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Original Research Article

Unusual presentations of late onset diaphragmatic hernia: a six year study

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ABSTRACT

Background: Late-presenting congenital diaphragmatic hernia (CDH) is a rare subset of CDH, most of the information is derived from small series or case reports. The aim of this study is to document the varied presentations of late-presenting CDH in a tertiary hospital.
Methods: Information about late-presenting CDH (diagnosed at later than 30 days of age) was identified from the data collected by a 5 year reterospective study and 1 year prospective study.
Results: Twenty five cases were studied over a period of 6 years with 5 years of reterospective study and 1 year of prospective study. Three cases had a morgagni hernia (2 right-sided, 1 central, 2 males and 1 female). There were 16 males (64%) and 9 females (36%). Major associated anomalies were identified in 6 cases (24%). Cardiac anomalies included ventricular septal defect (n = 4), atrial septal defect (n = 2), gastroesophageal reflux was found in 3 cases, 1 of which required surgery. Presenting symptoms were respiratory in 8 (32%), gastrointestinal in 13 (52%), both in 4 (16%). The hernia was left-sided in 17 (68%), right-sided in 5 (20%), and central 3 (12%). Patients with gastrointestinal symptoms invariably had left-sided hernias (n = 10), whereas patients with respiratory symptoms (n = 8) seemed equally likely to have right- or left-sided lesions. A primary repair without patch was done in all cases with 100% survival.
Conclusions: Presenting symptoms of late-onset CDH can be respiratory or gastrointestinal, but presentation with gastrointestinal problems was more common in left-sided hernias, whereas respiratory symptoms were seen with equal preponderance in left and right-sided lesions. The prognosis is excellent once the correct diagnosis is made.

Keywords: Congenital diaphragmatic hernia, Diagnosis, Late presentation

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a well-known cause of severe respiratory distress in the newborn and is associated with a high mortality rate. There is a milder form of CDH that does not present with symptoms until later in life. The presenting symptoms, diagnosis, management, and complications of late-presenting CDH differ considerably from those of neonatal CDH. It is generally assumed that acquired herniation of the abdominal viscera occurs through a congenital diaphragmatic defect that had been occluded by spleen or liver.[1,2] Previously normal chest radiographs and findings of the defect covered by peritoneum without any sign of inflammation support this assumption.[2,3] The outcome is generally favorable, but misdiagnosis can result in significant morbidity and mortality. Information about late-presenting CDH is derived from many case reports or small series except a recent collective review by Baglaj et al [1,3-9]. The largest series in the literature is a report of 26 cases by Berman et al, followed by 2 series of 22 cases.[3,7,8] In this study, we report a review of the

demographics, presenting symptoms, and outcomes of late-presenting CDH in a tertiary care centre.

METHODS

Records of 25 paediatric patients with CDH were recorded from data collection centre between 2011 to 2017. The database was screened for patients with late-presenting CDH. All patients presenting later than 30 days of age were included. The patient demographics, treatment, and outcomes were reviewed retrospectively. Out of twenty five patients, three patients had atypical presentation.

Case 1

One female child had right side CDH with caecal and ascending colon volvulus with ascending colon perforation and was diagnosed late at age of 12 years with features of faecopneumothorax, intestinal obstruction and septicaemia. Intraoperatively pt. had volvulus of right colon with perforation in ascending colon and generalized faecal contamination of right thorax and abdomen. Patient underwent Exploratory laparotomy with repair of diaphragm, resection anastomosis of perforated colonic part.

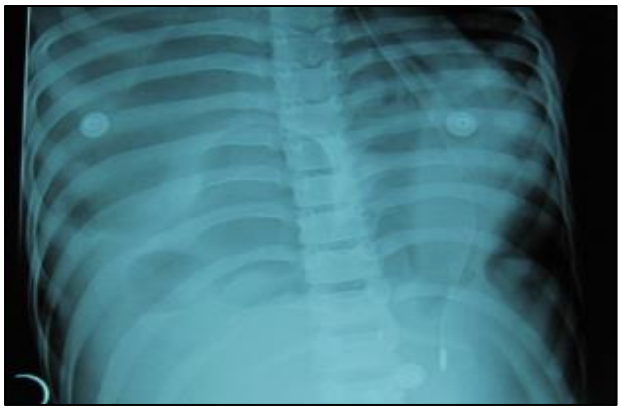

Figure 1: Chest x-ray.

