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Case Report

Congenital esophageal diverticulum in a very low birth weight infant: case report and review of literature

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ABSTRACT

Background. A diverticulum is an outpouching of a tubular organ that is classified as congenital and acquired according to the involved layers of the gastrointestinal wall. Congenital true diverticulum has been very rarely seen in neonatal period and it is very difficult to diagnose it especially in premature infants.

Case. A male infant was born with birth weight of 1000 g at 28th gestational week, was hospitalized for prematurity and respiratory distress. During follow up intermittent CO_2 retention was observed in blood gases. On the 17th day of hospitalization, esophageal dilatation was detected on X-ray and barium swallowed esophagram showed a saccular pouch on the distal esophagus. The patient was operated on 26th day of life and pathological specimen revealed true diverticulum of esophagus. The patient died due to respiratory failure and septic shock during hospitalization.

Conclusion. To the best of our knowledge this case is the smallest and youngest preterm infant diagnosed with congenital esophageal diverticulum. Prolonged and intermittent CO_2 retention such as in our case can be an atypical symptom of congenital diverticulum and it should be suspected in the differential diagnosis. Congenital esophageal diverticulum may be also seen in extremely preterm infants and can present with unusual symptoms.

Key words: congenital, esophageal diverticulum, newborn, premature.

A diverticulum is an outpouching of a tubular organ. Esophageal diverticulum is classified according to the location such as upper (pharyngoesophageal, Killion-Jamiesson or Zenker), middle or lower (epiphrenic). Besides anatomical location, esophageal diverticulum has also been described as pulsion or traction type according to the pathogenesis. Traction diverticula are true diverticula seen in mid-esophagus, associated with inflammation in mediastinum leading an external pressure on the esophagus. Pulsion diverticula are also classified into two groups; one is Zenker diverticule and the other is epiphrenic that both occur due to increased pressure on the upper and lower esophageal sphincter. Esophageal diverticula are also classified as; congenital (containing all layers of intestinal wall) and acquired (occurring in mucosal and submucosal layers via a defect in the muscular wall as herniation) like Zenker diverticulum.[1,2]

Congenital true diverticulum has very rarely been reported in newborns. Brintall and Kredelbagh[3] described two newborns with posterior hypopharyngeal congenital diverticule. None of them survived. These diverticula are usually small and asymptomatic; therefore, it is really difficult to diagnose especially in newborns. To the best of our knowledge here we reported the smallest and youngest infant diagnosed with congenital diverticulum.

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Case Report

A male infant was born by cesarean section at 28 weeks' gestation to a 22-year-old mother with Apgar 5 and 7 at 1 and 5 minutes. The birth weight was 1000 g (10-50th centile), birth length was 36 cm (50th centile) and head circumference was 25 cm (10-50th centile). The patient was hospitalized and monitored. Physical examination revealed respiratory distress and prematurity findings. No other congenital anomalies were observed with clinical and radiological findings. Early rescue surfactant was administered. He was given ampicillin and gentamicin therapies for sepsis. He could not be extubated during the first days of life due to the intermittent CO_2 retention that was not was compatible with the chest X-ray findings. Although chest radiograms did not reveal reticulogranular image and ventilator parameters were upgraded, there was no dramatical change in CO_2 levels during this period. This condition continued approximately once or twice a day. Ventilatory support status and blood gases changes are summarized in Table I. On the postnatal 17th day, a dilatation at esophagus was seen on chest radiogram (Fig. 1) and barium swallowed esophagram showed a saccular pouch on middle-distal esophagus (Fig. 2). Computed tomography of thorax illustrated a hypodense lesion with cystic characteristics with 30x14x15 mm in diameters forcing lung posteriorly in paraesophageal zone in the left hemithorax. After consultation with pediatric surgeons, esophagogastroduodenoscopy was performed in the operation room under general anesthesia and no evidence of diverticulum was detected. In addition to the persistence of unexplained intermittent CO_2 retention and persisting dilatation in the mid-esophageal region, the infant was operated on 26th postnatal day of life. After left thoracotomy at 5th intercostal area, grayish-like color mass with 3x1.5x 2 cm diameter was visualized between aorta and vertebra in posterior mediastinum left to distal esophagus. The mass was resected and no continuation or opening to any other part of esophagus or mediastinum was observed. The esophagus was primarily repaired and closed. Pathological evaluation of the mass revealed a true diverticulum containing all layers of esophagus. However, after the operation, the patient died on postnatal 34th day due to severe respiratory failure and septic shock. Informed consent was obtained from the parents.

