial hyperlucency or microcysts. Subsequent CT imaging, however, showed evolution to segmental hyperlucency in areas previously solid and in 2 cases a central bronchocele was noted. Ten children underwent resectional surgery (segmentectomy n=4, lobectomy n=6) at a median age of 1 (range 0.4–5.2) year and the gross appearance of the resected specimen confirmed hyperinflated (not cystic) segments. Histological review showed localised abnormally dilated alveolar spaces in 7 cases. Adjacent areas consistent with type 2 CCAM were also seen (n=3).

Conclusion: CSE lies within the spectrum of both CCAM and sequestration but there is a definite post-natal evolution and volume change which presage symptoms. This may be associated with segmental bronchial atresia and progressive air trapping via collateral airways such as the interalveolar pores of Kohn.

received October 15, 2009
accepted after revision
November 14, 2009

Bibliography

DOI <http://dx.doi.org/10.1055/s-0029-1246129>
Eur J Pediatr Surg 2010; 20: 78–81 © Georg Thieme Verlag KG Stuttgart · New York
ISSN 0939-7248

Correspondence

Prof. Mark Davenport
King's College Hospital
Department of Paediatric Surgery
SE5 9RS Denmark Hill
London
United Kingdom
Tel.: +44 (0)20 3299 3350
Fax: +44 (0)20 3299 4021
markdav2@ntlworld.com

Introduction

Routine antenatal ultrasound scanning in the UK has increased the detection rate of parenchymal lung lesions [4]. Most commonly, these lesions are congenital cystic adenomatoid malformations (CCAM) or bronchopulmonary sequestrations (BPS) [1,2,4,14], or on occasion, hybrids with features of both [3]. More rarely, these lesions have been reported to be congenital lobar emphysema [10] or bronchogenic cysts [9]. Increasing familiarity with these pathologies suggests that there is a new type of parenchymal lung lesion not adequately covered by the above differential descriptions. Our aim is to characterise this lesion and propose the name *congenital segmental emphysema* (CSE).

Methods

A retrospective review was performed of all cases diagnosed with congenital parenchymal lung lesions at our institution between the period of Jan 1994 and Dec 2007. The following data were compiled: antenatal ultrasonographic features and behaviour; post-natal clinical symptomatology, post-natal imaging and operative records. Histological specimens were reviewed retrospectively. Pre-natal lesions were described according to the Adzick classification [1]. Post-natal CCAM was described according to a modified Stocker classification [15].

In retrospect, the hallmark for the diagnosis of CSE appeared to be radiological hyperlucency (as distinct from the macroscopic cystic appearance of CCAM) which appeared to evolve from segmental and sub-segmental areas apparently solid

Table 1 Summary of clinical findings.

	Sex	Antenatal findings	Postnatal findings (CT)	Surgery	Age at surgery (yrs)	Notes
1	F	microcystic	LUL* – hyperlucent & bronchocele	SEG (LUL)	3.4	adjacent CCAM. bronchial atresia
2	F	microcystic	LLL – solid and hyperlucent	–	N/A	
3	F	microcystic & hyperechogenic	LLL – solid to hyperlucent	lobectomy	8.5	
4	M	microcystic	LUL – hyperlucent and cystic	lobectomy	2.5	
5	M	undefined	RML/RLL – solid to hyperlucent	SEG (RML/RLL)	0.4	
6	M	microcystic & hyperechogenic	LUL/LLL – solid/cystic/hyperlucent	SEG (LU/LL/Li)	2.1	adjacent CCAM
7	M	microcystic	RLL – hyperlucent	SEG (RLL)	0.9	
8	M	microcystic	L midzone – segmental cystic and hyperlucent	–	N/A	
9	F	hyperechogenic	LUL – solid and hyperlucent	lobectomy	0.7	adjacent CCAM
10	M	hyperechogenic	LUL – solid and hyperlucent	lobectomy	0.8	bronchial atresia
11	F	'possibly' solid	LLL – solid/cystic/hyperlucent	lobectomy	1.1	accessory vessel
12	M	microcystic	RLL – hyperlucent & bronchocele	lobectomy	2.5	

Key: *affected lesions were all sub-lobar but are described as the lobe of origin

LUL: left upper lobe. Li: lingual. LLL: left lower lobe. RML: right middle lobe. RL: right lower lobe. SEG: segmentectomy

on initial CT imaging. This became our criteria for diagnosis of this lesion. Data are given as median and range.

