

**Clinical Image**

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**Surgical emphysema and pneumothorax: Unfamiliar combination in Ehlers-Danlos syndrome****Vibha Singh; Arijit Jotdar\***; Annanya Soni; Rudra Prakash; Dhruv Kapoor

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**Case description**

A 53-year-old gentleman presented with sudden onset swelling over the neck for the past 5 days. It was associated with dyspnoea on exertion. He was a known patient of Ehlers-Danlos syndrome since sixteen years of age. Initially, the swelling appeared over the neck which gradually progressed over to the anterior chest wall and the nape of the neck. There was no history of trauma, fever, surgery or similar complaints in the past. He was a known smoker. On examination, there was diffuse subcutaneous emphysema over the neck, and the anterior chest wall (Figure 1). Skin crepitus was present. The skin was hyperextensible, fragile and easily stretchable (Figure 1). The patient was vitally stable. On rigid laryngoscopy, bilateral vocal cords were mobile. We admitted the patient and performed a CT scan of the neck and chest to assess the extent of the emphysema. CT scan confirmed an extensive subcutaneous emphysema over the neck, extending posterior to the oesophagus. There were multiple emphysematous bullae over the apical region of the right lung and an area of pneumothorax just in the vicinity of the bullae (Figure 2). The patient was planned for incision and evacuation of the air. A horizontal incision was given over the most prominent part of the swelling and the air was milked out

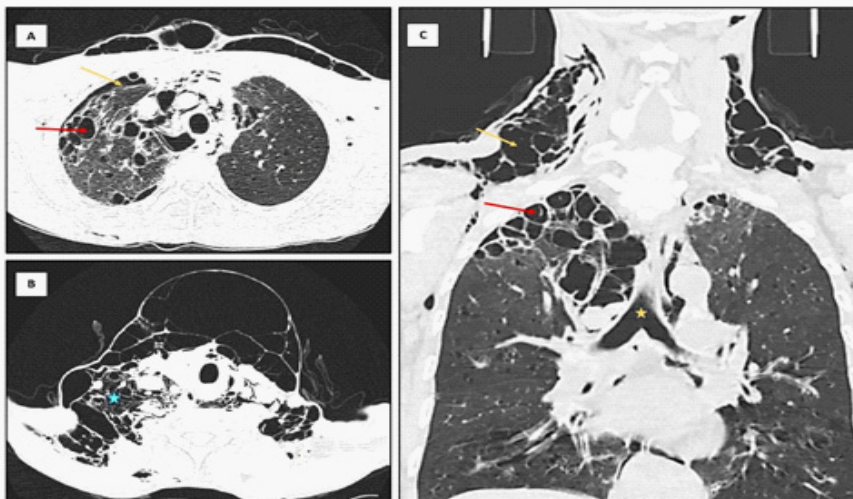
gently. An Intercostal Chest Drain (ICD) was inserted and the condition was closely monitored. The recovery was uneventful. He was discharged after seven days of hospital stay.

Spontaneous pneumothorax with subcutaneous emphysema is one of the lesser-known manifestations of the Ehlers-Danlos syndrome. The pathophysiology is proposed to be the weakening of pleura and pulmonary arteries [1,2]. The lung biopsy of such patients revealed fragility of the alveolar walls that contain type III collagen. This leads to the interstitial emphysema and bleb formation [2]. The resulting respiratory manifestations can be spontaneous pneumothorax and haemothorax [3]. Due to high chance of recurrence of pneumothorax in such patients, video-associated thoracoscopic pleurodesis and application of fibrin glue is suggested.

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**Figure 1:** (A, B, C) Surgical emphysema present over the anterior and lateral aspects of the neck extending from chin to the anterior chest wall. (D) Hyper-elastic skin that can be stretched easily.



**Figure 2:** (A) Axial section of CT scan showing Zone of pneumothorax in right lung (yellow arrow) and emphysematous bulla (red arrow), (B) Axial section showing emphysematous bulla in apical lung region on right side communicating with subcutaneous emphysema (Blue star), (C) Coronal section of CT scan showing multiple emphysematous bullae in the apex of right lung (red arrow), surgical emphysema over neck (yellow arrow) and the carina (yellow star).

## References

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