Competing interest: None.

Correspondence to: Dr Z G Li, heijing_1105@yahoo.com

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REFERENCES

- 1 Haneji N, Nakamura T, Takio K, Yanagi K, Higashiyama H, Hayashi Y. Identification of alpha-fodrin as a candidate autoantigen in primary Sjögren's syndrome. *Science* 1997;**276**:604–6.
- 2 Maeno N, Takei S, Imanaka H, Oda H, Yanagi K, Hayashi Y, et al. Anti-alpha-fodrin antibodies in Sjögren's syndrome in children. *J Rheumatol* 2001;**28**:860–4.
- 3 Witte T, Matthias T, Oppermann M, Helmke K, Tishler M, Schmidt RE, et al. Prevalence of antibodies against alpha-fodrin in Sjögren's syndrome: comparison of 2 sets of classification criteria. *J Rheumatol* 2003;**30**:2157–9.
- 4 Vitali C, Bombardieri S, Jonsson R, Alexander EL and the European Study Group on Classification Criteria for Sjögren's syndrome. Classification criteria for Sjögren's syndrome: a revised version of the European criteria proposed by the American-European Group. *Ann Rheum Dis* 2002;**61**:554–8.
- 5 Yanagi K, Ishimaru N, Haneji N, Saegusa K, Saito I, Hayashi Y. Anti-120-Kd alpha-fodrin immune response with Th1-cytokine profile in the NOD mouse model of Sjögren's syndrome. *Eur J Immunol* 1998;**28**:3336–45.
- 6 Nagraju K, Cox A, Casciola-Rosen L, Rosen A. Novel fragments muscarinic acetylcholine receptor generated during cytotoxic lymphocyte granule-induced cell-death. *Arthritis Rheum* 2001;**44**:2376–86.

"Bubbles in the brain": an unusual complication of dermatomyositis

C Iking-Konert, B Ostendorf, G Jung, A Becker, M Schneider

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Dermatomyositis (DM) is an inflammatory microangiopathy, affecting particularly the skin and muscles. The incidence ranges from 0.2 to 1.0/100 000; an overlap with other connective tissue diseases such as systemic sclerosis has been described, as has an association with cancer. Clinically DM is identified by a characteristic rash and a varying muscle weakness. The clinical diagnosis is confirmed by typical muscle histology.¹

A 60 year old man was admitted to our department with a 2 week history of muscle weakness, myalgias, dysphagia, a heliotrope rash on the eye lids, and bilateral acral finger ulcers. Creatine kinase (3879 U/ml, normal <171 U/ml) and C reactive protein (170 mg/l; normal <5 mg/l) were raised; autoantibodies (antinuclear antibodies (ANA), extractable nuclear antigen (ENA), anti-synthetase antibodies, rheumatoid factor, antineutrophil cytoplasmic antibodies (ANCA)) were negative. A muscle biopsy showed perivascular inflammation, an infiltrate predominantly of CD4+ T cells, and perifascicular atrophy, confirming the suspected diagnosis of DM. Despite the advanced age of the patient, the rapid onset of the manifestations, and the lack of autoantibodies, cancer associated myositis could be ruled out. Consequently, corticosteroid treatment (1 mg/kg body weight (b. wt)) was started.

The patient developed a fever, and a chest x ray examination showed a skin emphysema and a pneumomediastinum. Computed tomography (CT), oesophagoendoscopy, and bronchoscopy disclosed no cause of the pneumomediastinum. In particular, again no gastrointestinal or intrapulmonary tumour, pneumothorax or fistula was found. The patient then complained of dizziness, position dependent headache, and neck stiffness; after a syncope his mental state was altered. CT ruled out a cerebral infarction or bleeding, but showed intracranial air inclusions (fig 1). No signs of a fracture were seen at the base of the skull, but magnetic resonance imaging (MRI) of the neck showed a spondylodiscitis with a paravertebral inflammation between cervical spine C5–7 (fig 2A). CT performed after swallowing a contrast agent detected a fistula containing air from the hypopharynx into the paravertebral region (fig 2B). To obtain a detailed evaluation an inflexible endoscopy was performed,

showing two ulcers and a covered perforation of the upper oesophagus.

We assumed that disease activity was uncontrolled and the oesophagus was affected, and higher doses of corticosteroids (2 mg/kg b. wt) and immunoglobulins (1 g/kg b. wt) were given. Because of the spondylodiscitis and the proof of bacteria in the cerebrospinal fluid a further immunosuppression was postponed and intravenous antibiotic treatment was started. The patient was discharged weeks later without any neurological sequelae. Two years later he died from fatal

Figure 1 A CT scan of the head shows the intracranial air inclusions preferentially in the lateral ventricles (black arrows), but also disseminated in the subarachnoid space (white arrows).

