Hamman's Syndrome - in Young Asthmatic Female

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Abstract

Spontaneous pneumomediastinum (SPM) is a rare condition where there is presence of air in the mediastinum without any precipitating trauma, surgery or other conditions. It usually develops after alveolar rupture and air penetration into the pulmonary interstitium, the hilum and the mediastinum. It is commonly seen in young males, asthmatics on inhalational drugs, or following severe vomiting, parturition, weight lifting. A young female, known case of bronchial asthma presented to us with history of breathlessness and cough of 3 days and mild swelling of the face and neck of 2 days duration. On examination, she was dyspneic, had subcutaneous swelling of the face, neck and chest bilaterally with palpable crepitus. Respiratory examination showed diffuse polyphonic rhonchi and crepitations. CT thorax showed pneumomediastinum, pneumoperitoneum, pneumothorax and pneumorrhachis. A diagnosis of Hamman's syndrome was made and patient was treated symptomatically and recovered.

Case Presentation

An 18-year-old female, known asthmatic since the age of 10 years presented to the Emergency in the evening hours with history of breathlessness and cough of 3 days duration. She was initially admitted in primary care facility and was started on antibiotics and nebulized bronchodilators. Her symptoms improved with treatment, but noticed painful swelling of face and neck after a day of starting treatment. There was no history of trauma, history of pleural fluid aspiration, history of recent surgery or similar history in the past.

On clinical examination her vitals were stable with 98% SPo2 in room air, B.P 110/70 mmHg, Pulse rate 80 beats/min and was afebrile. There was swelling of cheeks, neck and chest (Figure 1) with palpable crepitations over these areas. Normal vesicular breath sounds with equal intensity and diffuse polyphonic rhonchi were heard all over the lung fields bilaterally.

Fig. 1: Fullness of the upper anterior chest and neck

Blood hematology and biochemistry were within normal limits. Chest X-ray (Figure 2) showed subcutaneous emphysema. Subsequent CT thorax showed (Figures 3, 4 and 5) pneumomediastinum, pneumoperitoneum, minimal pneumothorax and pneumorrhachis and ruled out causes like perforation of esophagus. A diagnosis of spontaneous pneumomediastinum (SPM) or Hamman's syndrome was made.

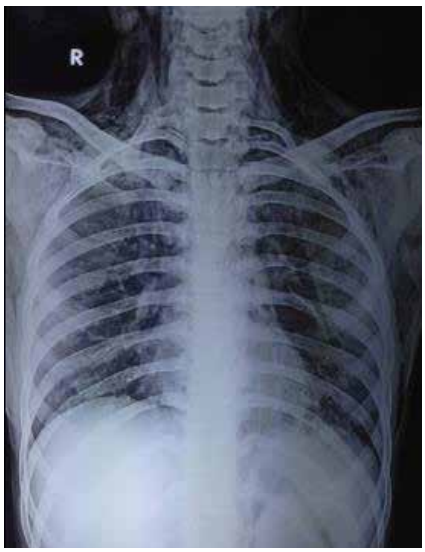

Fig. 2: Chest X-ray showing subcutaneous emphysema

Patient was managed with nebulised salbutamol, oxygen, rest and symptomatic measures. Her symptoms improved and subcutaneous emphysema resolved over a week's period. She was discharged and was advised out-patient follow-up.

Discussion

Pneumomediastinum or mediastinal emphysema refers to the presence of air in the mediastinum.[1] SPM, which was initially described by Laennec in 1819, was further characterized in case series by Hamman in 1939.[2,3] It is a rare condition defined by the presence of air in the mediastinum with no apparent causes like trauma or surgery, which may precipitate pneumomediastinum. Pneumomediastinum may also occur following barotrauma, tracheobronchial injury, procedures like bronchoscopy and cardiothoracic surgeries, frequent retching and vomiting, weight-lifting, parturition, and straining against a closed glottis.[3,4] A history of bronchial asthma and associated coughing bouts has been reported as a possible factor in the development of SPM in up to 25% of cases.[3] In patients with bronchial asthma pneumomediastinum may occur secondary to air trapping due to airway narrowing or mucous plugging, especially after bouts of cough. This sudden increase in intra-alveolar pressure causes rupture of alveoli with escape of air into the pulmonary interstitial spaces. Air then extends along the perivascular sheaths, eventually dissecting into the mediastinum producing mediastinal emphysema-the Macklin effect.[3,5] There may be associated pneumothorax in sporadic cases. Dissection along the carotid perivascular sheaths produces subcutaneous emphysema in the soft tissues of the head and neck.

