A child with primary spontaneous mediastinal emphysema – A rare presentation of a self-limiting disease

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Abstract

Spontaneous mediastinal emphysema is a rare occurrence in children while it is relatively more common in adolescents. It leads to subcutaneous emphysema, pneumomediastinum and pneumopericardium. We report a nine year old boy, a non-asthmatic who presented with fever and incessant cough for three days. He developed sudden onset chest pain and crepitant swelling of the chest-wall, neck and face. He was observed in the intensive care unit. Cardiopulmonary monitoring was done. Antibiotic, nebulization and oxygen supplementation were given. Recovery was uneventful. We discuss the clinical features, aetiopathogenesis, diagnosis and management of this rare condition.

Keywords: Asthma; Children; Mediastinal emphysema; Spontaneous pneumomediastinum

Introduction

Spontaneous pneumomediastinum (SPM) or mediastinal emphysema refers to free air or gas in the mediastinum. Spontaneous pneumomediastinum is a benign condition. Spontaneous pneumomediastinum has a bimodal occurrence at ages below seven years and adolescents aged 13-17 yrs. Incidence of SPM varies between 1:800 to 1:42000 patients in the emergency departments. The actual incidence could be higher in the range of 1:14000 when routine screening for SPM is done in patients with chest pain and idiopathic dyspnea as reported by Yellin et al[1]. When it is present it is usually associated with asthma. SPM mainly affects adolescent males with tall, thin body habitus. SPM was first reported by Hamman in 1939. It can be primary when no cause can be identified or secondary where causes like cystic fibrosis or asthma, respiratory tract infection and bronchiolitis are present.

Case Report

An 11 year old boy was admitted in the ICU with spontaneous swelling of the neck and face for one day duration. He complained of mild breathing difficulty. There was no fever. He also complained of severe bouts of cough. He had no history of asthma or trauma. Foreign body ingestion/aspiration was ruled out. On examination of the child there was obvious swelling of the supra clavicular fossae, upper anterior chest wall, neck and facial puffiness extending till his lower eyelids (Fig. 1). The swelling was crepitant (subcutaneous emphysema). The child was afebrile. Pulse was 100/minute, BP was 90/60mmHg. His room air saturation was 85%. There was no cyanosis. Auscultation revealed normal vesicular breath sounds in all areas of the lung and Hamman's sign (mediastinal crunch) was positive. Chest radiograph postero-anterior (PA) view revealed extensive pockets of air over the lateral aspects of the chest, neck arm and supraclavicular fossae and Spinnaker-sail sign was present (Fig. 2). Both lung fields were clear. High resolution computed tomography (HRCT) of the chest showed moderate pneumomediastinum with pneumopericardium (Fig. 3 and 4). Air pockets were seen dissecting along the superficial & deep fascial planes of the neck, chest wall, upper limb, abdominal wall and face. Epidural pneumatosis was present. Total count was 12,000 cells/cu mm with mild shift to left. Other routine investigations were normal. He was treated in the intensive care unit with supplemental oxygen support, and he was managed conservatively. Injection Amoxicillin and Clavulanic acid was administered intravenously thrice daily for three days. Fiber-optic bronchoscopy and oesophagoscopy was done and found to be normal. The subcutaneous emphysema subsided spontaneously. Patient was shifted to the ward on the third day. He had no other complaints and he was discharged on the sixth day. On the follow-up visit at one month the child was completely free from symptoms. Pulmonary function test (PFT) was not done.

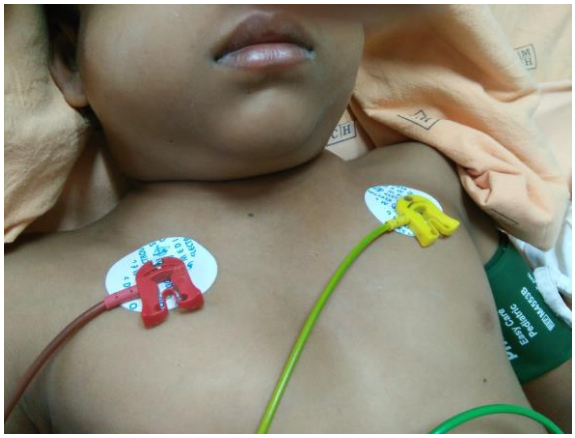

Fig. 1: Clinical picture of patient reveals swelling of the face, neck and upper chest wall due to subcutaneous emphysema

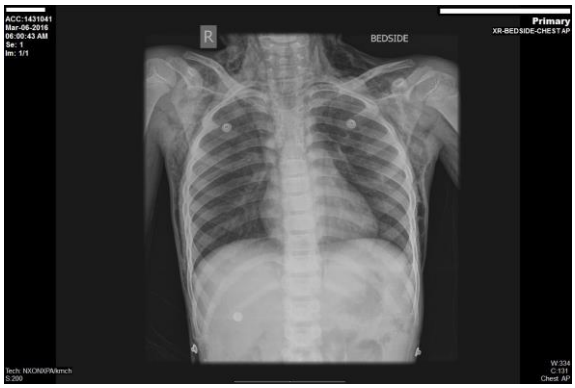

Fig. 2: Chest X-Ray shows air pockets in the subcutaneous tissues in the chest and neck and positive Spinnaker-sail sign

