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Idiopathic dilatation of the pulmonary artery in a patient with dermatomyositis complicated by interstitial pneumonitis

Idiopathic dilatation of the pulmonary artery (IDPA) is an uncommon anomaly occurring in 0.6% of patients with congenital heart disease,[1 2] and may be bilateral or unilateral.[3–9] We report on a patient with IDPA who concomitantly developed polymyositis (PM) and interstitial pneumonitis (IP).

On 25 August 1997 a 63 year old woman was referred to our hospital because of dyspnoea, pyrexia, arthralgia, raised levels of aspartate aminotransferase (AST) and lactate dehydrogenase (LDH), and bilateral diffuse shadows on chest *x* ray examination.

On admission, a fine crackling sound was audible in the lung fields. We could not confirm Gottron's sign and heliotrope erythema. Laboratory examination disclosed increases of LDH (1194 IU/l), AST (61 IU/l), creatinine kinase (CK; 2078 IU/l), and aldolase (36.8 IU/l) in the serum samples. Autoantibodies were negative except for antinuclear antibody (1/40). Lymphopenia (600/μl) and accelerated erythrocyte sedimentation rate (ESR; 75 mm/1st h) were seen. The radiological examination showed diffuse granular shadows and dilatation of the pulmonary arteries (PAs) (Fig 1A). Findings of electrocardiography, echo cardiography, and blood gas analysis (BGA) were unremarkable. Pulmonary function was compatible with IP: percentage predicted value for vital capacity was 71.7, for forced expiratory volume per second 110.0, and for carbon monoxide transfer factor was 61.8.

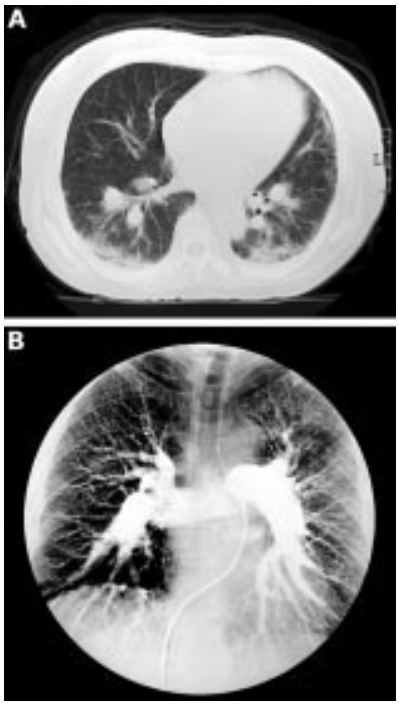

Figure1 Computed tomography findings and pulmonary arteriography.

We performed transbronchial lung biopsy, a right heart catheterisation study, and electromyography. The lung biopsy samples were compatible with IP (fibrosis with Masson body). The catheterisation study disclosed dilatation of the PAs without pulmonary hypertension (PH; mean pulmonary pressure was 20 mm Hg; fig 1B). An electromyograph of the right biceps muscle manifested low amplitude and short duration. We thus diagnosed her as having IDPA complicated with PM related IP.

During such examinations, pyrexia failed to subside, and BGA on 31 August deteriorated (Pa_{O_2} 67.4 mm Hg and Pa_{CO_2} 40.3 mm Hg). Prednisolone (40 mg/day) was started from 17 September. The fever subsided, and the laboratory data improved on the day of discharge (31 October): LDH decreased from 1082 to 738 IU/l, CK from 1885 to 223 IU/l, AST from 60 to 18 IU/l, ESR from 116 to 27 mm/1st h, and white blood cell count from 19 000 to 9600/μl. Data of pulmonary function tests including BGA were unchanged. While taking 30 mg prednisolone a day, she visited crowded stores and thereafter developed pyrexia with severe dyspnoea. BGA showed hypoxaemia (Pa_{O_2} 45.2 mm Hg and Pa_{CO_2} 35.3 mm Hg). Laboratory data on readmission showed LDH 1581 IU/l, CK 169 IU/l, AST 25 IU/l, ESR 122 mm/1st h, C reactive protein 212 mg/l, white blood cell count 15 100/μl, and platelets 60 000/μl. We diagnosed her as having exacerbated IP induced by infection and started treatment with pulse methylprednisolone (1000 mg/day for three days) followed by antibiotics, antimycotics, pentamidine, and immunoglobulin (high titres for cytomegalovirus).

Despite such treatments, she died of respiratory failure on 10 November. Necropsy disclosed (*a*) polymyositis showing focal atrophy, fibrosis, and contraction band necrosis; (*b*) IP with cytomegalic inclusion body; (*c*) dilatation of the bilateral PAs; and (*d*) disseminated intravascular coagulation.

Because dilatation of the PA can be caused by rheumatic diseases through secondary PH, diagnosis of IDPA is difficult among such patients. Our patient had PM associated IP; we thus initially supposed that she had both IP and PH complicated with PM. Pulmonary catheterisation showed the absence of PH, indicating IDPA. This case indicated that IDPA should be ruled out in patients manifesting PH.

