Paediatric Boerhaave's syndrome: a rare but crucial diagnosis

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Boerhaave's syndrome is typically seen in middle-aged male population. We report a case of a child who presented with sudden onset of left-sided oedema of the face and periorbital region. The child had a history of fever and throat pain, which started a day prior to the onset of swelling. On examination, the child had crepitus over the swelling on the left side of the face. Radiology showed pneumomediastinum and pneumothorax, and were treated with chest tube insertion. We present this case because of its rarity, unique presentation and need for prompt diagnosis, and discuss the literature.

Keywords:
Boerhaave's syndrome, oesophageal rupture, pneumothorax

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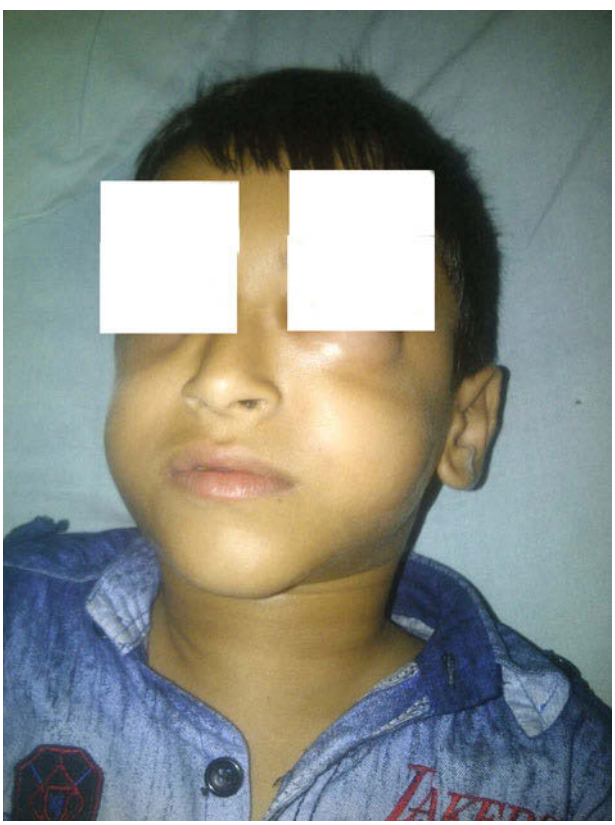
Introduction

Spontaneous rupture of the oesophagus occurs when intraoesophageal pressure increases against a closed cricopharyngeus during excessive retching or vomiting. This occurs commonly during binge eating and alcoholism [1]. Both these conditions are extremely rare in paediatric population, and hence the incidence of Boerhaave's syndrome is extremely low in children [2]. However, other conditions resulting in excessive vomiting can cause this condition in children. Although rare, if left untreated it can cause life-threatening complications such as mediastinitis and sepsis.

Case report

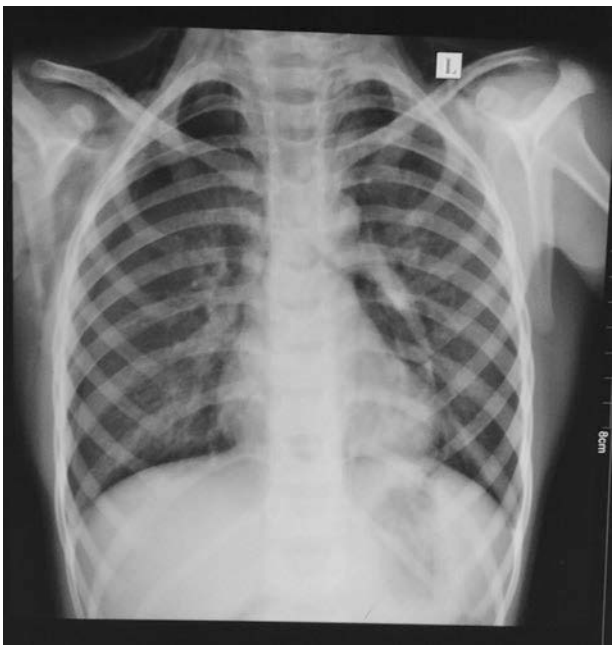
A 9-year-old boy presented to the emergency department complaining of acute onset painless swelling over the left side of his face. The child gave a history of fever and throat pain since 2 days, for which he was on antibiotics and analgesics. He had no breathing difficulty or difficulty in swallowing. There was no history of trauma. He gave a history of incomplete immunization. On examination, the child had left-sided facial and periorbital subcutaneous emphysema extending from the left upper eye lid and zygoma to the submandibular space (MEP_L_fig1 Fig. 1). The periorbital swelling prevented the child from opening his eyes. There was no tenderness or local rise of temperature over the facial swelling. His oral cavity was normal. Breath sounds were slightly reduced in all quadrants over the left side chest. He had no respiratory distress. He maintained normal saturation in room air. An ultrasonography of the neck was carried out, which showed no significant findings except air in the subcutaneous planes in the face and neck. A chest radiograph was carried out, which revealed left-sided pneumothorax (MEP_L_fig2 Fig. 2). A pulmonology consultation was taken, and a computerized tomography