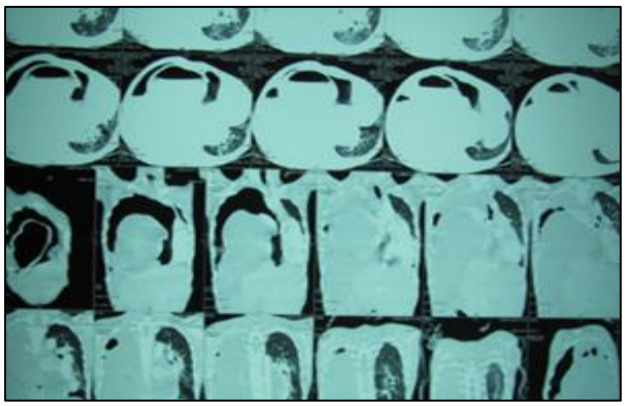

Figure 2: CT Chest pic of right CDH with perforation.

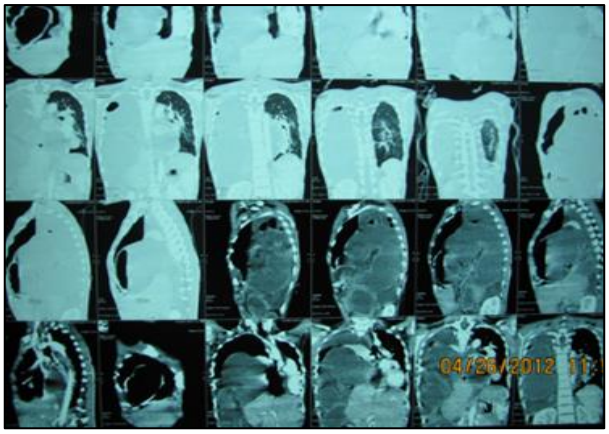

Figure 3: CT chest.

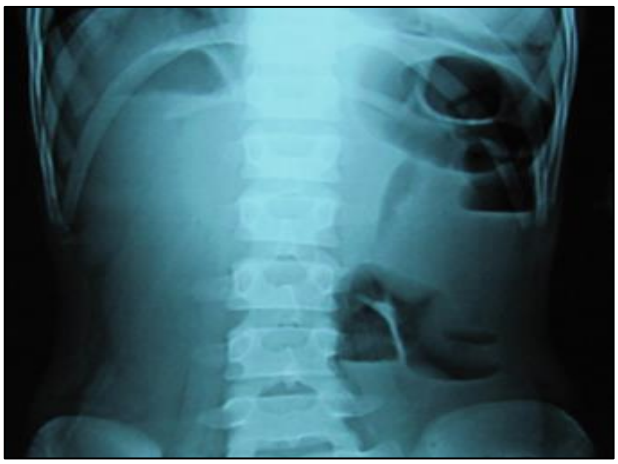

Figure 4: Abdominal x-ray.

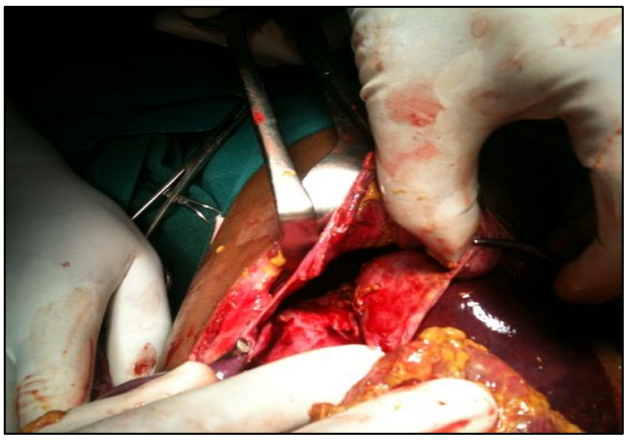

Figure 5: Intraop pic of right CDH.