Discussion

Herein, we described the smallest and youngest preterm infant diagnosed with congenital esophageal diverticulum. The clinical presentation of the case revealed unexplained intermittent respiratory acidosis and respiratory distress that resolved and reoccurred spontaneously. Most of the

Table I. Ventilatory support status (synchronized intermittent mandatory ventilation / continuous positive airway pressure / high frequency oscillation ventilation) and blood gases (pH / pCO_2) alterations of the patient before and after the interventions during the hospital stay.

Date of Blood Gases	pH	pCO_2 (mmHg)	Ventilatory support status
1st day (Before surfactant therapy)	7.22	64	SIMV mode
1st day (After surfactant therapy)	7.36	42	SIMV mode
3rd day	7.25	59	SIMV mode
7th day (Before extubation)	7.40	38	SIMV mode
7th day (After extubation)	7.21	92	Nasal CPAP mode
17th day	7.18	104	HFOV mode
26th day	7.26	59	SIMV mode
34th day	6.92	142	HFOV mode

pH: power of hydrogen, pCO_2: partial pressure of carbon dioxide, SIMV: synchronized intermittent mandatory ventilation, CPAP: continuous positive airway pressure, HFOV: high frequency oscillation ventilation.

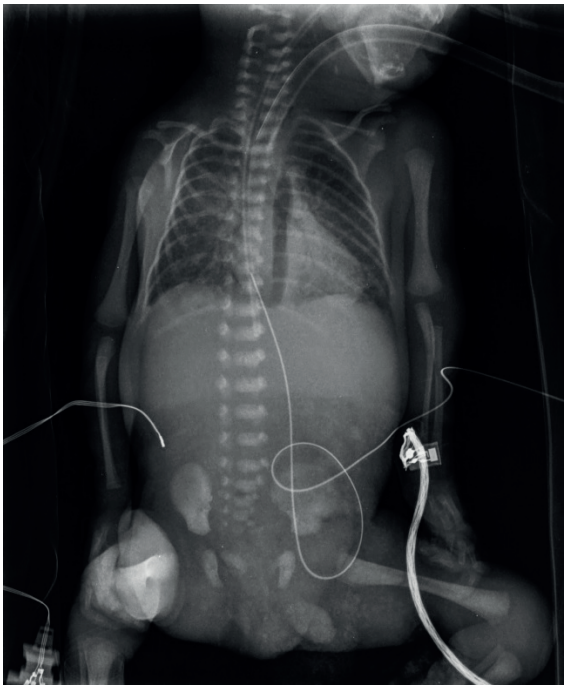

Fig. 1. Dilatation at esophagus seen on chest radiogram.

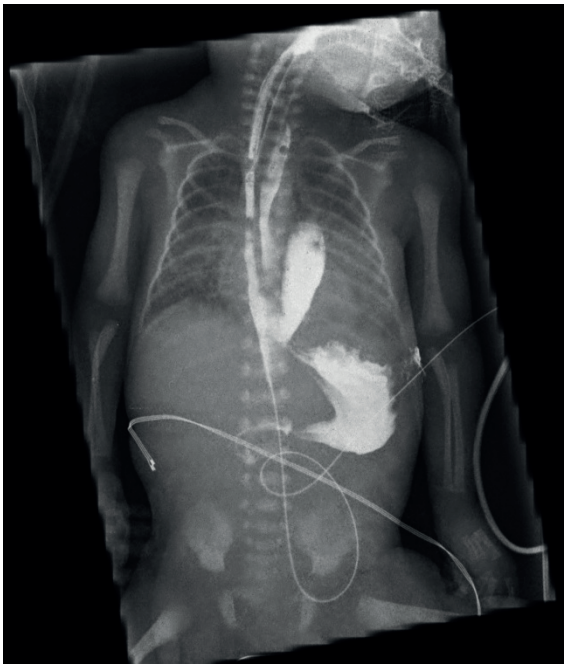

Fig. 2. Saccular pouch on middle-distal esophagus in barium swallowed esophagram.