Results

In the 13-year period, 130 infants had antenatally detected parenchymal lung lesions. From this retrospective series, 12 (8 male) infants (9.2%) fitted our criteria for the diagnosis of CSE. Most (n=9; 75%) had left-sided lesions.

Antenatal findings

Median age at antenatal detection was 20 (19–24) weeks. Descriptive features included non-progressive microcystic (n=6), non-progressive hyperechogenic (n=2) or features of both (n=2). The diagnosis was believed to be consistent with CCAM in all except one case. Case 11 appeared to have some ambiguity, and a provisional label of BPS was used antenatally as the predominant feature on antenatal ultrasound was that of a solid lesion (Table 1).

All except one of the infants were born at term without incident and were initially asymptomatic. One infant (of triplets) was born at 34 weeks' gestation and required a short period (<48 h) of non-invasive ventilation for respiratory distress at birth due to prematurity. Initial assessment with plain chest radiography was normal in all cases.

Four children became symptomatic (exercise-induced dyspnoea and recurrent upper respiratory tract infections) under subsequent out-patient review. The remainder appeared asymptomatic.

Radiological findings

All infants underwent post-natal CT imaging at about one month according to our institutional protocol for asymptomatic infants. This tended to show areas of solid segmental parenchyma with areas of hyperlucency and/or microcysts. Subsequent CT scans however showed progressive hyperlucency in areas previously solid. (Fig. 1, 2) and the development of a centrally located bronchocele (n=2). Table 1 refers to the lobe within which the abnormal segment was located (sometimes multiple).

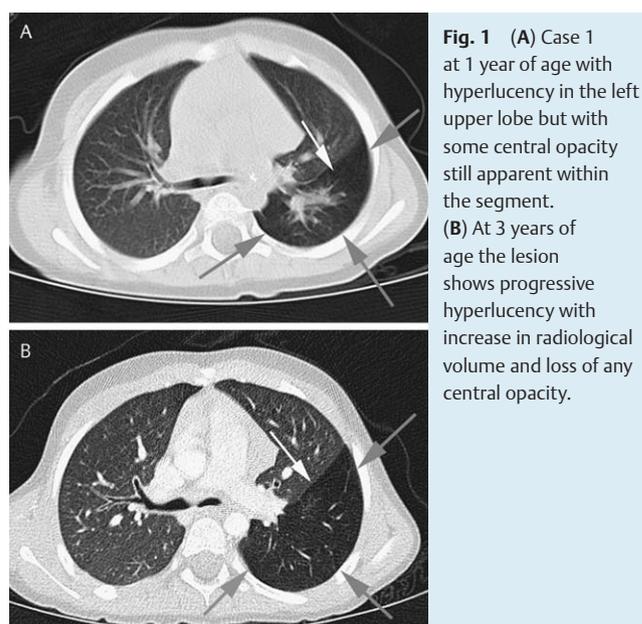


Fig. 1 (A) Case 1 at 1 year of age with hyperlucency in the left upper lobe but with some central opacity still apparent within the segment. (B) At 3 years of age the lesion shows progressive hyperlucency with increase in radiological volume and loss of any central opacity.

Operative findings

Ten children underwent thoracotomy at a median age of 1 (0.4–5. 2) year. Resectional surgery included segmentectomy (n=4) and formal lobectomy (n=6). The gross appearance at operation confirmed the hyperinflated segmental appearance described with imaging. Other pathologies were certainly evident and included Type II CCAM (n=3) and, an accessory vessel from the aorta (n=1, diagnosed antenatally as BPS).

There were no postoperative complications and at follow-up (median 6 [2–60] months), all patients are asymptomatic. Two children did not undergo surgery, one of whom (Case #2) has declined follow-up, but was well at last evaluation. The other child (Case #8) is now 12 years of age, remains asymptomatic and the most recent CT scan shows the lesion to be unchanged in character; if anything, it has undergone volume loss (Fig. 3).

Histological findings

Retrospective examination of all material excised consistently showed localised areas of dilated air spaces. Actual bronchial atresia and bronchocele formation proved more difficult to identify histologically, although the latter was easily appreciated at operation as central mucus-filled cavities. The lung parenchyma



Fig. 2 Case 3 at one year of age (A) showing solid areas in the left lower lobe. (B) Lesion becomes progressively hyperlucent, with an increase in radiological volume, at 5 years of age. (It should be noted that this child became symptomatic with exertional dyspnoea at this point).