Case 2

Another child 6yrs old presented with left fecopneumothorax along with the usual x-ray findings of left CDH. Patients underwent exploratory laparotomy with repair of diaphragm, primary repair of perforated colonic part.

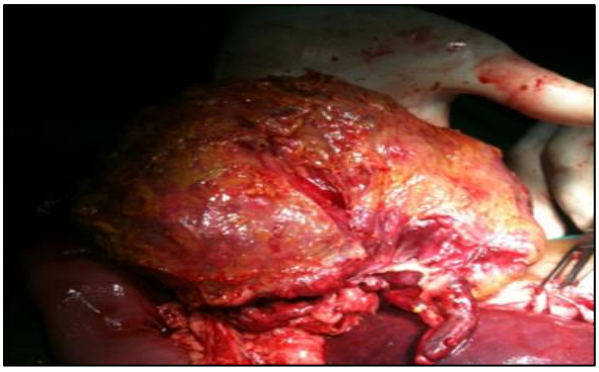

Figure 5: Right colon pulled out of chest.

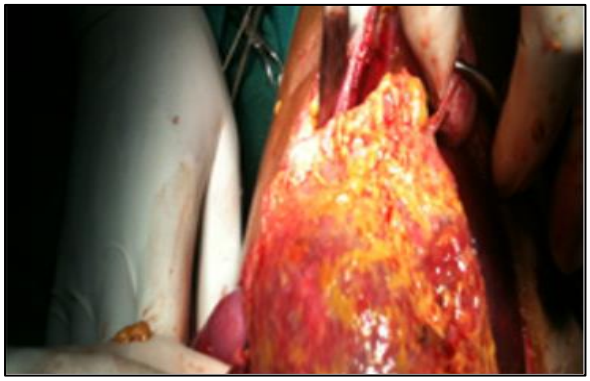

Figure 6: Perforated right colon being inspected.

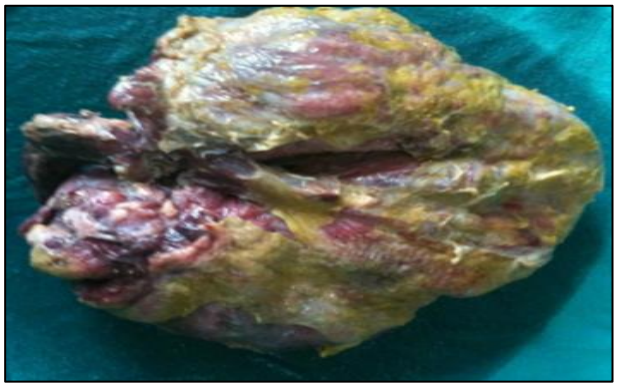

Figure 7: Excised gangrenous right colonic specimen.

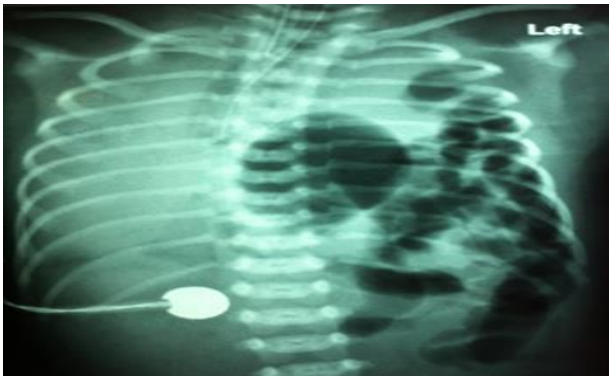

Figure 8: Chest X-ray of left CDH.

