BMJ

BMJ 2012;344:e1181 doi: 10.1136/bmj.e1181 (Published 15 March 2012)

PRACTICE

A PATIENT'S JOURNEY

Polymyositis

A formerly active man who developed polymyositis at age 47 explains how this rare and debilitating autoimmune disorder has affected his life

Paul Truepenny *patient*[1], Viswanath Kaushik *consultant rheumatologist*[2], Heidi Lempp *medical sociologist*[3]

[1]Spilsby, UK; [2]Department of Rheumatology, Lincoln County Hospital, Lincoln LN2 5QY, UK; [3]Department of Academic Rheumatology, King's College London School of Medicine, London, UK

This is one of a series of occasional articles by patients about their experiences that offer lessons to doctors. The *BMJ* welcomes contributions to the series. Please contact Peter Lapsley (plapsley@bmj.com) for guidance.

Life before diagnosis

About two years before my diagnosis in 2006 I became progressively more tired. I was a farm manager, a physical, outdoor job, and was used to working long hours during busy periods. I began to find manual tasks increasingly tiring and found myself needing more sleep and waking up feeling less refreshed. This in turn affected my concentration and ability to plan my days effectively. Also, I would often come home and fall asleep before our evening meal, be woken by my wife to join the family for supper, and then either fall asleep in my chair again or go straight to bed. I found it increasingly difficult to have a social life, and, if I did make the effort, I was often too exhausted to enjoy it.

I had no pain and just put the symptoms down to ageing; I was 47 at the time. In July 2006 things came to a head: my work became too difficult to manage and my family life was reduced to saying "hello" and "good night." One night I came home with barely the energy to enter the house. I then realised that something must be seriously wrong. I told my wife I could not continue like this. She made an appointment for me with my general practitioner the following day.

My diagnosis, initial treatment and impact on my life

The GP ordered blood tests, which came back showing only a high creatine phosphokinase (CPK) level. This indicated that I might have had a heart attack, which did not fit with the rest of my symptoms. The doctor repeated the tests the next day and found that my CPK was even higher. I was admitted to the local hospital for further blood tests and an electrocardiogram. They were all inconclusive apart from the raised CPK. As my family has a history of rheumatoid arthritis, I was referred to a consultant rheumatologist.

Luckily at this time I had private health insurance with my job, and obtained an appointment the following week. Within the first few minutes of the appointment Dr Kaushik relayed his suspicions that I might have polymyositis. A muscle biopsy confirmed the diagnosis. At this time I was unable to work. Dr Kaushik prescribed a high dose of steroids to try to control the inflammation and I felt some improvement. However, with gradual reduction of the dose of steroids to a long term sustainable level, the tiredness returned. I was now also starting to feel more muscle fatigue and severe, sudden, cramp like pains that limited my mobility.

My family history

Predisposition to polymyositis and rheumatoid arthritis are thought to be hereditary. Environmental factors, such as shock or viral infection, are thought to act as triggers that cause the immune system to attack the body. It may be the same gene that causes both conditions and affects other factors that determine what is attacked by the auto-immune system. My father received a diagnosis of rheumatoid arthritis at age 32 and was severely physically affected by the condition all his life. My sister was diagnosed as having rheumatoid arthritis at 26 and her quality of life is severely restricted. In addition, both my daughters were diagnosed with juvenile arthritis as teenagers. The elder is now 23 years old and in full remission. The younger is 19 and takes methotrexate, which allows her to lead a normal life.

My early retirement

In 2007, after six months off work and on a high dose of steroids, starting at 40 mg and reducing slowly, I tried a gradual

Correspondence to: V Kaushik Viswanath.Kaushik@ulh.nhs.uk

return to my job. I did mainly office work, initially for two to three hours a day. I steadily increased this to eight hours of light work. However, as soon as I reduced the dose of steroids to 10 mg the fatigue increased and limited me to moving slowly around the house.

Dr Kaushik and I developed a shared care arrangement between King's College Hospital in London, my local hospital, and my GP. This works well. I see Dr Kaushik every six months, although this appointment is often delayed by long waiting lists. If I have a flare up and need advice, I contact Dr Kaushik by phone or email and he fits me into his outpatient clinic, often on the same day. My visits to London vary from two-monthly to six-monthly, depending on my condition, potential changes in drugs, or the need to re-assess dosage.

My wife always accompanies me to my appointments. This is very important for both of us. When I am in pain, I find it difficult to concentrate, which can lead to confusion and misunderstanding that my wife can help clarify. She is the first person with whom I discuss illness related issues and she can then raise them during our consultations. The trips to London are not easy. They require early starts, an hour in the car to the local station, one hour on the train, and 30 to 50 minutes to get across London by bus.

In 2007, 12 months after my first visit to my GP and discussions with my family and my two consultants, I decided to retire on medical grounds. This was a very difficult decision as I had worked for the company for over 25 years. The house we lived in came with the job, so my retirement caused a big family upheaval. The overriding factors, after returning to work, were the pain, fatigue, and mental pressure it put me under. I was in no doubt that these would have affected my long term health and family life. Looking back, we took the right decision; it has allowed me to rest and make best use of my life with polymyositis.

My life after retirement

During the past four years my health has been very unpredictable. It was difficult to find the most effective drugs and dosages to control the disease and, just as importantly, to work out what my body can tolerate. Taking high doses of steroids for a year has resulted in a weight increase from 95 kg to a peak of 132 kg. This has made simple things difficult; for example, bending down to put on socks and shoes, sitting up from lying down, standing up from sitting and standing a short time. I am trying to lose weight but there is a limit to how little one can eat.

