

Response

To the Editor:

We thank Dr Garcia-Zamalloa for his comments regarding our case series of patients with arthritis-associated pseudochylothorax who did not have significant pleural thickening (October 2009). We agree that the low number of reported cases has hitherto prevented a robust description of the pathogenesis and natural history of pseudochylothorax. Hopefully, our case series serves to add to our understanding of the disease.

Conventionally, pseudochylothorax is viewed as a single disease entity; this preconception should be challenged. The majority (54%) of the published cases of pseudochylothorax were associated with TB, followed by rheumatoid arthritis (9%).2 A large number of other conditions has also been implied, although each has been linked with very few cases of pseudochylothorax. Pseudochyle is defined by a high pleural fluid cholesterol composition (presence of cholesterol crystals on microscopic examination or cholesterol concentration >200 mg/dL).3 This definition makes no assumption about the underlying pathogenetic mechanisms leading to pseudochyle formation. The previous literature has always associated pseudochylothorax with significant pleural thickening (a complication relatively common in TB) and assumed the fibrotic pleura plays a role in the accumulation of cholesterol within the pleural cavity. Our report of six cases of pseudochylothoraces—all associated with arthritis—without significantly thickened pleura contradicts such belief. Rather than one unifying mechanism resulting in pseudochyle formation, it is possible that pseudochyle actually represents a common end point of a variety of disease pathophysiologic conditions; those associated with TB may arise from a separate mechanism from those associated with arthritis. Expansion of such a theory could provide a plausible explanation for the diversity in rates of formation of pseudochyle and other pathologic findings (such as degree of pleural thickening), as seen in our series.

Differing pathogenetic mechanisms may also underlie the differing clinical outcomes. Regardless of underlying pathology, it would seem reasonable to propose better outcomes of pseudochylothoraces with nonfibrotic pleura, as seen in our cohort, over those with dense pleural thickening.

Like Dr Garcia-Zamalloa, we suspect that pseudochylothorax is underdiagnosed. Up to 20% of pleural effusions remain undiagnosed in routine clinical practice, and pleural fluid cholesterol concentration is infrequently evaluated, especially in the absence of extensive pleural thickening. A prospective study of pleural fluid cholesterol concentration (and cholesterol crystals) in undiagnosed pleural effusions will help determine the degree of underdiagnosis of this condition.

Specifically with regard to arthritis-associated pseudochylothoraces, our patients demonstrated pleural fluid volume control or reduction with intensified rheumatoid arthritis treatment. One could postulate that many patients with inflammatory arthritis may develop pseudochylothorax at some point during the evolution of their disease; the pseudochyle may remain uninvestigated and undiagnosed if subsequent intensification of arthritis treatment results in resolution of the pleural effusion.

Improved physician awareness of pseudochylothorax should improve rates of diagnosis. Cumulative knowledge may lead to new and testable hypotheses regarding the disease pathogenesis and, ultimately, much-needed studies on better treatment of pseudochylothorax.

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Solitary Fibrous Tumors of the Pleura
An Estimation of Population Incidence

To the Editor:

Primary tumors of the pleura present as either diffuse malignant mesotheliomas or localized solitary fibrous tumors of the pleura (SFTPs), the majority of which are benign. We have used nationwide registries in Iceland to study SFTP and establish an age-standardized incidence rate ([ASR] world standard population), which has not been previously reported. All patients diagnosed with histologically proven SFTP in Iceland from 1984 to 2007 were identified through a centralized pathology registry. For comparison, information on mesotheliomas was obtained from the population-based Icelandic Cancer Registry.

Eleven patients were diagnosed with SFTP (mean age 60 years; eight women) compared with 33 with mesothelioma (mean age 68 years; four women) (Table 1). The ASRs for SFTP and mesothelioma were 1.4 and 4.0 per million (95% CI, 0.54-2.2 and 2.6-5.4), respectively. All cases of SFTP had a benign histology. Only three cases were symptomatic (dyspnea, pneumonia, pleuritic pain), with eight incidentally detected. Apart from one case diagnosed at autopsy, all patients with SFTP were treated with complete surgical resection of the tumor. Average tumor diameter was 6.5 cm (range, 1.3-20 cm) and weight was 130 g (range 53-3260 g), with eight of the tumors arising from the visceral and...
three from the parietal pleura. At 70 months’ median follow-up, there has been no recurrence or disease-related death.

More than 800 cases of SFTP have been reported in the literature. Most cases occur between the sixth and seventh decades, with equal frequency in both sexes. Patients with benign SFTP have symptoms in 54% to 67% of cases compared with 75% with malignant tumors. Predictors of malignant behavior include increased mitoses and cellularity, nuclear pleomorphism, tumor size, necrosis, and stromal or vascular invasion.

