An 11-year-old boy was presented to the emergency department with history of skin lesions for 2 months as well as mild dyspnea, and recent progressive neck swelling. The patient was a known case of major thalassemia who had undergone hematopoietic stem cell transplantation (HSCT) eight months before and was on immunosuppressive therapy. On physical examination, he was lethargic with normal vital signs. There were several scattered brownish macules over his skin. On palpation, there was diffuse crepitation all over his chest, neck, and upper abdomen. In a laboratory study, there was normocytic normochromic anemia and increased lactate dehydrogenase (300 IU/L). A chest X-ray (Figure 1) and computed tomography (CT) scan (Figure 2) were performed.

**What is your diagnosis?**

See the next page for your diagnosis.
The X-ray and CT scan showed diffuse subcutaneous emphysema (white arrowhead), pneumothorax (white arrow), pneumomediastinum (black arrow), and pneumopericardium (black arrowhead). The diagnosis of thoracic air-leak syndrome (TALS) due to chronic graft versus host disease (cGVHD) was made. Although a pulmonary complication in cGVHD is a common condition, TALS is encountered rarely. TALS complicates less than 3% of HSCT patients and mostly caused by bronchiolitis obliterans syndrome (BOS). In most cases, blunt alveolar rupture (even due to trivial trauma, such as coughing), initially leads to pulmonary interstitial emphysema and then dissecting retrogradely along the perivascular sheaths into the hilum (Macklin effect). Spontaneous pneumothorax is the most common presentation, which is followed by other conditions such as subcutaneous emphysema, pneumomediastinum, and pneumopericardium. It cannot be treated easily and considered as a poor prognostic indicator. The patient was referred to oncology ward, but unfortunately was deteriorated and died few days later due to sepsis.

References