Over time I began to realise that with this unpredictable, chronic condition I have had to change my lifestyle. There are periods, sometimes days, sometimes weeks, when I feel well, my pain levels are low, and my fatigue is manageable; however, I have to be careful not to overdo things, or the pain and fatigue soon return. My main goal each day is to find a balance, to achieve as much as I can without overdoing it. During, what I call my "uphill with the hand brake on" days, everything is hard work. For example, simple activities such as showering and dressing can take up to an hour. I am determined not to spend the day in front of the television, but on the worst days that may be my limit. When "the hand brake is off" and the incline of the hill seems less, I try to keep myself busy. I enjoy outdoor photography and creating greetings cards for family and friends.

It is rewarding to find ways to make daily activities easier for myself. For example, a monopod takes the weight of my camera and helps keep it steady, and a walker with a seat allows me to rest. If I go shopping, I can sit down before I get too tired, which enables me to pace myself and do more. If we go out with friends on an "uphill" day, I take my wheelchair and they take turns in pushing me.

The support and understanding of my family are important to me. My wife is a rock and my daughters are great. They often take the brunt of my frustrations when pain levels are high and I become irritable. We have a great bunch of friends around us who always want to know how I am, involve me in their plans, and are aware of my limitations.

My advice to healthcare professionals

Six years on, I have developed a new approach to dealing with healthcare professionals: I now plan what my wife and I want to say when we go to our appointments. I have learnt that the more accurate the information I give medical and nursing staff the better they can understand and respond to my condition. I am not afraid to ask questions or ask for clarification. I now expect my consultations to be within an equal partnership. My aim is to help healthcare staff to understand me as a person and how this rare disease affects my life so that I can receive the best treatment and care available.

Competing interests: All authors declare no support from any organisation for the submitted work; no financial relationships with any organisations that might have an interest in the submitted work in the previous three years; no other relationships or activities that could appear to have influenced the submitted work.

Provenance and peer review: Not commissioned, not externally peer reviewed.

1 Oddis CV, Conte CG, Steen VD, Medsger TA Jr. Incidence of polymyositis-dermatomyositis: a 20-year study of hospital diagnosed cases in Allegheny County, PA 1963-1982. *J Rheumatol* 1990;17:1329-34.

Accepted: 25 November 2011

Cite this as: *BMJ* 2012;344:e1181

A clinician's perspective

Polymyositis is an autoimmune condition characterised by inflammation in the muscles. It is rare, with an incidence of around five to 10 cases per million.[1] It most often presents between ages 30 and 60, with women being more affected than men in a ratio of 2:1. The usual presenting feature is weakness in the muscles. Initially the thigh and arm muscles are affected, leading to difficulties in rising from a low chair, climbing steps, lifting objects, and combing hair. Fatigue, muscle aches, and cramps may also be present. The symptoms can fluctuate, with good days and bad days.

The commonest finding on clinical examination is weakness in the larger muscle groups such as the quadriceps, with fairly preserved strength in small muscles such as those in the hands. Around a third of patients may have a rash, which can include a blue-purple discolouration on the upper eyelids, a flat red rash involving the face and upper trunk, or raised purple-red scaly patches over the knuckles. A rash suggests dermatomyositis.

The diagnosis is based on raised concentrations of muscle enzymes—such as creatine kinase, lactate dehydrogenase, transaminases, and aldolase—and characteristic mysositic changes on electromyogram. The diagnosis can be confirmed by a biopsy from an affected muscle that shows changes consistent with active inflammation. People with polymyositis have a 1.5-fold increase in the incidence of malignancy, compared with the unaffected population, and this problem is usually seen in the first two years of the diagnosis. Further investigations, such as computed tomography, may be needed to exclude a malignant condition.

Initial treatment of polymyositis is with steroids to control the inflammation in the muscles. This needs to be started as soon as the diagnosis is confirmed because "time is muscle"; the longer the delay in starting treatment, the lesser the chance of recovery. A high dose of steroid is recommended, calculated as 1-2 mg per kg of the patient's body weight. Later, patients are started on further treatments such as methotrexate or cyclosporine. In people with recalcitrant myositis, treatments such as intravenous immunoglobulins may be needed.

Diagnosis of polymyositis can be difficult and not everyone will present with classic symptoms. This was the case with Mr Truepenny. He had symptoms for longer than usual before presentation and his CPK was only moderately raised. However, his family history of various autoimmune conditions was compelling. My first priority was to arrange an urgent electromyogram and muscle biopsy before starting treatment. When the muscle biopsy came back conclusive I started him on high dose steroids. Recovery was slower than expected. Because this condition is rare, I decided to refer Mr Truepenny to a neuromuscular centre at Kings College Hospital for a second opinion. They agreed with the diagnosis and since then Mr Truepenny has been managed between the two hospitals.

Viswanath Kaushik

Learning points

- Subtle features need to be recognised in the history because the presentation is not always textbook
- Organise a muscle biopsy to confirm the diagnosis
- Have a low threshold for referring to specialist centres for another opinion

Useful resources

Myositis Support Group UK (www.myositis.org.uk)—UK charity specifically for the inflammatory myopathies: dermatomyositis, polymyositis, inclusion body myositis, and juvenile dermatomyositis. It aims to provide information to patients and their families, to help give them a better understanding of their illness, to relieve the isolation felt by an individual with a rare illness, to guide patients in the right direction for treatment, to raise awareness of the conditions, and to raise funds to promote and support research

The Myositis Association (www.myositis.org)—is an organisation in the USA that provides information, support, advocacy, and research for those concerned about myositis

The Cure JM Foundation (www.curejm.com)—was established in October 2003 with the goal of raising awareness of this rare disease and funding research to find a cure for juvenile myositis