Original Article

Congenital cystic lesions of the lungs: The perils of misdiagnosis – A single-center experience

V. Shankar Raman, Sandeep Agarwala, Veereshwar Bhatnagar, Shasanka Shekhar Panda, Arun Kumar Gupta[1]

Department of Paediatric Surgery and [1]Radiodiagnosis, All India Institute of Medical Sciences, New Delhi, India

ABSTRACT

Background: A majority of cystic lesions in the western world are detected antenatally, whereas, the diagnosis in our setup occurs once the child becomes symptomatic. Surgical management is primarily dictated by the presence of symptoms, recurrent infection, and rarely by the potential risk of malignant transformation. **Materials and Methods:** A retrospective analysis was carried out on all consecutive patients with cystic lung lesions managed at our center from January 2000 through June 2011 for antenatal diagnosis, presentation, diagnostic modalities, treatment, and complications. **Results:** Forty cystic lung lesions were identified. Only 8% were antenatally detected. Out of 40, the final diagnosis was congenital cystic adenomatoid malformation in 19, congenital lobar emphysema in 11, and bronchogenic cysts and pulmonary sequestration in five each. Of these, 20% had received a course of prior antitubercular therapy and 30% had an intercostal drain inserted prior to referral to our center. Postoperative morbidity in the form of bronchopleural fistula, pneumothorax, and non-expansion of the residual lung was noted in 10% of the patients. **Conclusion:** Antenatal diagnosis of these lesions is still uncommon in third world countries. Prior to referral to a pediatric surgical center a large number of patients received antitubercular drugs and an intercostal drain insertion, due to incorrect diagnosis.

KEY WORDS: Antenatal diagnosis, antitubercular drugs, congenial cystic lung lesions, intercostal drainage

Address for correspondence: Prof. Sandeep Agarwala, Department of Paediatric Surgery, All India Institute of Medical Sciences, New Delhi - 110 029, India.
E-mail: sandpagr@hotmail.com

INTRODUCTION

Cystic lesions of the lungs encompass a wide spectrum of rare lung lesions comprising of congenital cystic adenomatoid malformation (CCAM), congenital lobar emphysema (CLE), bronchopulmonary sequestration, and bronchogenic cyst. CCAM with an incidence of 1 in 10,000 to 25,000 pregnancies is the most common lung cyst.[1] An abnormal antenatal ultrasound scan has now become the most common mode of presentation in Europe and the United States. In a small proportion of the cases these lesions may cause polyhydramnios, hydrops, and even intrauterine death.[2] Fetal intervention, like thoracoamniotic shunts, laser ablation, and resectional surgery, is a possibility in some of these conditions.[3] Postnatally a child may be asymptomatic, develop respiratory distress or recurrent infections.[4] Symptomatic lesions need to be managed by surgical resection.[5] Management of an asymptomatic lesion is controversial; while some advocate surgical excision,[6] others recommend resection, only when symptoms occur.[7] This study retrospectively reviews our experience of congenital cystic lung lesions in terms of antenatal diagnosis, etiologies, presentation, management and complications.

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/0970-2113.152616

MATERIALS AND METHODS

The records of all patients managed at our center with a diagnosis of cystic lung lesions from January 2000 to June 2011 were reviewed. The following data were compiled: Antenatal diagnosis, symptomatology, treatment received prior to referral to our center, imaging findings, location, and type of lesions, type of surgery, postoperative ventilation, complications, and outcomes.

RESULTS

For the 10-year study period, 40 symptomatic patients with a diagnosis of congenital cystic lung lesions were

managed at our center. Only three (8%) patients were diagnosed antenatally. Among the remaining, 34 had no antenatal ultrasound scan performed and it was reported as normal in three patients. Out of the three patients with an antenatal diagnosis, two became symptomatic at birth and one at 48 months. Twenty-nine of the 40 (73%) patients were male and 11 (27%) were female and the median age of presentation was 25 months.

