HHT classification. Rare cases with ischaemic manifestations have been previously reported, though not well-documented, and usually with coronary artery aneurysm. Our HHT patient had a well-documented myocardial infarction with normal coronary arteries, as shown by our biology data, ECG, TTE, coronarography and RMI results. The ischaemic myocardial lesions might result from our patient’s inherited thrombophilia: factor V Leiden and MTHFR. In 2002, a case of recurrent ischaemic stroke was reported by Undas et al. in a patient with HHT and factor V Leiden antiphospholipid antibodies. However, this association between inherited thrombophilia and myocardial infarction remains the subject of debate. Rare cases of coronary artery aneurysms without stenosis have been previously reported. With angiographically normal arteries, we hypothesize that in our patient, HHT may be complicated by an endothelial dysfunction. This mechanism of endothelial dysfunction should be verified by an acetylcholine test. Nevertheless, in our opinion, inherited thrombophilia and oral contraceptive therapy have probably contributed to the myocardial ischaemic lesions in our patient.

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Patients with bronchiectasis: look for specific causes

Sir,

Over 90% of cases of cystic fibrosis are diagnosed in infancy or childhood, although older patients with phenotypically mild disease are occasionally discovered. We describe a patient with a long history of chronic suppurative lung disease, who was discovered to have cystic fibrosis in her seventh decade.

A 64-year-old non-smoking female Caucasian with radiologically-confirmed bronchiectasis had been regularly followed up for 6 years at our outpatient respiratory clinic. Prior to this, her medical care had been carried out elsewhere. As a child while living in Africa, she reported frequent episodes of ‘chest infections’, and contracted tuberculosis at the age of 21. Years later, the diagnosis of post-tuberculous bronchiectasis was made, based on the clinical history of chronic sputum expectoration and characteristic chest radiograph appearances. Her other past medical history included mild asthma and allergic bronchopulmonary aspergillosis. There was no clinical evidence of exocrine pancreatic insufficiency (normal bowel habit, body mass index 30.1 kg/m²), hepatobiliary disease (normal ultrasonographic appearances), nasal problems or diabetes. There was no family history of chronic lung disease and she had conceived two healthy children—now adults—without difficulty. There were no risk factors for immunodeficiency, and haematological and biochemical laboratory tests (including immunoglobulin levels) were normal. The forced expiratory volume in 1 s was 77% predicted, and forced vital
capacity 83% predicted. During the preceding 5 years, the patient had required 2–3 monthly courses of intravenous anti-pseudomonal chemotherapy administered at home by an indwelling subcutaneous vascular access port. During a routine clinic review, the patient mentioned that her great-nephew had recently been diagnosed with cystic fibrosis (mutations Q43X and D1152H). Subsequent genetic testing of our patient revealed her to be a compound heterozygote for two recognized cystic fibrosis alleles: delta F508 and D1152H.

Delta F508 is the most common mutation worldwide and occurs in up to 80% of cystic fibrosis patients in the UK. D1152H is far less common, having first been identified in Ashkenazi Jews and Northern Europeans, with most data on this mutation arising from studies on infertile males.1 The many different phenotypes observed in cystic fibrosis are believed to relate to the effect of the specific mutation on the production of the cystic fibrosis transmembrane conductance regulator protein. Low values of the protein (class I mutations) are associated with severe disease, and intermediate values (class V) with mild disease.2–5 D1152H is a class IV mutation, and is generally associated with late presentation, mild pulmonary disease, pancreatic sufficiency, normal sweat chloride values and advanced survival.4,5

It is not common for cystic fibrosis to be diagnosed after adolescence, let alone beyond 60 years of age. Although the clinical implication in our patient was limited, her family has received appropriate genetic counselling. Moreover, despite the absence of extra-pulmonary symptoms, advanced age and other plausible causes of bronchiectasis being present (previous ‘chest infections’, tuberculosis and allergic bronchopulmonary aspergillosis), the diagnosis of cystic fibrosis was still made in this particular patient.

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Angioedema may not be a class side-effect of the angiotensin-converting-enzyme inhibitors

Sir,

Angioedema is a well-documented and potentially life-threatening side-effect of treatment with angiotensin-converting-enzyme (ACE) inhibitors, occurring in 0.1–0.2% of patients treated with these drugs.1 Given the growing number of patients with hypertension or heart failure treated with these drugs, and the long duration of treatment, the frequency of this complication is probably set to rise. Although most cases of angioedema occur within the first week of treatment, recent reports indicate that late-onset angioedema may be more prevalent than initially thought. This side-effect of ACE inhibitors is not an allergic reaction and can occur after many years of uneventful drug use.2 Black patients appear to be at increased risk.

We report the case of a 57-year-old Caucasian man, who was admitted to hospital because of severe dyspnoea. Clinical examination revealed intense swelling of the lips and the tongue that did not allow intubation, and emergency tracheotomy was performed to relieve airway obstruction. He had been treated for hypertension with an ACE inhibitor (ramipril 2.5 mg/day), with no side-effects over the last three years. His blood pressure was not well-controlled, however, and a family physician decided to change from ramipril to another ACE inhibitor (trandolapril 2.0 mg/day). Two days later, the patient presented with symptoms of angioedema.

Angioedema is a swelling involving the deeper layers of the skin or submucosal tissue, and usually presents as episodic attacks of swelling of the face, lips, tongue and airways, although it may also