



CASE REPORT

Improved ventilatory function associated with relief of chest pain in a patient with Kartagener's syndrome treated by subcutaneous analgesia

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Introduction

Kartagener's syndrome was defined in 1933 as a distinctive clinico-pathological entity characterized by a triad of signs: situs inversus, bronchiectasis and paranasal sinusitis. The condition is a part of the syndrome of primary ciliary dyskinesia, where 50% of the patients have a situs inversus totalis. In primary ciliary dyskinesia, inflammation is not restricted to the ciliated conducting airways, but also occurs in the alveolar space.¹

Repetitive episodes of pleuritic chest pain are a feature of patients with bronchiectasis and middle lobe syndrome.^{2–4} Pleuritic chest pain can occur in up to 21% of patients with bronchiectasis and 19% of patients with middle lobe syndrome and characteristically occurs in an area overlying a bronchiectatic lobe.^{2,3} This presentation can be confused with other conditions, such as pulmonary embolus.⁴ However, the natural history of pleuritic chest pain and its response to analgesia in these conditions is poorly described. We report a patient with Kartagener's syndrome with intractable pleuritic chest pain, in whom peripheral nerve stimulation with subcutaneous electrodes led to a dramatic therapeutic response.

Case report

A 37-year-old female was admitted for assessment of intractable pleuritic chest pain. Kartagener's syndrome was diagnosed at age 13 years when she presented with recurrent respiratory tract infections. The patient had pansinusitis, situs inversus, bronchiectasis and primary infertility, but had successfully conceived with the aid of in vitro fertilisation. She had an appendectomy at 15 years of age and a cholecystectomy at 34 years, both left-sided. She had suffered from recurrent respiratory tract infections and chronic pleuritic left-sided chest pain since 31 years of age which could not be controlled on combinations of non-steroidal anti-inflammatory drugs and opiates. The pain was constant and was located in the left anterior chest wall, prevented full inspiration, and was exacerbated by intercurrent respiratory tract infections and cough. At the time of admission, the patient was depressed and anxious because of the pain, and felt that her pain was severely interfering with her quality of life.

Physical examination revealed crackles at the left base, wheeze and healed left-sided cholecystectomy and appendectomy incisions. Chest radiograph and high resolution computerised tomography of thorax showed dextrocardia and left middle lobe (morphologically a right middle lobe) bronchiectasis and atelectasis (Fig. 1). Fibreoptic

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bronchoscopy showed transposed bronchial anatomy with inflamed left middle and right lower lobes containing purulent secretions. Bronchial washings cultured *Haemophilus influenzae*, which was sensitive to broad spectrum antibiotics. A pair of temporary subcutaneous quadrode electrodes (Advanced Neuromodulation Systems, Texas, USA) was inserted under anaesthesia with fentanyl 150 mg, propofol 30 mg and midazolam 3 mg intravenously. Electrodes were inserted at the point of maximum hyperalgesia, as determined by cotton wool and pin-prick testing (midaxillary (T_{4,5}) and midclavicular areas (T_{6,7})). The patient experienced immediate analgesia. Pulmonary function testing demonstrated an immediate and sustained rise in forced expiratory volume in 1 s (FEV₁) from 1.3 to 2.12 l (63% increase) and forced vital capacity (FVC) from 1.89 to 3.08 l (62% increase; Fig. 2). In view of

the clinical response, the temporary device was removed 7 days after insertion and a set of quadrode electrodes (ANS) were inserted and connected to an internal power source (Genesis, ANS) under general anaesthetic. Chest radiograph showed improvement in the atelectatic changes in the left middle lobe. She remained pain free after the procedure, declined further analgesics, and spirometry values have remained significantly improved at 2 weeks and 2 months follow-up (Fig. 2).

Discussion

This phenotype of primary ciliary dyskinesia results from axonemal abnormalities of respiratory cilia and sperm flagella, mainly an absence of dynein arms, and is inherited as an autosomal recessive disorder.⁵ Diagnosis of primary ciliary dyskinesia is clinical and is confirmed by studies of ciliary motility and ultrastructure. Radiologically, the disease progresses from bronchial wall thickening to parenchymal changes including atelectasis, consolidation, and bronchiectasis. There is a predilection for anatomic middle lobe abnormalities. The radiological appearance and clinical state have similarities to cystic fibrosis, although they are less severe and less progressive. Pulmonary function testing most commonly reveals mild airflow obstruction.⁶ Management is directed to microbial suppression and clearing of retained secretions. The natural history of intractable pleuritic chest pain in primary ciliary dyskinesia is unknown.

Thoracic paravertebral block has been successfully used in the management of intractable

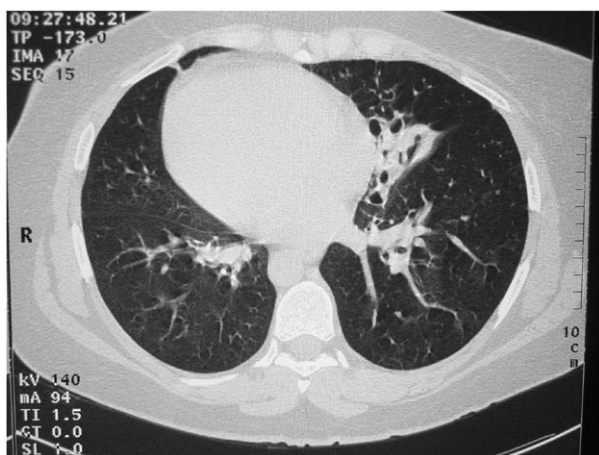


Figure 1 Dextrocardia with left middle lobe bronchiectasis.

