AIR OESOPHAGOGRAM: A FREQUENT, BUT NOT A SPECIFIC SIGN OF OESOPHAGEAL INVOLVEMENT IN CONNECTIVE TISSUE DISEASES

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SUMMARY

Objective. This study investigates the role of the air oesophagogram in conventional chest X-rays for the diagnosis of oesophageal dysmotility in patients with connective tissue diseases.

Methods. Fifty-one patients with connective tissue diseases were studied by oesophageal manometry and lateral and posterior–anterior chest X-rays. The presence or absence of oesophageal air on chest X-rays was evaluated separately in the upper, middle and distal segment of the oesophagus. Forty-seven chest X-rays of patients without connective tissue diseases, who had undergone oesophageal manometry for the evaluation of oesophagus-related symptoms and who had normal oesophageal function, were analysed as a control.

Results. A total of 23/51 patients with connective tissue diseases showed oesophageal dysfunction in manometry; 16/51 patients (31%) had air in two or more oesophageal segments on the lateral chest X-ray. There was a significant association of manometrically proven oesophageal dysmotility and air in two or three oesophageal segments (P < 0.05; specificity 48%, specificity 82%). However, the prevalence of an air oesophagogram showed no significant difference between patients with connective tissue diseases and the control group (10/47; 21%).

Conclusion. The radiological sign of an air oesophagogram is neither sensitive nor specific enough to omit oesophageal motility studies in patients with connective tissue diseases.

KEY WORDS: Systemic sclerosis, Oesophagus, Air, Manometry, X-ray, Connective tissue disease.

IMPAIRMENT of oesophageal motor function is well recognized in several connective tissue diseases. The most common systemic disease causing oesophageal oesophageal dysmotility is systemic sclerosis. Oesophageal dysfunction caused by atrophy and fibrosis of the smooth muscle portion has been reported to occur in 50–80% of these patients [1–4]. In polymyositis and mixed connective tissue disease, the oesophagus appears to be involved in 60–85% of cases [5, 6], whereas in systemic lupus erythematosus and primary Sjögren’s syndrome much lower prevalences of 25–36% have been reported [7–9].

In 1966, Dinsmore et al. [10] were the first to describe air on conventional chest roentgenograms within the intrathoracic portion of the oesophagus in 12 of 16 patients with systemic sclerosis. They explained this finding by diminished peristalsis and limited collapsibility of the ‘scleroderma oesophagus’. They proposed that an air oesophagogram is strongly suggestive of systemic sclerosis, especially in the absence of other conditions known to induce this phenomenon, such as achalasia, laryngectomy or after thoracic surgery. Other studies have reported an air oesophagogram in 1–20% of patients with systemic sclerosis [11–15].

The aim of this study was to assess systematically the diagnostic significance of an air oesophagogram in patients with connective tissue diseases and suspected oesophageal involvement by comparing the findings in chest roentgenograms with oesophageal manometry as a ‘gold standard’ of oesophageal motility testing.

PATIENTS AND METHODS

Charts of all patients who had undergone oesophageal manometry for the diagnosis of oesophageal dysmotility in suspected or proven connective tissue disorders from 1992 to 1996 were reviewed. Fifty-one patients for whom both lateral and posterior–anterior (p.a.) chest X-rays were available were enrolled in the study. Forty patients were female, 11 male; mean age was 54 ± 13.5 (range 24–84) yr. Final diagnoses of patients were systemic sclerosis (n = 31), localized scleroderma (n = 3), mixed connective tissue disease (n = 4), unclassified connective tissue disease or overlap syndromes (n = 6), systemic lupus erythematosus (n = 4), primary Sjögren’s syndrome (n = 2) and dermatomyositis (n = 1).

Forty-seven chest X-rays of patients without evidence for connective tissue diseases who had undergone oesophageal manometry for oesophagus-related symptoms in the same laboratory, and who had shown no manometric evidence for tubular oesophageal dysfunction, were analysed as a control group. Indications for manometry had been dysphagia or globus sensation (n = 13), chest pain of presumed non-cardiac origin (n = 21), gastro-oesophageal reflux (n = 11) and others (n = 2). The mean age of the control group was 52 ± 15.1 (range 15–79) yr and thus was comparable to the patient group with connective tissue disorders; however, there were significantly more men in the control group (32 male, 16 female).

