CASE REPORT

Non-traumatic trans-diaphragmatic intercostal hernia and cor pulmonale in a patient with poorly controlled obstructive sleep apnoea

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SUMMARY

Trans-diaphragmatic intercostal hernia is a rare entity. Patient with multiple medical comorbidities, including obstructive sleep apnoea, presents with shortness of breath, leg oedema and a bulging swelling through the right chest wall. CT shows partial herniation of the right lung and liver through intercostal space and an echocardiogram reveals right heart failure. He was treated initially with continuous positive airway pressure with poor response and subsequently treated with adaptive servo ventilation with much better symptomatic relief and treatment tolerance.

On examination, he had bilateral leg oedema and visible swelling protruding through the right chest wall which was more pronounced on forced expiration, Valsalva manoeuvre and coughing with evidence of dullness at the lung bases ([video 1](#)).

INVESTIGATIONS

Echocardiogram showed mildly impaired left ventricular function and severely impaired right ventricular function with raised pulmonary artery pressures (69–84 mm Hg).

A CT ([figure 1](#)) chest showed partial herniation of the right lung and the liver through a defect in the chest wall.

BACKGROUND

Trans-diaphragmatic intercostal hernia (TDICH) is a rare clinical entity and few cases have been reported. It occurs when there is a defect simultaneously in the diaphragm and the chest wall leading to herniation of abdominal organs into the thoracic cavity and through a defect in the chest wall. Patients are usually asymptomatic but can present with dyspnoea, chest pain and a noticeable swelling in the chest.

Such hernias occur most commonly as a result of some form of traumatic incident, blunt or penetrating, with or without associated rib fractures or through the thoracotomy site.

The chest wall swelling is more noticeable during manoeuvres that increase intrapleural pressure such as forced expiration, coughing and Valsalva manoeuvre.¹

Ideally, symptomatic patients are managed by surgical correction of the diaphragmatic and chest wall defect;¹ however, there is not much evidence regarding non-surgical management. Therefore, we report the case of a patient with a history of progressive breathlessness and other comorbidities precluding him from undergoing major surgical intervention for correction of the symptomatic TDICH.

DIFFERENTIAL DIAGNOSIS

Lipoma
Osteochondroma
Chondrosarcoma
Chest wall abscess
Pleural-based mass

TREATMENT

He was initially treated with continuous positive airway pressure followed by very short duration of Bi-level ventilation. Adaptive servo ventilation (ASV) was established with good tolerance and response. The patient was also on diuretics.

The patient was reviewed by the thoracic surgeons with a view to surgically repairing the defect; however, he was deemed to be high surgical risk.

Video 1 This is a video compilation of the effect of inspiration, expiration and coughing manoeuvre on the trans-diaphragmatic intercostal hernia. Zero to 27 s demonstrates inspiration and expiration, followed by coughing manoeuvre for 4 s (27–31 s). From 32 to 36 s, the video demonstrates inspiration and expiration followed by 3 s of coughing manoeuvre. The video ends with demonstration of inspiration and expiration for the last 7 s (40–47 s).

CASE PRESENTATION

A patient aged 66 years with no history of trauma presented with progressive shortness of breath and bilateral leg swelling.

Medical history includes chronic back pain, hypertension, atrial fibrillation, ST elevation myocardial infarction, congestive cardiac failure, peripheral vascular disease and obstructive sleep apnoea (OSA).

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Figure 1 CT image.

OUTCOME AND FOLLOW-UP

He continued to use ASV device and tolerated it well. His symptoms were alleviated and he had a good quality of life.

DISCUSSION

The diaphragm is primary muscle of breathing and normally when actively contracts creates negative intrapleural pressure leading the air through the lungs; however, when there is defect in the diaphragm, this negative pressure can lead the abdominal content to herniate into the chest cavity.² With the creation of negative intrapleural pressure, the defective segment in the chest wall will cave in behaving like flail segment seen in significant acute chest trauma. During expiration, the positive intrapleural pressure will cause the lung herniation through the chest wall defect more pronounced through the chest leading to less effective pressure to exhale.

The natural active process of breathing is a negative pressure one and using positive airway pressure modality via non-invasive ventilation will overcome the problem created by the underlying anatomical defects.

In our case, the patient had severe OSA and TDICH, both of which are likely to have contributed to his clinical picture and the development of cor pulmonale through chronic hypoxaemia. Ideally, for patients with symptomatic anatomical defects, surgical repair is the standard treatment; however, if that is not feasible, we recommend non-invasive positive pressure ventilation. In our patient with underlying significant OSA, we found ASV is better tolerated.

Learning points

- Trans-diaphragmatic intercostal hernias are rare and pose a diagnostic and management challenge.
- First-line treatment of large symptomatic trans-diaphragmatic intercostal hernias is surgical correction.
- In cases where surgical intervention is not possible, ventilatory support remains the alternative modality of treatment.
- Early diagnosis and management of obstructive sleep apnoea is important in order to avoid long-term complications.

Contributors SM is the corresponding author and has overseen the entire writing up of case report. SBM has contributed in selecting and modifying the video and also the context of the article. JS has written the initial draft of the case report. This interesting case along with the video was presented in Trust-wide grandround presentation.

Competing interests None declared.

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