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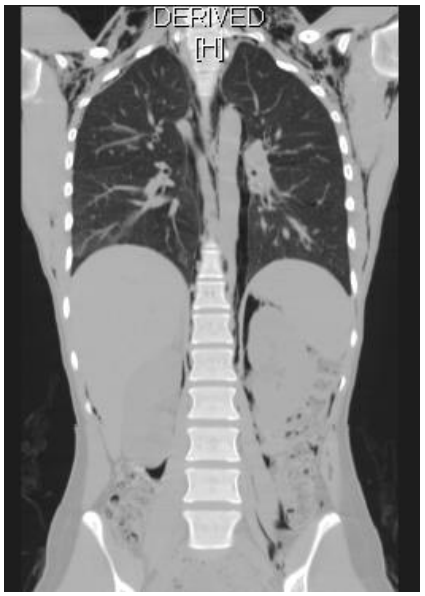

Fig. 3: CT thorax showing subcutaneous emphysema

Most common clinical presentations of Hamman's syndrome are chest pain, dyspnea and subcutaneous emphysema. Other symptoms like neck pain and swelling, dysphagia, hoarseness of voice may also be seen.[1,6,7] Apart from subcutaneous emphysema, Hamman's sign or crunch may be heard in about a fifth of the cases - which is characterised by precordial systolic crepitations synchronous with cardiac cycle, intensified during inspiration and with faint cardiac sounds.[2,3,6,7]

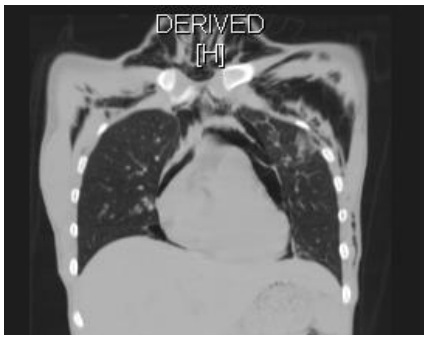

Fig. 4: CT thorax – pneumopericardium, pneumomediastinum

Diagnosis can be made from X- rays of chest and cervical region - if no trauma or perforation suspected - which will reveal air in the mediastinum in the form of transparent bands around the heart, and subcutaneous neck emphysema and occasional pneumothorax. Chest CT may be required if minimal pneumomediastinum is present (which may not be visualized in X-ray) and it also helps anatomically to locate air in the mediastinum.[7,8] Bronchoscopy and esophagoscopy may be indicated if tracheobronchial or esophageal perforation (Boerhaave syndrome) is suspected.

SPM often follows a benign course and is often under diagnosed due to a clinical presentation similar to many respiratory pathologies. The diagnosis of pneumomediastinum should be