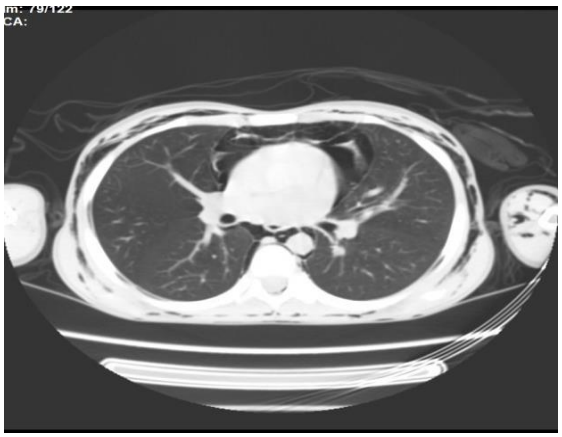

Fig. 3: HRCT reveals extensive pneumomediastinum

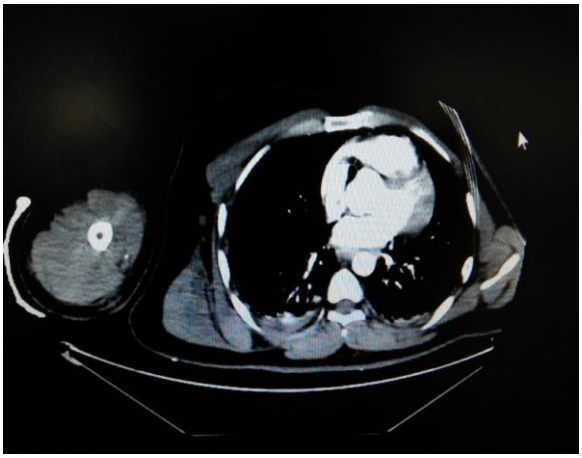

Fig. 4: HRCT shows pneumopericardium

Discussion

The pathophysiology behind SPM is alveolar rupture due to increased intra-alveolar pressure and the free air then tracks by dissection along the broncho-vascular sheath and reaches the mediastinum. This is called the 'Maclin effect'. The free air can remain within the mediastinum or can extend to the neck by through the communication between the mediastinum and the sub-mandibular space, the retro-pharyngeal space and vascular sheath of the neck as reported Zylak et al[2]. Retroperitoneal communication occurs through the sternocostal attachment to the diaphragm, peri-aortic and peri esophageal planes. Air tracks through these tissue planes resulting in pneumopericardium, pneumothorax, pneumoperitoneum and pneumo-retroperitoneum[3]. It can also enter into the neural foramina and spinal canal causing spontaneous pneumorrachis.

Patient usually are tall with thin habitus and present with dyspnea and chest pain[1]. Extensive subcutaneous emphysema is the hallmark clinical finding in patients with SPM. Other symptoms include fever, dysphonia, and throat and jaw pain. Chia-Ying Lee et al. in his study of eighteen patients discussed the etiology of SPM. They found trigger factors in 50% of children. Most common causes where infection (bronchopneumonia, bronchiolitis) and diabetic ketoacidosis in one patient. In their study nine boys developed idiopathic SPM after strenuous physical activity[4]. Stack and Caputo reported an incidence of 0.3% of 12,000 asthmatic patients while Egglestan et al. reported an incidence of 5% for SPM[5,6]. Differential diagnosis of SPM include esophageal rupture and foreign body aspiration[7]. Management of patients with SPM includes ambulatory monitoring, analgesics for chest pain, supplemental oxygen for hypoxia and broncho-dilators for wheezing. Pre-disposing factors if any like vomiting, strenuous exercise and activities that induce forced expiration should be avoided. Pre-disposing disease should be treated appropriately. PFT can be done on follow up visit to detect the presence of

hyper-active airway.

SPM has also been reported from India by a few studies in children and adult with H1N1 infection[8,9]. SPM is a rare complication of H1N1 infection. This is a form of secondary SPM unlike the one in our case report which is a case of primary SPM.

Conclusions

SPM is a benign condition. According to the etiology it can be primary when it's idiopathic and secondary when concomitant disease is identified. Usually for patients with primary spontaneous pneumomediastinum ambulatory monitoring suffices. Underlying diseases, if identified should be treated. PFT can be done in the follow-up cases to identify risk factors. Inciting maneuvers should be avoided to prevent recurrence. Esophageal pathology and foreign body aspiration should be ruled out.

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Conflicts of interest: None declared

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