Patients with PM can die of acute respiratory failure induced by infection. To prevent respiratory failure, rheumatologists should be careful about opportunistic infections in patients receiving high doses of corticosteroids or immunosuppressive drugs. Because respiratory failure can be fatal, patients with PM should stay away from crowds while they are taking high doses of immunosuppressive agents.

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Arthritis associated with non-alcoholic steatohepatitis

Non-alcoholic steatohepatitis (NASH) is an entity characterised by mild abnormal liver function tests, a negative serological investigation, and histopathological findings similar to those seen in alcohol induced liver disease in patients who do not consume significant amounts of alcohol. Patients are usually asymptomatic but may complain of fatigue, malaise, or vague right upper quadrant pain. A low calorie diet is helpful in most of these patients. We present a case report of a patient with arthritis associated with NASH, which resolved completely after the introduction of a low calorie diet.

A 37 year old man presented with symmetrical arthritis during the past year affecting the elbows, wrists, shoulders, and knees. He had morning stiffness, which resolved after a one hour period of activity. On physical examination there was moderate tenderness of the elbows and wrists without swelling or limitation of the range of motion. Liver and spleen were not enlarged. Laboratory studies showed increased liver enzymes; aspartate aminotransferase 53 IU (normal <49 IU), alanine aminotransferase 98 IU (normal <51 IU), glutamyl transpeptidase 37 IU (normal <34 IU), alkaline phosphatase 64 IU (normal <131 IU), and total bilirubin 16 μmol/l. Serum cholesterol was 6.00 mmol/l, triglycerides 1.62 mmol/l, total protein 77 g/l, and albumin 50 g/l.

Sedimentation rate was 20 mm/1st h, C reactive protein was normal, rheumatoid factor, antinuclear antibodies (ANA), hepatitis B surface antigen, and antibodies to hepatitis C virus (HCV), smooth muscle, and mitochondria were all negative. He had IgG

antibodies to Epstein-Barr virus, and cytomegalovirus. An *x* ray examination of the hands and knees was negative. An ultrasound of the liver and spleen was normal.

His primary physician reported that the abnormal liver function tests had been present for approximately two years. A liver biopsy showed prominent macrovesicular fatty changes, aggregates of parenchymal inflammation, and mild fibrosis with minimal inflammatory infiltrate at some of the portal spaces. Fibrosis and Mallory bodies were not seen at the central vein areas. Iron staining was negative. These findings are consistent with a diagnosis of NASH.

The patient started on a low calorie diet and a few weeks later his liver function tests normalised with complete resolution of his joint symptoms. A few months later he stopped following the low calorie diet and within one week he began to have joint symptoms and the liver enzymes were found to be raised. Reintroduction of the low calorie diet again resulted in complete resolution of his symptoms and abnormal liver function tests.

Polyarthralgia or polyarthritis is one of the extrahepatic manifestations of liver diseases, and many chronic liver diseases are associated with rheumatic diseases. Examples include the association between chronic infection with hepatitis B virus and polyarteritis nodosa,[1] the association between chronic infection with HCV and mixed cryoglobulinaemia,[2] the association between chronic infection with HCV and Sjögren's syndrome, and the association of primary biliary cirrhosis and scleroderma.

Immune complexes may be one of the mechanisms of arthralgia associated with liver diseases, and patients with chronic liver disease due to HCV[3] infection have been shown to have an increased prevalence of ANA, rheumatoid factor, and anti-smooth muscle antibodies.

Increasing evidence shows that several cytokines mediate hepatic inflammation and cholestasis in alcoholic and non-alcoholic steatohepatitis. Among these cytokines tumour necrosis factor α (TNFα) is a key factor.[4] TNFα mediates not only the early stages of fatty liver but also the transition to more advanced stages of liver diseases like steatohepatitis and cirrhosis. On the other hand, TNFα is an important inflammatory disease mediator in a wide spectrum of articular diseases, and inhibition of this cytokine led to a significant improvement of symptoms and signs in rheumatoid arthritis (RA).[5] So TNFα may be a common mediator in NASH and arthritis.

Liver disease, however, may occur owing to connective tissue diseases which cause the joint symptoms, as has been found in patients with RA,[6] Felty's syndrome, Still's disease, systemic lupus, giant cell arteritis, polyarteritis nodosa, and polymyalgia rheumatica. Histological liver changes are usually non-specific. Our patient, however, had no evidence of RA or other rheumatic diseases.

There are numerous reports connecting the ingestion of certain food items and arthritis,[7] with symptomatic improvement after the exclusion of certain types of food from the diet (elimination treatment). Dietary supplementation has resulted in mixed results in patients with RA, with no response to the addition of vitamin C, or zinc, but clinical benefits of diets enriched with fish oil[8] or plant oils. Complete remission has been reported with a diet which included chicken type II collagen.[9] However, low calorie diets have not been shown to reduce joint symptoms in patients with collagen vascular diseases.

We are unaware of previous publications showing an association between NASH and arthralgia or arthritis, and in fact joint symptoms were not reported in two large series of patient with NASH.[10 11] Still the unique response on two occasions of our patient's symptoms and abnormal liver function tests to a low calorie diet supports our hypothesis that the joint symptoms were associated with the NASH.

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