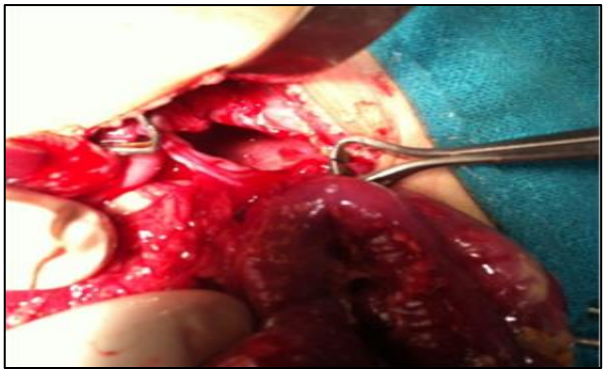

Figure 9: Left colon pulled out of left chest

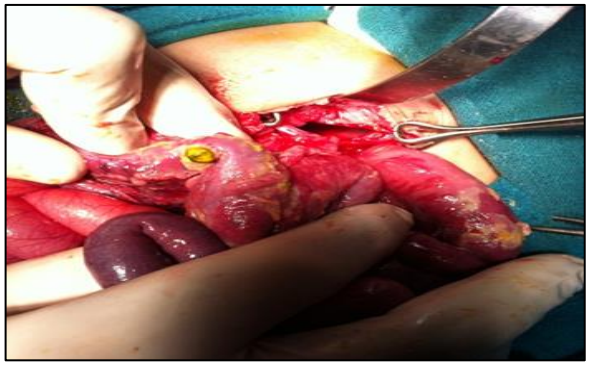

Figure 10: Left colonic perforation - part which herniated into chest.

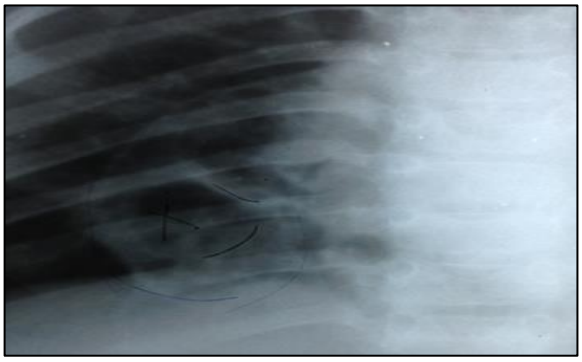

Figure 11: Xray chest of Morgagni hernia.

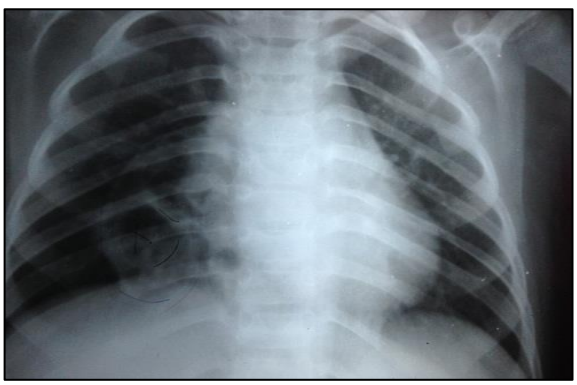

Figure 12: X-ray chest of Morgagni hernia.

Case 3

Another pt. of morgagni hernia, who was a 8 year old male child presented with features of vomiting, not gaining weight, ocassional cough. Patient was diagnosed on chest x-ray and hence repair done through transabdominal route.

RESULTS

Twenty five cases were studied over a period of 6 yrs with 5 years of reterospective study and 1 year of prospective study. Three cases had a morgagni hernia (2 right-sided, 1 central, 2 males and 1 female). There were 16 males (64%) and 9 females (36%). There were 11 males and 6 females with a left CDH and 3 males and 2 females with a right CDH. Males dominated in both groups with a male-to-female ratio of 1.8 in the left CDH and 1.5 in the right CDH. The mean age at diagnosis was 372 days (n = 25, range - 32 days to 15 years). The age at diagnosis was not different between the left CDH (390±1078 days, n = 17) and the right CDH (346 ± 704 days, n = 5). The hernia was left-sided in 17 (68%), right-sided in 5 (20%), central in 3.

Table 1: Presenting symptoms.

Symptoms	n	Details
Respiratory symptoms	8	
Acute	4	URI/pneumonia, 2; respiratory distress, 1; cough, 1;
Chronic	4	Tachypnea, 1; chronic respiratory distress, 2; silent aspiration, 1
Gastrointestinal symptoms	13	
Acute	10	Vomiting ± abdominal pain, 7; tarry stool + abdominal pain, 1; abdominal pain 2
Chronic	3	FTT, 2; constipation, 1
Both	4	
Acute	2	pneumonia +/or respiratory distress with vomiting +/or abdominal pain
Chronic	2	FTT + recurrent URI, poor feeding + cough, SOB + abdominal pain

URI, upper respiratory tract infection; SOB, shortness of breath; FTT, failure to thrive.