Oesophageal manometry had been performed in a standardized procedure after an overnight fast.
with an eight-lumen water-perfused polyvinylchloride tube (outer diameter 4.5 mm; Synectics Medical, Stockholm). Lower oesophageal sphincter pressure was recorded by a station pull-through method using four distal circular openings. Tubular oesophageal function was assessed using five openings at a distance of 5 cm each, thus covering 20 cm of the oesophageal body. Values from 10 dry and 10 wet swallows (using 5 ml of water) were averaged separately. Manometric tracings were interpreted using standard criteria [16]. Oesophageal function was classified as normal when there were peristaltic contractions with an amplitude exceeding the lower range of normal values [16]. Oesophageal function was classified as abnormal when there was (1) aperistalsis or (2) a marked hypoperistalsis (e.g. weak or multiple peaked contractions with an amplitude below the lower range of normal values) in at least two of the five recording sites of tubular oesophageal function.

Chest X-rays of all patients were reassessed by a consultant radiologist blinded to the diagnosis and to the results of oesophageal manometry. The presence or absence of air were evaluated separately in the upper, middle and distal segment of the oesophagus, each on p.a. and lateral views. An air oesophagogram was defined as the presence of air in at least two oesophageal segments. Sensitivity, specificity and predictive values of radiologically detectable air in more than one or more than two oesophageal segments were calculated separately using oesophageal manometry as the gold standard. The $\chi^2$ test with Yates continuity correction was applied to investigate the association of an air oesophagogram with oesophageal dysmotility. All results are expressed as the mean ± s.d.

RESULTS

A total of 23 of 51 patients (45%) had manometric evidence for an oesophageal involvement of the underlying connective tissue disease: 17 patients showed distal aperistalsis, six patients exhibited a marked hypocontractility of the distal oesophagus. The majority of patients with manometrically proven oesophageal dysfunction suffered from systemic sclerosis ($n = 18$); the other five patients had mixed connective tissue disease ($n = 1$), unclassified connective tissue disease ($n = 2$), dermatomyositis ($n = 1$) and Sjögren’s syndrome ($n = 1$).

Chest X-rays revealed air in the oesophagus in 21 of 51 patients (41%) (Fig. 1): five patients had air in one, 14 in two, and two in all three oesophageal segments. In 15 of these patients, air could only be seen in the lateral view; in only six patients could oesophageal air be detected in both the lateral and p.a. view.

In patients with connective tissue diseases, the sign of air in one or more oesophageal segments had a sensitivity of 52% and a specificity of 68% for oesophageal dysfunction compared to oesophageal manometry as a ‘gold standard’ (Table I). When air was present in two or three segments, specificity improved to 82% with a decrease in sensitivity to 48% and a positive predictive value of 69% (Table II). There was a significant association of oesophageal dysmotility with air in at least two oesophageal segments ($\chi^2$ test, $P < 0.05$). Lower oesophageal sphincter pressure in patients with manometrically proven oesophageal dysfunction showed no significant difference whether an air oesophagogram was present or not (11.5 ± 8.7 vs 16.4 ± 12.5 mmHg; $P = 0.44$).

In the control group of patients with oesophagus-related symptoms, but manometrically proven normal tubular oesophageal function and no evidence for connective tissue disease, 17 patients (36%) had air in at least one and 10 patients (21%) had air in at least two oesophageal segments. This was not significantly different from all studied patients with connective tissue diseases. Non-cardiac chest pain ($n = 6$), heartburn ($n = 3$) and dysphagia ($n = 1$) were the main presenting symptoms in control patients with a positive air oesophagogram. Lower oesophageal sphincter pressure did not differ in control patients with and without air oesophagogram (20.0 ± 10.3 vs 24.4 ± 13.9 mmHg).