All patients were investigated with a standard chest X-ray (CXR) and computed tomography (CT) scan. The CXR was accurate in identifying a cystic lesion in 18 patients (45%) compared with a CT scan, which picked up the lesion in all 40 patients (100%). Nineteen (47.5%) children became symptomatic in the neonatal period, 13 (32.5%) between one month and one year, seven (18%) between one and five years, and one (2%) beyond five years of age. Respiratory distress was the most common presenting symptom in children less than one year of age, while respiratory infections were common in children presenting after one year of age. Eight (20%) children had received a full course of prior antitubercular therapy due to misdiagnosis. Twelve (30%) children had an intercostal drain (ICD) inserted, prior to referral to our center, due to a mistaken diagnosis of pneumothorax. All patients underwent open surgical resection by posterolateral thoracotomy, out of which eight were performed in the neonatal period. The average duration of the postoperative ICD drainage was 3.5 days (1-11 days) and the average duration of hospital stay was five days (4-11 days). The final histological diagnosis was CCAM in 19 (48%), CLE in 11 (28%), bronchogenic cyst in five (12%), and pulmonary sequestration in five (12%). There were no hybrid lesions. Out of 40 patients, 21 (52%) had lesions on the right side and 19 (48%) on the left side, and four patients had multilobar involvement. Two CCAMs involved all the lobes, one right CCAM involved both upper and middle lobes, and one right CLE involved both middle and lower lobes [Figure 1]. Thirty patients underwent lobectomies, two underwent pneumonectomies, and eight underwent excisions for a bronchogenic cyst and for extralobar pulmonary sequestrations [Table 1]. Seven patients required postoperative ventilation with a median duration of 48 hours.

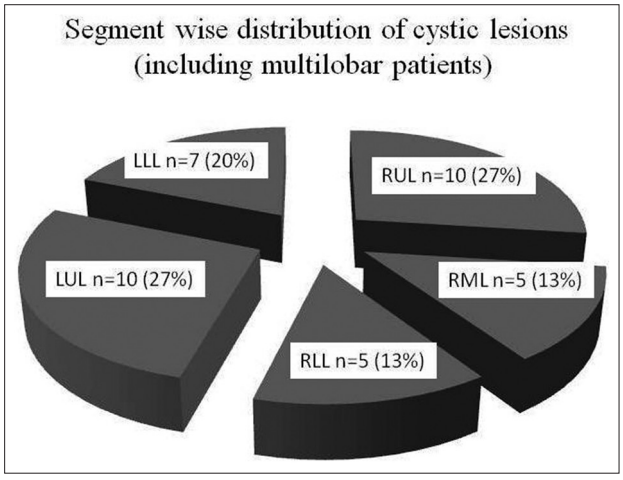

Figure 1: Lobar distribution of the cystic lesions (including four patients with multilobar involvement)

Complications included persistent non-expansion of the residual lobe requiring bronchoscopy and suction in two patients, pneumothorax necessitating re-ICD reinsertion in one patient, persistent hyperinflation necessitating a redo thoracotomy, and excision of a small foregut duplication cyst in one patient (this patient had been operated for a CLE), persistent bronchopleural fistula requiring redo thoracotomy and repair in one patient. There were no wound infections or mortality. The median period of follow up was 36 months and all patients had been asymptomatic.

DISCUSSION

Antenatal diagnosis of cystic lung lesions is the norm in the western setup, leading to prenatal counseling and planned delivery.[4] Only three patients in our series were diagnosed antenatally. The antenatally diagnosed lesions need to be evaluated postnatally with a CT scan, as the CXR may appear normal in spite of a pathological lesion.[8] The reported sensitivity of CXR as a test for the presence of CCAM is about 61%, while it is 100% for a CT scan.[9] In our series the CXR picked up the lesions in only 18 patients (45%), whereas, the CT scan picked up all the chest lesions (100%). Occasionally, a CT or magnetic resonance (MR) angiography may be required for better delineation of the vascular supply in case of pulmonary sequestration.[10]