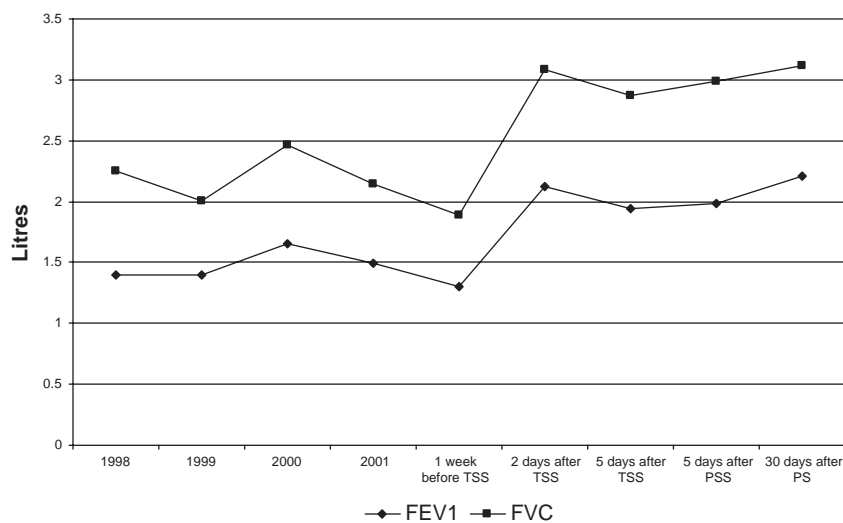


Figure 2 The rise in FEV₁ and FVC is demonstrated after insertion of the temporary subcutaneous stimulator (TSS) device which is sustained after its replacement with a permanent device (PSS) 30 days later.

pleuritic chest pain.⁷ Electrical nerve stimulation is a diagnostic procedure, which involves stimulation of peripheral nerves by a needle electrode inserted through the skin. The nerve stimulation does not prevent pain but only alleviates pain as it occurs. The procedure is used to treat chronic, intractable pain conditions such as intractable angina.⁸ Normally, this lead is implanted in the epidural space but this was relatively contraindicated in this patient because of the risk of epidural abscess. Electrical nerve stimulation is an accepted modality for assessing suitability for ongoing treatment with an implanted nerve stimulator. The physician should be able to determine whether the patient is likely to derive a significant therapeutic benefit from continuing use of an implanted nerve stimulator within a trial period of one month. If pain is effectively controlled by percutaneous stimulation, implantation of electrodes is warranted.

Chronic chest pain is a common problem in patients with bronchiectasis, cystic fibrosis, and middle lobe syndrome. Analgesics may lead to serious and difficult to manage side effects in these patients.^{4,9} Few interventions have a significant impact on quality of life or lung function in patients with most pulmonary disorders. Although this technique unmasked the patient's true lung function, rather than improving a decline in lung function, the clinical result was the same. This procedure, used in a patient with Kartagener's syndrome and chronic pain due to left middle lobe

bronchiectasis, improved both lung function and quality of life. The authors believe that the technique should be considered as a therapeutic option in patients with chronic pleuritic chest pain.

References

1. Skold CM, Barck C, Johard U, Dahl M, Eklund A. The immotile cilia syndrome: characterization of the inflammatory response in nonsmoking patients with dysfunction of the cilia. *Respiration* 1996;**63**:325–32.
2. Munro NC, Currie DC, Garbett ND, Cole PJ. Chest pain in chronic sputum production: a neglected symptom. *Respir Med* 1989;**83**:339–41.
3. Kwon KY, Myers JL, Swensen SJ, Colby TV. Middle lobe syndrome: a clinicopathological study of 21 patients. *Hum Pathol* 1995;**26**:302–7.
4. Owens GP, Rogers RM. Bronchiectasis masquerading as pulmonary embolus. *Am J Med* 1985;**79**:667–9.
5. Pennarun G, Chapelin C, Escudier E, et al. The human dynein intermediate chain 2 gene (DNAI2): cloning, mapping, expression pattern, and evaluation as a candidate for primary ciliary dyskinesia. *Hum Genet* 2000;**107**:642–9.
6. Greenstone M, Rutman A, Dewar A, Mackay I, Cole PJ. Primary ciliary dyskinesia: cytological and clinical features. *Q J Med* 1988;**67**:405–23.
7. Paniagua P, Catala E, Villar Landeira JM. Successful management of pleuritic pain with thoracic paravertebral block. *Reg Anesth Pain Med* 2000;**25**:651–3.
8. Moore R, Chester M. Neuromodulation for chronic refractory angina. *Br Med Bull* 2001;**59**:269–78.
9. Ravilly S, Robinson W, Suresh S, Wohl ME, Berde CB. Chronic pain in cystic fibrosis. *Pediatrics* 1996;**98**:741–7.