Fig. 3 Case 12 showing CT scan at aged 1 yr (A) and 2.5 yrs (B, C). There is segmental emphysema affecting the right upper lobe posteriorly in all, with an expanding centrally-placed bronchocele, not apparent on earlier scan. Contents shown to be sterile at the time of surgery.

of the resected specimens was fully differentiated, suggesting that this is not CCAM or BPS, as these are characterised by the presence of immature lung tissue. However, areas more typical of Type II CCAM were histologically identifiable in adjacent lung tissue in at least three of the resected specimens (Cases # 1, 6 and 9) (○ Fig. 4).

Discussion

We describe a sub-type of congenital parenchymal lung pathology which we term *congenital segmental emphysema*, as this appears to the main radiological feature (if allowed to evolve), typically (but not exclusively) with a segmental rather than lobar distribution. Progressive hyperlucency on CT-imaging is the hallmark, confirmed at operation and by histological examination as hyperinflated (not cystic) segments. Nonetheless these lesions still appear to be within the spectrum of CCAM and BPS as these can exist simultaneously but, in contrast to both, there is a definitive post-natal increase in the volume of the segment due to hyperinflation, leading to symptoms in some cases.

Recently published work [11,16] from Philadelphia, a foetal medicine centre comparable to our own, suggests recognition of this same entity, although they clearly favour the term “peripheral bronchial atresia” [11]. In the first reported series of asymptomatic cases subjected to resectional surgery (largely before their first birthday), classical CLE accounted for only about 2% of all lesions but other variants of “emphysematous lung” lesions were also listed which could well include cases we would now recognise as CSE [16]. In the second report [11], 16 infants (11 coming to surgical resection) fitted radiological criteria of non-cystic hyperlucency (i.e. CSE). In those for whom histological data is available, actual PBA was identified in 82% and about half had pathological features of CCAM. A further retrospective histological review of the entire series showed an additional 14 patients with actual PBA, but without radiological hyperlucency. We much prefer our term as this is the main feature obvious on delayed post-natal CT imaging, and seemingly actual PBA is not exclusively associated with hyperlucency.

Bronchial atresia is not new and has long been recognised as a distinct clinical entity [13], but it has recently been implicated by Riedlinger et al. as a common and possibly aetiological factor

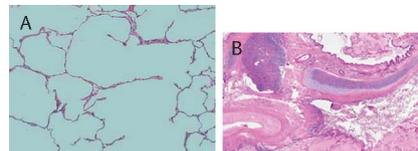


Fig. 4 Photomicrographs of resected lobe showing grossly dilated airspaces (alveoli) with stretching of the interstitium consistent with emphysematous change (A) together with collapsed bronchus (arrowed) and fully differentiated lung parenchyma (B) (H&E, magnification ×400).

in CCAM, BPS and lobar emphysema [12]. Certainly this would be an attractive pathological mechanism for CSE, although we only actually noted it in two cases. We suspect that segmental bronchial atresia is difficult to demonstrate in retrospect using standard histological techniques. In contrast, a bronchocele distal to the atresia appears consistently present and may be obvious both radiologically and at surgery, although less so histologically. We speculate that the characteristic evolution of CSE from solid into hyperlucency is actually due to progressive air entrapment from collateral adjacent airways via the pores of Kohn [6], if left long enough.

Classical congenital lobar emphysema (CLE) is a relatively unusual cause of antenatally detected lung lesions. A retrospective review from Toronto identified 20 children with radiological CLE, although an antenatal lesion was detected only in eight [8]. The authors took a predominantly conservative approach, but unlike our experience no further radiological changes could be demonstrated, indeed resolution occurred in one case. The commonest site in their series was the left upper lobe. The variable pathological features of symptomatic and post-natally detected lobar emphysema have been documented by Mani et al. from the Armed Forces Institute of Pathology [7], who noted the almost invariable location in the upper lobes and the degree of overlap with polyalveolar lung. Left-sided lesions were found in three quarters of our patients in this series, with over 40% being in the left upper lobe; nonetheless CSE also occurred consistently in the lower lobes.