esophageal diverticula are acquired and seen in middle-aged group, it has been very rarely been reported in infants.[4]

Holderman[5] first described a 5-day old infant misdiagnosed with congenital diverticulum in 1927, it was thought to be a class 3 tracheoesophageal fistula. Rush and Stingily[6] reported a newborn with supraclavicular mass causing respiratory obstruction. The autopsy revealed a large congenital cricopharyngeal diverticulum. Also, diverticulum in a newborn was reported by O'Bannon[7] on autopsy including a pouch anteriorly and inferiorly proximal to distal esophagus. In an extended review of literature, Poncher and Milles[8] could not find any examples of congenital esophagus diverticulum in 1933. In a more recent paper, an 8-day old term infant with stridor and poor feeding was diagnosed with upper esophageal diverticulum.[9] However, to our best of knowledge, no preterm infant had a diagnosis of esophageal diverticulum. Therefore, our case represents the smallest preterm infant diagnosed with congenital esophageal diverticulum in the literature.

Development of congenital diverticulum was suggested based on esophageal motor dysfunction by D'Abreu[10] in 1949 and it was manometrically confirmed by Cross et al.[11] in 1961. Kaye[12] reported esophageal motor dysfunction in 12 patients diagnosed with diverticule in 1974. Further theories supporting the development of diverticula were suggested by Ishigami et al.[13] in 1965. Our case had unexplained intermittent respiratory acidosis that did not response to ventilatory therapy and was not compatible with radiological findings. Therefore, motor dysfunction, impaired relaxation and delayed passage in esophagus might have caused increased pressure on the esophagus.[12] The weakness of the muscular layer of esophagus and pouch formation might have resulted in increased pressure against the lungs leading to worsening of pulmonary function. The blood gases might be found normal possibly when the size of the pouch and associated pressure against lungs decreased.

Clinical features are broad for esophageal diverticula. It can be asymptomatic but dysphagia is a common finding. When

symptoms occur, they are likely to be caused by associated underlying motility disorder. Other clinical features associated with esophageal diverticulum include feeding problems, weight loss, regurgitation, stridor, belching, bleeding, and cough. In a recent case report; esophageal diveticulum with bronchoesophageal fistula was diagnosed in case of unexplained cough or recurrent pneumonia.[14] In our case, intermittently increasing CO_2 levels were detected as the main and unusual symptom. We suggest that dilatation of esophagus and sac formation may have led to increased pressure on lungs. We also think that it resolved spontaneously when the pressure decreased.

According to adult American Gastroenterological Association guidelines, barium swallowed esophagogram is the gold standard diagnostic method.[15] On barium swallowed esophagogram, diverticulum will be illustrated as a distended, barium filled sac above the diaphragm. We showed the same similar findings in our patient. There was dilated, pouch-like formation seen in the mid-esophageal area. Endoscopy and bronchoscopy may also be useful for both confirming and assessing the degree of esophageal inflammation and obstruction. Carcinomas or other diseases should also be excluded by endoscopy. Although we performed an endoscopy, we could not diagnose the diverticulum. Endoscopy may be dangerous due to increased risk of perforation with misdirection of the scope into the diverticula and is not recommended in case of large diverticula because of incomplete emptying of pouch remnants.[9] In our case, thorax CT showed the diverticulum as a hypodense lesion with cystic characteristics, therefore we suggest to use of other diagnostic tools such as CT in suspected cases. This CT finding can be interrupted with esophageal duplication cyst. But approximately 90 percent of esophageal cysts do not communicate with the esophageal lumen.[16] In our case the lesion originated from the esophageal lumen. We could precisely differentiate this lesion from duplication cyst by pathological evaluation.

The optimal management of these cases are early and prompt surgery. Surgical approach for treatment of diverticulum is mainly myotomy, supported by various studies in which 80-100% of patients had good outcomes.[17] Left-sided thoracotomy is mostly preferred to visualize the esophagus. Our case underwent a left thoracotomy and after the resection of the sac, all esophageal layers were closed anatomically. Although the surgery was successful, the patient died due to problems of prematurity and neonatal sepsis. The prognosis varies according to several factors including the presentation, gestational age, and associated abnormalities. However, the prognosis may be poorer in preterm infants such as in our case. Therefore, we recommend prompt surgery in appropriate conditions after the diagnosis.