Major associated anomalies were identified in 6 cases (24%). Cardiac anomalies included ventricular septal defect (n = 4), atrial septal defect (n = 2), gastroesophageal reflux was found in 3 cases, 1 of which required surgery.

Presenting symptoms could be identified in 25 cases (Table 2). They were classified as respiratory (upper respiratory tract infection, pneumonia, respiratory distress, cough wheezing, etc) in 8 (32%), gastrointestinal (vomiting, abdominal pain, failure to thrive, constipation, etc) in 13 (52%), both in 4 (16%).

Table 2: Presenting symptoms in relation with side of the hernia and age at diagnosis.

Symptoms	Right	Left	Age at diagnosis
Respiratory	4	4	447±768
Gastrointestinal	3	10	625±1457
Both	0	4	1006±2178

Side of the hernia and the age at diagnosis as they relate to the presenting symptoms. Of 13 patients with gastrointestinal symptoms, 10 had left-sided hernias whereas patients with respiratory symptoms seemed equally likely to have right- or left-sided lesions ($P < .05$ by Fisher's Exact probability test). The age at diagnosis was not different between the cases with respiratory and gastrointestinal symptoms (Table 3).

A primary repair without patch was done in all 25 cases with 100% survival; only complication being Incisional hernia in a 12 year old female with right colonic volvulus with perforation which was repaired twice. The surgical approach was subcostal in all the cases. A hernia sac was present in 6 cases (24%).

DISCUSSION

In this combined prospective and retrospective aAnalysis of 25 late-presenting CDH, the side distribution, male-to-female ratio and symptomatology was studied. The rare presence of a hernia sac (24%) and a moderate incidence of major associated anomalies (24%) are also the features shared with neonatal variant. These findings are compatible with the assumption that the defect of late-presenting CDH is congenital.[2]

Several features of late-presenting CDH were noted in previously published studies. They include a high incidence of right-sided defects.[1,4,-6] Presentation primarily with respiratory symptoms in the younger patients and gastrointestinal symptoms in the older ones.[1,5,7] Younger age of patients with right CDH as compared with those with left-sided defects.[5,9] We could not confirm any of these features in our study. There was no association between age and clinical presentation nor age and side of the defect.

Late-presenting CDH is characterized by a variety of clinical symptoms. The patients may present with a wide

range of acute or chronic respiratory or gastrointestinal symptoms or may be completely asymptomatic. We found that virtually greater proportion of right CDH cases presented with respiratory symptoms, whereas gastrointestinal symptoms were seen predominantly in left CDH cases. This is in agreement with the review of Baglaj et al, in which two thirds of infants with right CDH had respiratory symptoms alone. We speculate that partial liver herniation, which is a common finding in the right CDH, may block the further herniation of hollow viscera, preventing the development of gastrointestinal symptoms.[9]

Although pneumonia is frequently the initial incorrect diagnosis in these children, it is usually not associated with severe morbidity. In contrast, an incorrect diagnosis of tension pneumothorax or pleural effusion is associated with inappropriate chest tube insertion and subsequent gastrointestinal perforation or bleeding from the thoracic liver.[3,10,11,12] Nasogastric tube placement confirms herniation of the stomach into the left side of the chest and is the initial treatment of choice when a tension gastrothorax is identified. The possibility of CDH should be considered in unusual cases of pneumothorax, especially in the absence of trauma, so that unnecessary procedures like chest tube drainage can be avoided.

We conclude that presenting symptoms of late-onset CDH can be respiratory or gastrointestinal, but presentation with gastrointestinal problems was more common in left-sided hernias, whereas respiratory symptoms predominated in right-sided lesions. The prognosis is excellent once the correct diagnosis is made. A routine chest radiograph should be included in the evaluation of the child with any unexplained acute or chronic respiratory or gastrointestinal problems so as one doesn't put a chest tube accidentally mistaking it for pneumothorax.

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Conflict of interest: None declared
Ethical approval: The study was approved by the institutional ethics committee

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