We took an initially conservative view of these lesions, avoiding intervention and promoting serial CT scans. Most were initially small, predominantly solid and innocuous-looking. However, in

up to half there was clear segmental expansion which in one child coincided with the onset of symptoms, cured by surgical excision. We feel it is likely that most will come to resectional surgery at some point, and in most cases this can be accomplished by segmentectomy rather than lobectomy. We appreciate that the need for surgical intervention is a controversial area [14] with some authors advocating resection for all antenatally detected lesions irrespective of symptoms [16], while others advocating almost only conservative treatment [8], particularly if presenting beyond the neonatal period [5]. The recognition pre-operatively that this is a segmental and hence localised disease allows lung-sparing procedures such as segmentectomy to be planned. If a conservative view is taken then, because CSE is a lesion in evolution, close follow-up in the form of clinical assessment and serial radiological imaging is crucial.

Conflict of Interest: None

Affiliations

¹King's College Hospital, Paediatric Surgery, London, United Kingdom

²King's College Hospital, Radiology, London, United Kingdom

³King's College Hospital, Pathology, London, United Kingdom

⁴King's College Hospital, Fetal Medicine, London, United Kingdom

⁵King's College Hospital, Department of Paediatric Surgery, London, United Kingdom

References

- 1 Adzick NS, Harrison MR, Crombleholme TM et al. Fetal lung lesions: management and outcome. *Am J Obstet Gynecol* 1988; 179: 884–889
- 2 Adzick NS, Harrison MR, Glick PL et al. Fetal cystic adenomatoid malformation: prenatal diagnosis and natural history. *J Pediatr Surg* 1985; 20: 483–488
- 3 Cass DL, Crombleholme TM, Howell LJ et al. Cystic lung lesions with systemic arterial blood supply: a hybrid of congenital cystic adenomatoid malformation and bronchopulmonary sequestration. *J Pediatr Surg* 1997; 32: 986–990
- 4 Davenport M, Warne SA, Cacciaguera S et al. Current outcome of antenatally diagnosed cystic lung disease. *J Pediatr Surg* 2004; 39: 549–556
- 5 Karnak I, Senocak ME, Ciftci AO et al. Congenital lobar emphysema: diagnostic and therapeutic considerations. *J Pediatr Surg* 1999; 34: 1347–1351
- 6 Kohn HN. Zur Histologie der indurierenden fibrinösen Pneumonie. *Münchener Medicinische Wochenschrift* 1893; 40: 42–45
- 7 Mani H, Suarez E, Stocker JT. The morphologic spectrum of infantile lobar emphysema: a study of 33 cases. *Paediatr Respir Rev* 2004; 5 (Suppl A): 313–320
- 8 Mei-Zahav M, Konen O, Manson D et al. Is congenital lobar emphysema a surgical disease? *J Pediatr Surg* 2006; 41: 1058–1061
- 9 Nobuhara KK, Gorski YC, La Quaglia MP et al. Bronchogenic cysts and oesophageal duplication: common origins and treatment. *J Pediatr Surg* 1997; 32: 1408–1413
- 10 Olutayo OO, Coleman BG, Hubbard AM et al. Prenatal diagnosis and management of congenital lobar emphysema. *J Pediatr Surg* 2000; 35: 792–795
- 11 Peranteau WH, Merchant AM, Hedrick HL et al. Prenatal course and postnatal management of peripheral bronchial atresia: association with congenital cystic adenomatoid malformation of the lung. *Fetal Diagn Ther* 2008; 24: 190–196
- 12 Riedlinger WF, Vargas SO, Jennings RW et al. Bronchial atresia is common to extralobar sequestration, intralobar sequestration, congenital cystic adenomatoid malformation and lobar emphysema. *Pediatr Develop Pathol* 2006; 9: 361–373
- 13 Schuster SR, Harris GB, Williams A et al. Bronchial atresia: a recognizable entity in the pediatric age group. *J Pediatr Surg* 1978; 13: 682–689
- 14 Stanton M, Davenport M. Management of congenital lung lesions. *Early Human Development* 2006; 82: 289–295
- 15 Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung: classification and morphologic spectrum. *Hum Pathol* 1977; 8: 155–172
- 16 Tsai AY, Liechty KW, Hedrick HL et al. Outcomes after postnatal resection of prenatally diagnosed asymptomatic cystic lung lesions. *J Pediatr Surg* 2008; 43: 513–517