The strength of this case can be suggested as the youngest and smallest preterm infant in the literature to be diagnosed and operated on very promptly. Although symptoms were non-specific our findings may represent a clue for both neonatologists and pediatric surgeons. The weak point of this paper is that the problems may have been associated with prematurity and also there was no genetic data about this congenital anomaly.

Esophageal diverticula are frequently acquired and seen in the middle-aged group, it has very rarely been reported in infants and even rarer in premature infants. Clinicians should keep in mind and be aware of this condition in the case of prolonged intubation and extubation failure with prolonged CO_2 retention that could not be related to any other situation.

Herein, we reported an unusual case of a true congenital esophageal diverticulum in a very low birth weight premature infant. Although congenital esophageal diverticulum is rare in neonates, the clinical findings may vary from respiratory problems to feeding problems. The main mechanism responsible from variable symptoms may be increased esophageal pressure. In addition to barium graphics, other diagnostic tools such as endoscopy or thorax

CT should be performed in suspected cases. Prolonged and intermittent CO_2 retention such as in our case can be an atypical symptom of congenital diverticulum. In conclusion, we suggest that congenital esophageal diverticulum should be kept in mind in the differential diagnosis of infants with respiratory and feeding problems even in preterm babies.

REFERENCES

1. Costantini M, Zaninotto G, Rizzetto C, Narne S, Ancona E. Oesophageal diverticula. Best Pract Res Clin Gastroenterol 2004; 18: 3-17.
2. Carrere N, Pradere B. Esophageal diverticula. J Chir (Paris) 2004; 141: 85-92.
3. Brintnall ES, Kridelbaugh WW. Congenital diverticulum of the posterior hypopharynx simulating atresia of the esophagus. Ann Surg 1950; 131: 564-574.
4. Dokucu AI, Fabre M, Otcu S, et al. Zenker's diverticulum in childhood: a new case. Eur J Pediatr Surg 2000; 10: 191-193.
5. Holderman HH. Congenital atresia of the esophagus with esophageal diverticulum and tracheoesophageal fistula. Arch Surg 1927; 14: 917-920.
6. Rush LV, Stingily CR. Congenital diverticulum of the esophagus; case report. Southern Med J 1929; 22: 546.
7. O'Bannon RP. Congenital partial atresia of the esophagus associated with congenital diverticulum of the esophagus; report of a case. Radiology 1946; 47: 471-477.
8. Poncher HG, Milles G. Cysts and diverticula of intestinal origin. Am J Dis Child 1933; 45: 1064-1078.
9. Lindholm EB, Hansbourgh F, Upp JR Jr, Cilloniz R, Lopoo J. Congenital esophageal diverticulum - a case report and review of literature. J Pediatr Surg 2013; 48: 665-668.
10. D'Abreu AL. Diverticula of the oesophagus. Br J Radiol 1949; 22: 423-426.
11. Cross FS, Johnson GF, Gerein AN. Esophageal diverticula. Associated neuromuscular changes in the esophagus. Arch Surg 1961; 83: 525-533.
12. Kaye MD. Oesophageal motor dysfunction in patients with diverticula of the mid-thoracic oesophagus. Thorax 1974; 29: 666-672.
13. Ishigami K, Matsuo Y, Yamane M, Suzuki H. Surgical treatment of diverticula of the mid-thoracic esophagus, with special references to their pathogenesis. Nihon Geka Hokan 1965; 34: 509-523.
14. Wang ZM, Zhang SC, Teng X. Esophageal diverticulum serves as a unique cause of bronchoesophageal fistula in children: a case report. Medicine (Baltimore) 2017; 96: e9492.
15. American Gastroenterological Association medical position statement on management of oropharyngeal dysphagia. Gastroenterology 1999; 116: 452-454.
16. Wootton-Gorges SL, Eckel GM, Poulos ND, Kappler S, Milstein JM. Duplication of the cervical esophagus: a case report and review of the literature. Pediatr Radiol 2002; 32: 533-535.
17. Cook IJ, Kahrilas PJ. AGA technical review on management of oropharyngeal dysphagia. Gastroenterology 1999; 116: 455-478.