CASE REPORT

Pectus excavatum severity underestimated due to lack of objective measures in radiological reports

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SUMMARY

During a routine physical examination, the degree of pectus excavatum (PE) is not always appreciated as the external appearance does not always reflect the severity of the deformity. In the patient in this case report, the severity had been underestimated for 33 years. The physicians, having requested standard two-view chest radiographs, had relied solely on the radiological reports, where the PE had been ambiguously described as 'moderate' or 'substantial'. In patients where PE has been observed, it is essential that an objective numeric measure of severity, using the Haller index, is included in radiological reports.

BACKGROUND

Pectus excavatum (PE) is the term used to describe a depression in the anterior chest wall as a result of a dorsal deviation of the sternum and the third to seventh rib or costal cartilage. It is the most common chest wall deformity, representing 90% of congenital chest wall deformities, and affects 1–8 in 1000 births, predominantly males (gender ratio between 2:1 and 9:1).¹

The Haller index, a numeric evaluation of the severity of PE, is defined as the ratio between the lateral distance of the chest wall (inner margins), and the narrowest anteroposterior distance between the vertebrae and sternum, and can be calculated from a CT scan or two-view chest radiographs.^{2–3}

The normal chest has a Haller index of 2.5. Values >3.2 are considered for surgical intervention and values >3.5 are considered severe.⁴

Severe cases of PE have been shown to be associated with reduced pulmonary function.⁵ As early as 1951, decreases in vital capacity (VC) of more than 50% were reported in 9 of 11 preoperative cases.⁶ In a more recent case report of a 29-year-old woman with severe PE (Haller index 17), there was a significant decrease in pulmonary function (VC 19% predicted), cor pulmonale and chronic respiratory acidosis. No other cause for these conditions could be found.⁷ Recurrent syncope caused by compression of the inferior vena cava has been described in a patient with severe PE (Haller index 18.8).⁸ Right ventricular dysfunction caused by PE has been described in a case with Haller index 4.2, a value much more likely to be seen.⁹

CASE PRESENTATION

A 41-year-old man (a coauthor of this paper) presented with postural dyspnoea that resolved while

supine. He had advanced facioscapulothoracic muscular dystrophy type 1, a neuromuscular disorder, but this condition is not commonly associated with respiratory insufficiency.¹⁰ Referral to a pulmonologist was arranged.

Standard two-view chest radiographs were taken. The radiologist reported a 'substantial' PE but no evidence of pulmonary pathology. At the time, the patient presented two-view chest radiographs taken 24 years earlier (aged 17 years), but these were not reviewed by the radiologist. Pulmonary function tests revealed a restrictive defect: VC 1.57 L (33% predicted). No cause was identified for the patient's symptoms.

Pulmonary function tests performed 8 years later (aged 49 years) showed a marked decline in VC with a value of 0.74 L (17% predicted). The patient was referred to a centre for home mechanical ventilation. Pulse rate and SpO₂ were normal, but hypercapnia with high levels of bicarbonate and base excess indicated metabolically compensated nocturnal hypoventilation. Nocturnal non-invasive positive pressure ventilation therapy was initiated. Although this resulted in diurnal normocapnia, this did not result in any improvement in the postural dyspnoea.

The patient observed that the episodes of dyspnoea were accompanied by tachycardia and that the pulse rate dropped significantly after lying down. This led him to speculate that some mechanical, posture-dependent compression could be a factor in causing the symptoms he was experiencing.

The patient was referred to a cardiologist, who requested a range of diagnostic tests including chest radiographs. The radiologist compared the two-view chest radiographs with those taken 8 years previously and reported 'substantial' PE. He concluded that the patient's condition was essentially unchanged. The cardiologist was unable to identify any cause of the patient's symptoms.

One year later (aged 50 years), research on PubMed led the patient to suspect PE as a possible factor in the cause of his symptoms. He requested referral to a cardiologist who was the coauthor of publications relating to complications of PE in adults.¹¹ The cardiologist reviewed the radiographs taken 9 years earlier (figure 1). It was the most severe case of PE he had personally encountered. A CT scan confirmed the most likely cause of the postural dyspnoea, first experienced by the patient 9 years prior—almost complete compression of the right atrium by the sternum (figure 2 Haller index=30).

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Figure 1 Lateral chest radiograph at age 41 years. Note the short sterno-vertebral distance (<1 cm).

DIFFERENTIAL DIAGNOSIS

- Restrictive lung disease
- Heart failure

Final diagnosis: Severe PE, Haller index 30, compression of the heart.

OUTCOME AND FOLLOW-UP

The patient declined surgical correction as the procedure would not enhance his pulmonary function and there was a prohibitive risk.

Figure 2 Transverse CT at age 50 years. Note the cardiac displacement and compression.

DISCUSSION

During a routine physical examination, the severity of PE is not always appreciated as the external appearance does not always reflect the severity of the deformity.

Standard two-view chest radiographs and CT scans are routinely used as diagnostic tools. The physician requesting the investigation rarely views the images and relies solely on the radiologist's report. Terms such as 'moderate' or 'substantial' are subject to misinterpretation and it is therefore essential that an objective numeric measure is included in the radiology report.

In the case of this patient, the extreme severity was clearly defined on the two-view chest radiographs taken 33, 9 and 1 years previously (retrospectively calculated HI >7, 30 and 30, respectively) but this extreme severity was repeatedly overlooked by the physicians ordering the radiological tests. Had the radiologists included a numerical measure in their reports, this would not have occurred. This case report highlights the need to include the Haller index in all radiological reports when PE is observed.

The mechanism of postural dyspnoea is explained as follows: postural changes that increase the concavity of the thoracic wall (eg, bending over) result in increased compression of the right atrium and/or ventricle.¹¹

Facioscapulohumeral dystrophy (FSHD) is an inherited myopathy with variable age at onset and variable rate of progression. Typically, the disease initially involves facial and scapular muscles, followed by spreading to lower limb muscles. Although 10–20% of patients become wheelchair-bound, about the same percentage remain asymptomatic.¹⁰

FSHD is a known risk factor for PE.¹² Atrophy of the pectoralis major muscle, which takes place early in the course of FSHD, may be a causative factor.¹³

Learning points

- The external presentation of pectus excavatum does not always show the extent of the deformity.
- In radiological reports, adjectives such as 'moderate' or 'substantial' can be misleading.
- It took 33 years to realise the severity of pectus excavatum in a patient who should have been referred for surgical correction at 17 years.
- The Haller index should always be reported when pectus excavatum is observed.
- Facioscapulohumeral dystrophy is a risk factor for pectus excavatum.

Contributors JK treated this patient and took the responsibility in gathering the data and wrote the review. PH wrote the manuscript and was involved in literature search and gathering the data. AG was involved in patient's care and wrote the manuscript. MdB reviewed the X-rays and commented on the manuscript.

Competing interests None declared.

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