PHOTO QUIZ

Two siblings with hepatosplenomegaly and pulmonary reticulation

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CASE REPORT

A 28-year-old woman with stable hepatosplenomegaly (diagnosed at the age of 2-3 years) presented with a one-year history of progressive dyspnoea. She denied other pulmonary symptoms. The patient had a younger sister, aged 25 years, who had also had hepatosplenomegaly since early childhood and complained of mild dyspnoea. Physical examination in both sisters confirmed hepatosplenomegaly. High-resolution computed tomography performed in both sisters (figure 1) showed the same aspects, characterised by thickening of the interlobular septa and intralobular interstitium in association with small foci of ground-glass opacity.

WHAT IS YOUR DIAGNOSIS?

See page 442 for the answer to this photo quiz.

Figure 1. High-resolution computed tomography images from two sisters aged 28 (A) and 25 (B) years. The images show similar aspects, with thickening of the interlobular septa and intralobular interstitium. Note also small foci of ground-glass opacities.