DISCUSSION

Only a few case reports and studies have been published about the air oesophagogram [11–15] since
TABLE I
Predictive value of oesophageal air (≥1 segment) compared to oesophageal manometry

<table>
<thead>
<tr>
<th>Patients with connective tissue diseases and normal manometry</th>
<th>Patients with connective tissue diseases and manometrically proven oesophageal dysmotility</th>
<th>All patients with connective tissue diseases</th>
<th>Controls without connective tissue diseases and normal manometry</th>
</tr>
</thead>
<tbody>
<tr>
<td>No oesophageal air</td>
<td>19</td>
<td>11</td>
<td>30</td>
</tr>
<tr>
<td>Air in ≥1 oesophageal segments</td>
<td>9</td>
<td>12</td>
<td>21</td>
</tr>
<tr>
<td>Total</td>
<td>28</td>
<td>23</td>
<td>51</td>
</tr>
</tbody>
</table>

Predictive values for patients with connective tissue diseases compared to oesophageal manometry: sensitivity 52.2%; specificity 67.9%; positive predictive value 57.1%; negative predictive value 63.3%.

TABLE II
Predictive value of oesophageal air (≥2 segments) compared to oesophageal manometry

<table>
<thead>
<tr>
<th>Patients with connective tissue diseases and normal manometry</th>
<th>Patients with connective tissue diseases and manometrically proven oesophageal dysmotility</th>
<th>All patients with connective tissue diseases</th>
<th>Controls without connective tissue diseases and normal manometry</th>
</tr>
</thead>
<tbody>
<tr>
<td>Air in ≤1 oesophageal segment</td>
<td>25</td>
<td>12</td>
<td>35</td>
</tr>
<tr>
<td>Air in ≥2 oesophageal segments</td>
<td>5</td>
<td>11</td>
<td>16</td>
</tr>
<tr>
<td>Total</td>
<td>28</td>
<td>23</td>
<td>51</td>
</tr>
</tbody>
</table>

Predictive values for patients with connective tissue diseases compared to oesophageal manometry: sensitivity 47.8%; specificity 82.1%; positive predictive value 68.8%; negative predictive value 65.7%.

its first description as a sign of oesophageal involvement in systemic sclerosis [10]. However, definitions of air oesophagograms (referred to either as segmental air collection or air outlining the entire oesophagus) and even radiographic techniques (p.a. or lateral views) differed considerably, resulting in widely scattered figures for the prevalence of this sign (1–75%).

To our knowledge, this is the first study to compare systematically the results of modern oesophageal motility measurements with the presence or absence of the oesophageal air sign. Oesophageal air in at least one oesophageal segment was detected in nine of 28 patients without manometric evidence for oesophageal dysmotility, leading to a specificity of only 68%. If only patients with air collections in two or three segments were considered positive, specificity improved to 83%.

We found oesophageal air in connective tissue diseases other than systemic sclerosis (e.g. dermatomyositis, Sjögren’s syndrome) which, to our knowledge, has not yet been described. However, the presumed pathophysiological mechanism of oesophageal hypomotility preventing oesophageal collapse is quite similar to patients with systemic sclerosis. Surprisingly, the prevalence of an air oesophagogram in the control group, which underwent manometry for oesophagus-related symptoms, was rather high despite manometrically proven normal tubular oesophageal function. Thus, besides oesophageal hypocontractility and fibrosis, other factors such as aerophagia may play a role in the development of an air oesophagogram.

In conclusion, air in the oesophagus observed in a lateral chest X-ray is a rather frequent finding in patients with connective tissue diseases. Only air columns extending over two-thirds of the oesophagus are associated with manometrically confirmed oesophageal dysmotility and may be interpreted as a sign of oesophageal involvement. However, oesophageal air collections are quite common in patients with oesophagus-related symptoms and no manometric signs of oesophageal dysfunction. Thus, the radiological sign of air oesophagogram is neither sensitive nor specific enough to omit oesophageal motility studies.

ACKNOWLEDGEMENT
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REFERENCES
7. Stevens MB, Hookman P, Siegel CI et al. Aperistalsis of the esophagus in patients with connective tissue dis-