In this study, we found that during 24 years in Iceland, 46 cases of primary pleural tumors were diagnosed, one-fourth of these being SFTPs, which all had a benign course. Despite the small number of patients, this population-based study enables the calculation of ASR. This is, to our knowledge, the first calculation of an age-specific, sex-specific, and population-based standardized incidence rate. This population-based study enables the calculation of an age-specific, sex-specific, and population-based standardized incidence rate for this disease, but it can be compared with the previously established incidence of 2.8 per 100,000 registered patients at a single institution. Because many patients with SFTP are asymptomatic, incidence rates are affected by the likelihood of incidental detection, primarily through medical imaging of the chest. Therefore, the ASR for SFTP in Iceland, with its single-payer health care system and comparatively easy access to imaging studies, is possibly in the higher range. More than 800 cases of SFTP have been reported in the literature. Most cases occur between the sixth and seventh decades, with equal frequency in both sexes. Patients with benign SFTP have symptoms in 54% to 67% of cases compared with 75% with malignant tumors. Predictors of malignant behavior include increased mitoses and cellularity, nuclear pleomorphism, tumor size, necrosis, and stromal or vascular invasion.

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Table 1—Summary of Clinical Data for 11 Patients Diagnosed With Solitary Fibrous Tumors of the Pleura in Iceland 1984-2007

<table>
<thead>
<tr>
<th>No.</th>
<th>Age, y/sex</th>
<th>Symptoms</th>
<th>History of Smoking/Pack-yr</th>
<th>History of Cancer</th>
<th>Surgical Resection</th>
<th>Max. Tumor Diameter, cm</th>
<th>Survival, mo</th>
<th>Alive Dec 31, 2007</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>53/F</td>
<td>N/A</td>
<td>Y/35</td>
<td>Lung cancer (NSCLC)</td>
<td>Y</td>
<td>5</td>
<td>77</td>
<td>N</td>
</tr>
<tr>
<td>2</td>
<td>61/F</td>
<td>Incidental finding on CXR</td>
<td>Y/20</td>
<td>N</td>
<td>Y</td>
<td>6.5</td>
<td>207</td>
<td>Y</td>
</tr>
<tr>
<td>3</td>
<td>66/M</td>
<td>Incidental finding at thoracotomy</td>
<td>Y/50</td>
<td>Urinary bladder cancer</td>
<td>Y</td>
<td>1.3</td>
<td>83</td>
<td>N</td>
</tr>
<tr>
<td>4</td>
<td>72/F</td>
<td>Obstructed breathing and dyspnea; Tumor diagnosed at autopsy.</td>
<td>Y/50</td>
<td>N</td>
<td>N</td>
<td>20</td>
<td>...</td>
<td>N</td>
</tr>
<tr>
<td>5</td>
<td>43/F</td>
<td>Pleuritic pain</td>
<td>Y/10-20</td>
<td>Malignant melanoma</td>
<td>Y</td>
<td>4.8</td>
<td>150</td>
<td>Y</td>
</tr>
<tr>
<td>6</td>
<td>69/M</td>
<td>Incidental finding on CXR</td>
<td>Y/54</td>
<td>N</td>
<td>Y</td>
<td>1.3</td>
<td>9</td>
<td>N</td>
</tr>
<tr>
<td>7</td>
<td>37/M</td>
<td>Incidental finding on CXR</td>
<td>Y/20-30</td>
<td>N</td>
<td>Y</td>
<td>3.2</td>
<td>108</td>
<td>Y</td>
</tr>
<tr>
<td>8</td>
<td>81/F</td>
<td>Incidental finding on CXR</td>
<td>Y/30</td>
<td>N</td>
<td>Y</td>
<td>9.5</td>
<td>37</td>
<td>N</td>
</tr>
<tr>
<td>9</td>
<td>68/F</td>
<td>Incidental finding on CXR</td>
<td>N</td>
<td>N</td>
<td>Y</td>
<td>10</td>
<td>62</td>
<td>Y</td>
</tr>
<tr>
<td>10</td>
<td>71/F</td>
<td>Repeated lower respiratory infections and obstructed breathing</td>
<td>Y/20</td>
<td>Multiple myeloma; lung cancer (NSCLC)</td>
<td>Y</td>
<td>17</td>
<td>13</td>
<td>N</td>
</tr>
<tr>
<td>11</td>
<td>32/F</td>
<td>Incidental finding on CXR</td>
<td>N</td>
<td>N</td>
<td>Y</td>
<td>9.9</td>
<td>4</td>
<td>Y</td>
</tr>
</tbody>
</table>

CXR = chest radiograph; F = female; M = male; Max = maximum; N = no; N/A = information not available; NSCLC = non-small cell lung cancer; Y = yes.

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