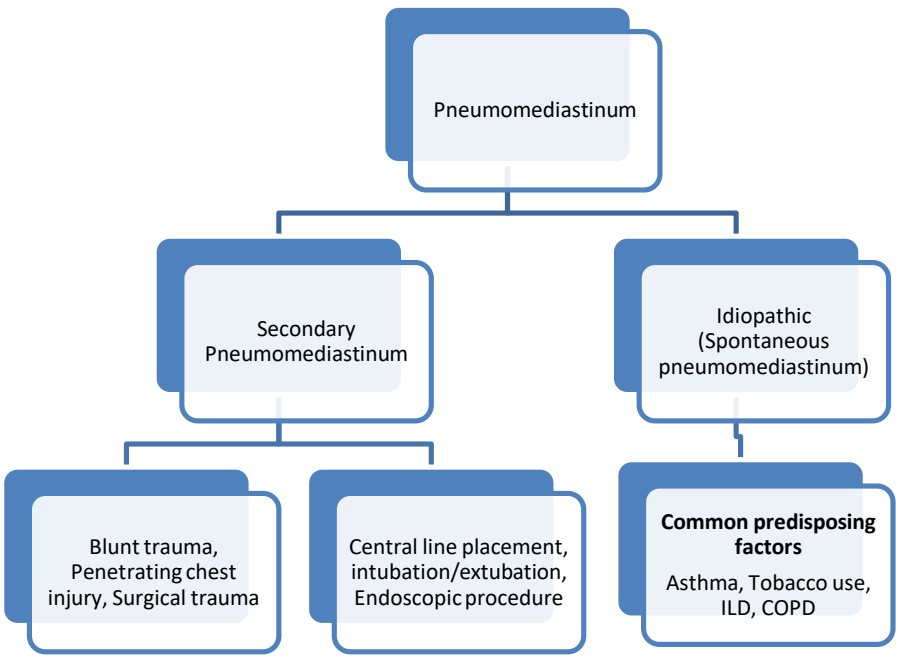

Fig. 6: Classification scheme of pneumomediastinum: Pneumomediastinum can be broken down into two main categories, secondary and idiopathic. Both categories have multiple causes[3]

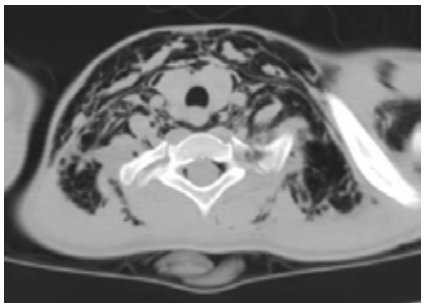

Fig. 5: CT thorax -pneumorrhachis

suspected when an asthma patient experiences substernal chest pain during an acute asthmatic attack. The presence of subcutaneous emphysema in the neck and a "crunching" sound heard over the heart during systole should raise the suspicion of the diagnosis even further. Though an infrequent entity it should be also considered in the differential diagnosis of acute chest pain especially among young people without risk factors for ischaemic heart disease.[9] It is however important to differentiate it from far more serious differentials such as Boerhaave syndrome. Occasional complications include tension pneumomediastinum, unilateral or bilateral pneumothorax, tension pneumopericardium and cardiac tamponade.[3,6,7]

Treatment is mainly conservative and most of the pneumomediastinum resolves spontaneously.[3,4,6,7] Observation of the patient for development of any complication, rest and cardio-pulmonary monitoring is the mainstay of management. Pure oxygen treatment increases the diffusion pressure of nitrogen in the interstitium and promotes rapid absorption of the free air.[3,7] Mediastinoscopy may be rarely required for cases of life threatening tension pneumomediastinum. Thoracotomy and drainage may be needed in cases of pneumothorax.[6,7]

Pneumorrhachis (epidural emphysema or epidural pneumatosis), which was first described in 1977,[10] denotes air in the spinal epidural space and is mostly traumatic or iatrogenic origin. Spontaneous combination of pneumomediastinum with epidural pneumorrhachis without thoracic trauma is very rare. Pneumorrhachis in bronchial asthma is extremely rare; only 14 cases were reported till 2009,[11] and we could find another 2 thereafter.[12,13] Mechanism of pneumorrhachis in bronchial asthma is thought to be due

to rupture of alveoli due to the acute increase in the intra-alveolar pressure and escape of air sequentially into the perivascular space, facial planes of the neck, into the posterior mediastinum and finally to the epidural space.[11]

Conclusion

It is important to keep a diagnosis of SPM in mind in cases of undiagnosed chest pain, and in cases of surgical emphysema. Once diagnosed, it may reassure the caregiver and temper the management plan to follow a path of expectant observation and watch for any sinister signs requiring surgical interventions.

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