Figure 1



Child with surgical emphysema of the left cheek.

Figure 2



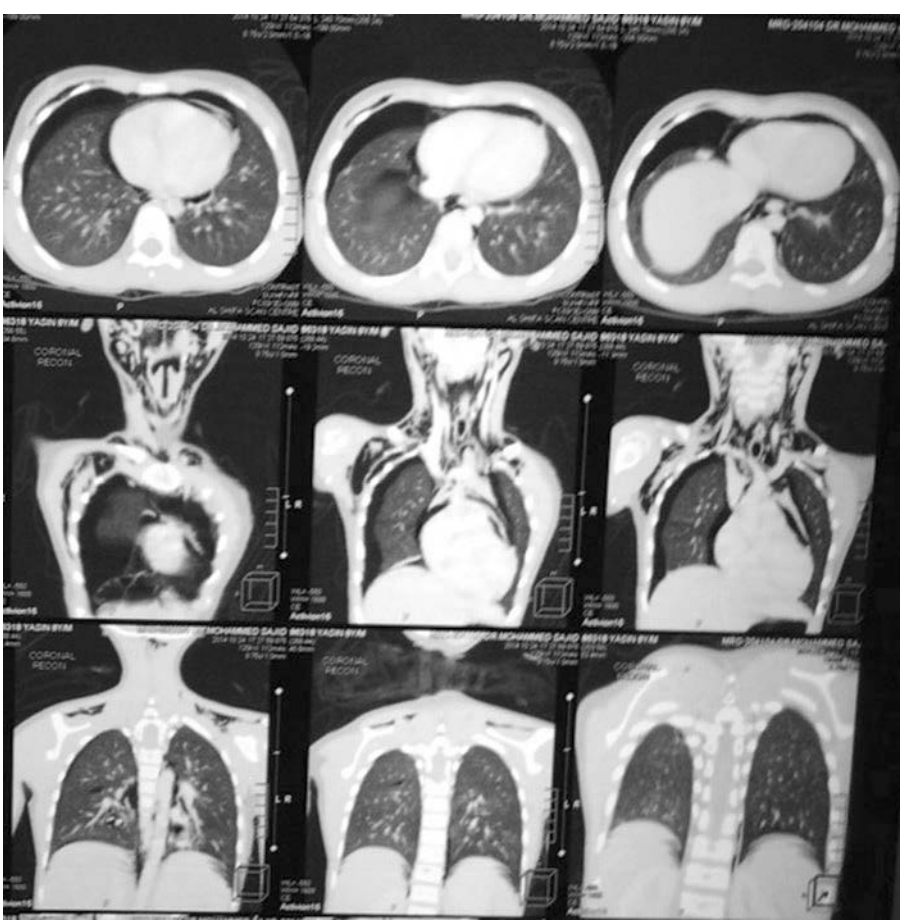
Chest radiograph showing pneumothorax.

of the neck and thorax was carried out, which revealed left-sided pneumothorax and pneumomediastinum (MEP_L_fig3 Fig. 3) and subcutaneous emphysema in the neck (MEP_L_fig4 Fig. 4). The reason for this presentation could not be explained. Hence, further detailed history was taken and the parents of the child conceded that this episode occurred immediately following a bout of retching and vomiting. Hence, the diagnosis of Boerhaave's syndrome was made. The child was given prophylactic intravenous antibiotics and intercostal chest tube was inserted as the emphysema was not resolving. He recovered within 3 days as the subcutaneous emphysema completely disappeared; chest radiograph showed the expansion of the lungs (MEP_L_fig5 Fig. 5). The intercostals chest drain was removed on the fourth day and the child was discharged on the sixth day. The patient was asymptomatic on follow-up.