Figure 2 (A) MRI using a contrast enhanced, T₁ weighted, spin echo sequence shows a spondylodiscitis at the level C5/6 and C6/7 (arrow A). Furthermore, there is a prevertebral inflammatory soft tissue enhancement (arrow B) in the region of the fistula seen on CT and an epidural inflammation at the level of the spondylodiscitis (arrow C). (B) A CT scan of the neck after a water soluble contrast agent (Solustrast; Schering, Berlin, Germany) has been swallowed depicts the fistula from the hypopharynx (black arrow A) into the paravertebral tissue (black arrow B) due to the oesophagus perforation. Air inclusions are also seen in the paravertebral space (white arrow).

tachyarrhythmia, probably due to a myocardial involvement of the disease.

As far as we know, this is the first case, reporting a pneumoencephalon in a patient with DM, probably caused by a trespass of air from the paravertebral inflammation into the cerebrospinal fluid system. Although the soft tissue inflammation and the spondylodiscitis could be explained by the perforation of the oesophagus, the direct leakage of air into the cerebrospinal fluid system itself could not be depicted by CT. Nevertheless, the discovery that bacteria were present in the cerebrospinal fluid in addition to the air inclusions clearly argues for a subarachnoid cervical fistula. The development of a pneumoencephalon through such fistulas has been described before—for example, in patients with spinal fractures² or after routine thoracotomy.³

The involvement of the striated muscles of the oropharynx and upper oesophagus in patients with DM is a common clinical manifestation (15–75%^{4,5}) and was obvious in our patient, who complained of dysphagia. A perforation or a fistula as a severe complication of an oesophagus involvement has so far only been reported in single cases in DM or in other connective tissue diseases such as CREST syndrome or scleroderma.^{6–8} A pneumomediastinum in DM has also been described previously.⁹ It has been suggested that it may be associated with a vasculopathy,⁹ pretreatment with corticosteroids or a pre-existing lung disease,¹⁰ but its precise aetiopathogenesis remains unclear. Comorbidity of a pneumomediastinum and a perforation of the oropharynx/upper oesophagus, however, has not previously been described.

In conclusion, while the involvement of the oropharynx/oesophagus in DM is quite common, a perforation of the oesophagus is a rare, but potentially life-threatening complication; physicians should be aware of it, especially when patients complain about dysphagia. In the case of a pneumomediastinum, a perforation of the upper oesophagus should be considered and ruled out by an inflexible endoscopy, because it might be missed by conventional endoscopy.

Authors' affiliations

C Iking-Konert, B Ostendorf, M Schneider, Department of Endocrinology, Diabetology and Rheumatology, Heinrich-Heine-University Duesseldorf, Germany

G Jung, Department of Diagnostic Radiology, Heinrich-Heine-University Duesseldorf, Germany

A Becker, Medizinische Klinik II, Department of Rheumatology, Klinikum Bremerhaven, Germany

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The patient's consent and permission to publish are not available (the patient died).

Correspondence to: Dr C Iking-Konert, Department of Endocrinology, Diabetology and Rheumatology, Heinrich-Heine-University Duesseldorf, Moorenstr 5, 40225 Düsseldorf, Germany; Iking-Konert@med.uni-duesseldorf.de

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REFERENCES

- 1 Dalakas MC, Hohlfield R. Polymyositis and dermatomyositis. *Lancet* 2003;**362**:971–82.
- 2 Uemura K, Meguro K, Matsumura A. Pneumocephalus associated with fracture of thoracic spine: case report. *Br J Neurosurg* 1997;**11**:253–6.
- 3 Brown WM 3rd, Symbas PN. Pneumocephalus complicating routine thoracotomy: symptoms, diagnosis and management. *Ann Thorac Surg* 1995;**59**:234–6.
- 4 de Merieux P, Verity MA, Clements PJ, Paulus HE. Esophageal abnormalities and dysphagia in polymyositis and dermatomyositis. *Arthritis Rheum* 1983;**26**:961–8.
- 5 Ertekin C, Secil Y, Yuceyar N, Aydgdu I. Oropharyngeal dysphagia in polymyositis/dermatomyositis. *Clin Neurol Neurosurg* 2004;**107**:32–7.
- 6 Dougenis D, Papathanasopoulos PG, Paschalis C, Papapetropoulos T. Spontaneous esophageal rupture in adult dermatomyositis. *Eur J Cardiothorac Surg* 1996;**10**:1021–3.
- 7 Thompson JW. Spontaneous perforation of the esophagus as a manifestation of dermatomyositis. *Ann Otol Rhinol Laryngol* 1984;**93**:464–7.
- 8 Lambert DR, Ilaneza PP, Gaglani RD, Lach RD, Beaver WL. Esophageal-atrial fistula. *J Clin Gastroenterol* 1987;**9**:345–9.
- 9 Barvaux VA, Van Mullem X, Pieters TH, Houssiau FA. Persistent pneumomediastinum and dermatomyositis: a case report and review of the literature. *Clin Rheumatol* 2001;**20**:359–61.
- 10 Kono H, Inokuma S, Nakayama H, Suzuki M. Pneumomediastinum in dermatomyositis: association with cutaneous vasculopathy. *Ann Rheum Dis* 2000;**59**:372–6.