Fever, rash, swollen joints, and sore throat in a young man

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Answers on p 211.

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An 18 year old man developed a sore throat followed within two days by pain and swelling of both wrists, knees, and ankles, the metacarpophalangeal joints of both hands, and the metatarsophalangeal joints of both feet. This was associated with erythematous rash over his hands, arms, chest, and thighs down to both knees (see figs 1 and 2). His past medical history was otherwise unremarkable.

On examination he was febrile with a temperature of 38.6°C orally. He had an erythematous rash over the dorsal aspects of both hands, both arms, both thighs down to the knees, and chest. There were small effusions in both knees. Both wrists, both ankles, the metacarpophalangeal joints of both hands, and the metatarsophalangeal joints of both feet were tender and slightly swollen. Examination of the rest of his peripheral joints and of his axial skeleton was normal. Abdominal examination showed moderate splenomegaly, but nothing else. Other systemic examinations were unremarkable.

Figure 1 Erythematous rash on hands.

Figure 2 Erythematous rash on arm.

Investigations on admission included:

- Haemoglobin = 119 g/l
- White cell count = $23.0 \times 10^9/l$ (neutrophils $21.6 \times 10^9/l$)
- Platelets = $338 \times 10^9/l$
- Erythrocyte sedimentation rate = 92 mm in one hour
- C reactive protein = 234 mg/l (normal 0–10 mg/l)
- Serum total bilirubin = 16 $\mu\text{mol/l}$
- Serum alanine transaminase = 166 IU/l
- Serum alkaline phosphatase = 465 IU/l
- Serum albumin = 33 g/l
- Antistreptolysin-O titres = normal
- Serum ferritin = 22 515 $\mu\text{g/l}$ (normal 40–350)
- Blood cultures and viral titres = negative
- Latex and antinuclear antibodies = negative
- White cell scan = normal
- Bone scan = normal

Ultrasound scan of the abdomen confirmed a modest degree of splenomegaly and nothing else.

Question

- (1) What is the most probable diagnosis?

A rare cause of respiratory failure

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Answers p 211.

A 68 year old man with a 60 year pack history (20 cigarettes per day for 60 years) of smoking was admitted via casualty with acute confusion, lethargy, and dyspnoea over three days. He denied any cough or haemoptysis, but was unable to give a clear history. Information from his wife indicated increasing exertional dyspnoea over two years with recent swelling of the ankles. She described increased spinal curvature over the past year with weakness of his arms and legs. On clinical examination he had marked kyphosis and was afebrile. Pulse was 100 beats/min with a blood pressure of 140/70 mm Hg. Heart sounds were normal. His respiratory rate was 14 beats/min with poor chest expansion and decreased breath sounds bilaterally. He was confused and slightly drowsy with a Glasgow coma scale of 14/15. Cranial nerves were intact. Upper limb examination was normal with no evidence of muscle wasting or fasciculation. Lower limb examination showed right foot drop. Full blood count and electrolytes were normal. Arterial blood gases on admission on 24% oxygen showed pH 7.40, arterial carbon dioxide tension 9.8 kPa, arterial oxygen tension 8.1 kPa, bicarbonate 44 mmol/l, and a base excess of +15. The chest radiograph is shown in fig 1.

Six weeks before this admission he had been seen in a respiratory clinic for investigation of his dyspnoea but no mention of muscular weakness was made. His past medical history included a neck injury after a road traffic accident 14 years previously with no associated neurological deficit. Chest examination at that time revealed kyphosis with markedly decreased chest expansion with reduced air entry.

Figure 1 Chest radiograph.

Figure 2 Cervical radiograph.

Cardiovascular examination was normal. Detailed neurological examination was not recorded. Full lung function tests showed peak flow 320 l/m (predicted 505 l/m). Forced expiratory volume in one second was 2.2 l/sec (predicted 2.65 l/sec). Forced vital capacity was 2.4 litres (predicted 3.65 litres), and transfer factor of carbon monoxide per unit of volume was 19.4 ml/min/mm Hg (predicted 17.6). Cervical radiography is shown in fig 2. Arrangements were made to review him after further investigation.

Questions

- (1) How would you interpret the arterial blood gases and the lung function tests?
- (2) What is the differential diagnosis?
- (3) What do the chest and cervical radiographs show?
- (4) What further investigations might be helpful?
- (5) What would be your initial management of this patient?
- (6) What is the long term respiratory management of this condition?

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