Discussion

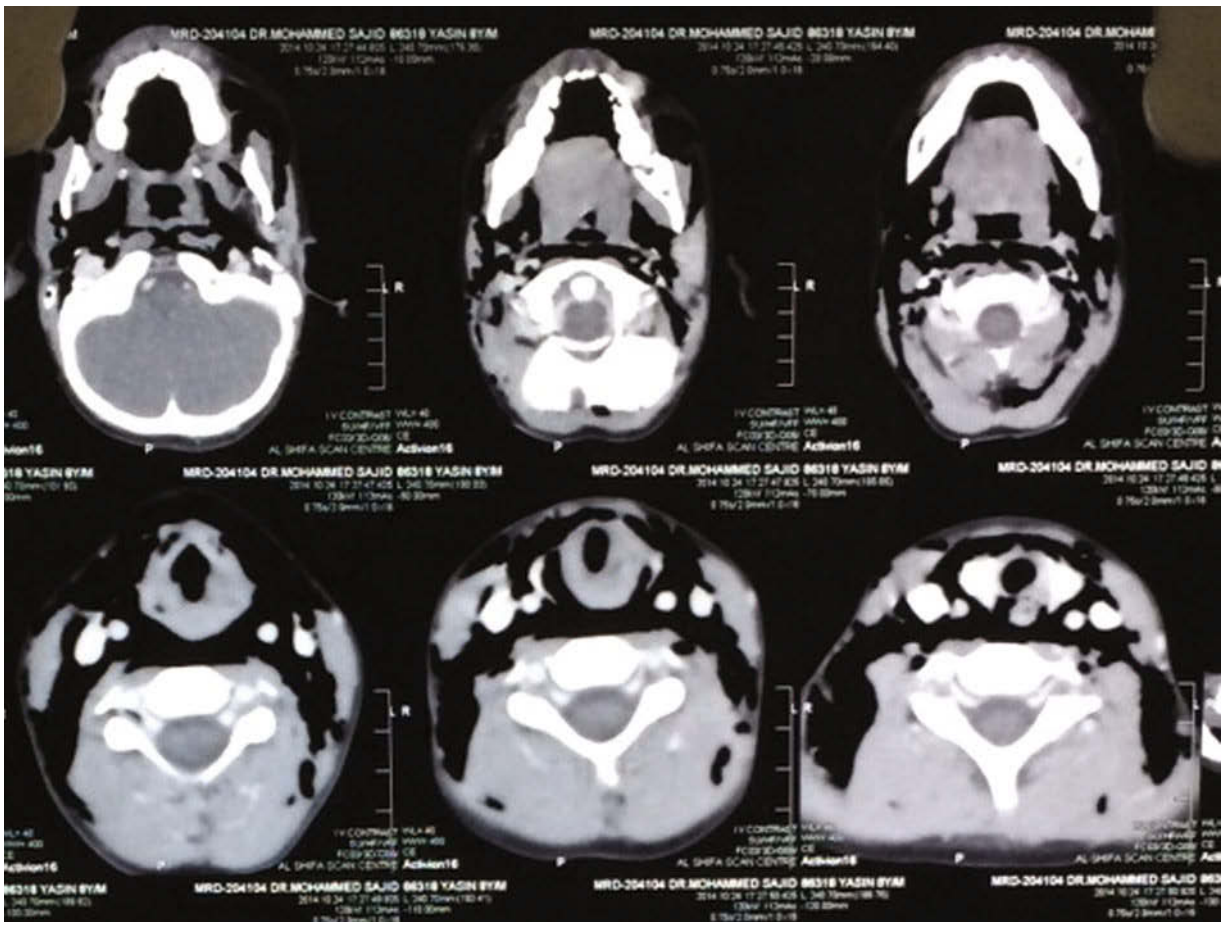
Boerhaave's syndrome was originally described by Herman Boerhaave in 1724 [3]. It commonly occurs

Figure 3



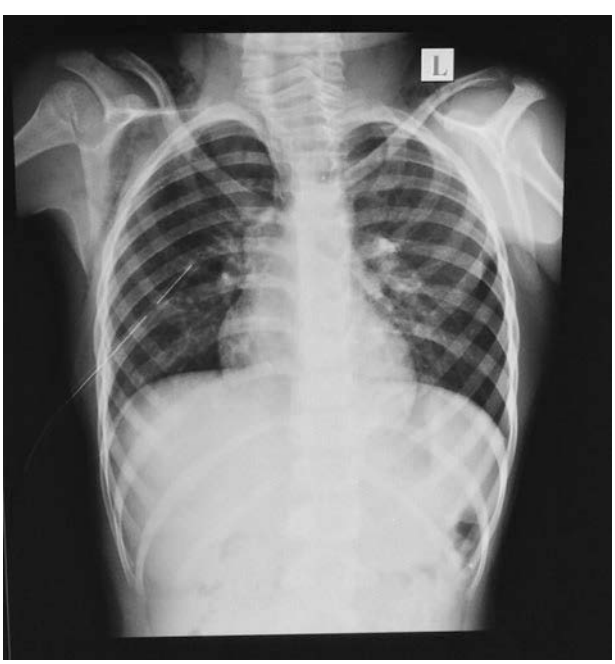
Computed tomography showing pneumothorax and pneumomediastinum.