Symptomatically, neonates with cystic lesions tend to manifest with respiratory distress while the older children present with recurrent infections.[4] Out of the 23 children with respiratory distress at presentation, 21 (92%) were neonates, and out of the 27 patients with recurrent respiratory infections, only 10 (37%) were neonates. Twelve (30%) patients had prior ICD insertion, due to mistaken diagnosis of pneumothorax. Likewise, eight (20%) patients had been misdiagnosed as tuberculosis and had received complete antitubercular therapy before referral to our center. Antenatal diagnosis, coupled with an awareness of congenital cystic lesions of the lung, could have prevented this wrong treatment.

Table 1: Surgical procedures performed on the various cystic lesions

	n	RUL	RML	RLL	LUL	LLL	*P*	Others
CCAM	19	5	2	4	1	5	2	
CLE	11	3	2		7			
PS	5		1			1		3
BC	5							5

n: Number of patients, RUL: Right upper lobectomy, RML: Right middle lobectomy, RLL: Right lower lobectomy, LUL: Left upper lobectomy, LLL: Left lower lobectomy, P: Pneumonectomy, CCAM: Congenital cystic adenomatoid malformation, CLE: Congenital lobar emphysema, PS: Pulmonary sequestration, BC: Bronchogenic cyst

Surgical resection is warranted for symptomatic lesions, but treatment recommendations for asymptomatic lesions are still unclear. The main arguments for surgical resection in asymptomatic lesions are the potential risk of recurrent infections, pneumothorax, and even malignant transformation.[7,11] About 4% of the pulmonary tumors are associated with congenital cystic malformations.[12] The tumors that develop are, sarcomas, pleuropulmonary blastoma, bronchogenic carcinoma, and mesenchymoma. Ozcan *et al*. in a case report and literature review reported 33 cases of primary pulmonary rhabdomyosarcomas, 15 of which arose in a pre-existing pulmonary cystic malformation.[13] We did not encounter any malignant degeneration in our series. Adzick *et al*. in their large series of 105 asymptomatic patients demonstrated that surgical resection is safe with no mortality and minimal morbidity.[14] Davenport *et al*., in their review and meta-analysis of the postnatal management of cystic lesions, concluded that although the risk of asymptomatic cases developing symptoms is small (3%), elective surgery in asymptomatic cases has significantly reduced postoperative complications compared to emergency surgery in these cases.[15] Also they mentioned that segmentectomy was associated with more residual disease (15%) compared to cases in which lobectomies were carried out (0%). All our patients underwent lobectomies and none have shown evidence of residual disease on follow up.

Minimally invasive surgery has emerged as the standard of care for many pediatric surgical conditions.[16] The stated advantages are: Better postoperative pain control, shorter hospital stay, and improved cosmesis, compared to standard thoracotomy. Thoracoscopic resection of CCAM has been reported to have a longer operative time, shorter hospital stay, and potentially reduced complications, with no additional costs, with the main risk factor for conversion to thoracotomy being a prior history of pneumonia.[17] All our patients underwent posterolateral thoracotomy and resection, with an average hospital stay of five days. Probably with the increasing use of video-assisted thoracoscopic surgery (VATS), thoracoscopic resection of cystic lesions may well become the standard of care in future.

CONCLUSION

Although the treatment of cystic lung lesions is quite straightforward, lesions that are not diagnosed antenatally are quite a diagnostic challenge when they become symptomatic, especially in our country (India), where pediatric surgical facilities are not readily available. Increased awareness of this uncommon entity coupled with a wider antenatal ultrasonography (USG) cover for pregnant mothers would result in prompt referrals to a pediatric surgical center, thereby, obviating unnecessary prolonged and needless medical management like ICD insertion and antitubercular therapy.