Figure 4



Computed tomography of the neck showing subcutaneous emphysema.

Figure 5



Chest radiograph after intercostal chest tube placement.

in men aged 30–40 years, following an emesis usually either after excessive food intake or alcoholic binge. The male to female ratio is between 2: 1 and 5: 1. The basic pathomechanism behind Boerhaave's syndrome is the oesophageal barotrauma resulting from increased intraluminal pressure during excessive emesis, which is transmitted to the oesophagus against a closed cricopharyngeus [4]. The spontaneous nature of rupture following vomiting distinguishes it from other common traumatic and pathological oesophageal trauma. The most common site of rupture is the left posterolateral wall of the lower third of the oesophagus, followed by the subdiaphragmatic area. The patient presents with thoracoabdominal pain, vomiting and surgical emphysema, which is known as Mackler's triad [3]. However, history is not always reliable, especially in the paediatric age group where this condition is least expected. In our case, the child did not have abdominal pain. He had a single episode of vomiting that caused the clinical scenario. His vomiting might have been triggered due to the throat infection or due to the antibiotic that he was

taking for the same. A careful history, positive clinical findings and the demonstration of perioesophageal air tracks on radiological imaging causing pneumomediastinum should promptly diagnose Boerhaave's syndrome [5,6]. Mallory–Weiss tear is another condition that presents with haematemesis in the setting of alcoholism and food intake. In this condition, it is nontransmural tear of oesophagus, where as in Boerhaave's syndrome it is the transmural rupture. Other differential diagnosis of Boerhaave's syndrome include acute pancreatitis, aortic dissection, oesophageal rupture, myocardial infarction and peptic ulcer disease.

Biochemical investigations do not have any particular value in diagnosing this condition. However, some studies have suggested that the patient can present with polycythemia due to third space sequestration of the fluid [7]. Radiology is the mainstay in diagnosing Boerhaave's syndrome. In our case, a chest radiograph was taken only because the chest sounds were reduced on the affected side due to pneumothorax. Further, a computerized tomography of the thorax was carried out, which showed pneumomediastinum along with pneumothorax. This finding along with the crucial history of the episode occurring following emesis helped us to reach the diagnosis. Contrast oesophagography has been shown to have a sensitivity, consistently reported as 70–75%, and is not preferred over computerized tomography for diagnosis [5]. Undetected and untreated cases may further progress to pneumothorax, hydrothorax, pyothorax and sepsis. In extreme cases, the perforation may cause trauma or inflammation of the vagal trunks causing impaired gastric emptying and motility [8].

Fluid resuscitation and broad spectrum antibiotics are the mainstay of management. In majority of the cases the condition spontaneously resolves. Children tend to do well compared with their elder counterparts. Primary repair of the oesophageal tear is required only in complicated cases with pyomediastinum or sepsis. This can be done either immediately or in a delayed manner. Repair can be done either by opting for an open thoracotomy or an endoscopic method [9,10]. However, these situations have a high mortality rate. In the literature, Boerhaave's syndrome has a mortality of 30% in surgically treated cases and 100% in untreated cases [11]. In our case, the child had nonresolving subcutaneous emphysema and pneumothorax, which required intercostal drain placement. The child immediately responded and completely recovered within 3 days.

Paediatric Boerhaave's syndrome is extremely rare with only 29 documented paediatric cases [12]. This rarity causes the clinician to often neglect this possibility of diagnosis, especially when proper history is not available. Untreated cases may later present with above-mentioned complications.

Conclusion

Boerhaave's syndrome is an extreme rare diagnosis in a paediatric age group. However, if left undiagnosed and untreated, it may lead to fatal complications such as pyomediastinum and sepsis. Hence, the possibility of this diagnosis in the paediatric population should be made aware. A good clinical suspicion and prompt use of radiology is required to make the diagnosis and effectively treat such patients.

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Conflicts of interest

There is no conflict of interest.

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