REFERENCES

1. Laberge JM, Flageole H, Pugash D, Khalife S, Blair G, Filiatrault D, *et al*. Outcome of the prenatally diagnosed congenital cystic adenomatoid lung malformation: A Canadian experience. Fetal Diagn Ther 2001;16:178-86.
2. Crombleholme TM, Coleman B, Hedrick H, Liechty K, Howell L, Flake AW, *et al*. Cystic adenomatoid malformation volume ratio predicts outcome in prenatally diagnosed cystic adenomatoid malformation of the lung. J Pediatr Surg 2002;37:331-8.
3. Harrison MR, Adzick NS, Jennings RW, Duncan BW, Rosen MA, Filly RA, *et al*. Antenatal intervention for congenital cystic adenomatoid malformation. Lancet 1990;336:965-7.
4. Davenport M, Warne SA, Cacciaguerra S, Patel S, Greenough A, Nicolaides K. Current outcome of antenally diagnosed cystic lung disease. J Pediatr Surg 2004;39:549-56.
5. vanLeeuwen K, Teitelbaum DH, Hirschl RB, Austin E, Adelman SH, Polley TZ, *et al*. Prenatal diagnosis of congenital cystic adenomatoid malformation and its postnatal presentation, surgical indications, and natural history. J PediatrSurg 1999;34:794-9.
6. Calvert JK, Boyd PA, Chamberlain PC, Syed S, Lakhoo K. Outcome of antenatally suspected congenital cystic adenomatoid malformation of the lung: 10 years' experience 1991-2001. Arch Dis Child Fetal Neonatal Ed 2006;91:F26-8.
7. Aziz D, Langer JC, Tuuha SE, Ryan G, Ein SH, Kim PC. Perinatally diagnosed asymptomatic congenital cystic adenomatoid malformation: To resect or not. J Pediatr Surg 2004;39:329-34.
8. Shanti CM, KleinMD.Cystic lung disease. Semin Pediatr Surg 2008;17:2-8.
9. Calvert JK, Lakhoo K. Antenatally suspected congenital cystic adenomatoid malformation of the lung: Postnatal investigation and timing of surgery. J PediatrSurg2007;42:411-4.
10. Doyle AJ. Demonstration of blood supply to pulmonary sequestration by MR angiography. AJR Am J Roentgenol 1992;158:989-90.
11. Shanmugam G, MacArthur K, Pollock JC. Congenital lung malformations--antenatal and postnatal evaluation and management. Eur J CardiothoracSurg 2005;27:45-52.
12. Hartman GE, Shochat SJ. Primary pulmonary neoplasms of childhood: A review. Ann ThoracSurg 1983;36:108-19.
13. Ozcan C, Celik A, Ural Z, Veral A, Kandiloğlu G, Balik E. Primary pulmonary rhabdomyosarcoma arising within cystic adenomatoid malformation: A case report and review of the literature. J Pediatr Surg 2001;36:1062-5.
14. Tsai AY, Liechty KW, Hedrick HL, Bebbington M, Wilson RD, Johnson MP, *et al*.Outcomes after postnatal resection of prenatally diagnosed asymptomatic cystic lung lesions. J Pediatr Surg2008;43:513-7.
15. Stanton M, Njere I, Ade-Ajayi N, Patel S, Davenport M. Systematic review and meta-analysis of the postnatal management of congenital cystic lung lesions. J Pediatr Surg2009;44:1027-33.
16. Georgeson KE, Robertson DJ. Minimally invasive surgery in the neonate: Review of current evidence. Semin Perinatol 2004;28:212-20.
17. Vu LT, Farmer DL, Nobuhara KK, Miniati D, Lee H. Thoracoscopic versus open resection for congenital cystic adenomatoid malformations of the lung. J Pediatr Surg2008;43:35-9.

How to cite this article: Raman VS, Agarwala S, Bhatnagar V, Panda SS, Gupta AK. Congenital cystic lesions of the lungs: The perils of misdiagnosis - A single-center experience. Lung India 2015;32:116-8.

Source of Support: Nil, **Conflict of Interest:** None declared.