SHORT REPORTS

Myeloma presenting with Pancoast's syndrome

Skeletal pain is the commonest initial manifestation of multiple myeloma. We report an unusual presentation with shoulder pain, Horner's syndrome, and apical lung mass mimicking bronchial carcinoma.

Case history

A 72-year-old Caucasian man was admitted complaining of increasing left shoulder pain and stiffness of nine months' duration. He had no back pain, symptoms of anaemia, cough, haemoptysis, fever, or weight loss. He had smoked 20 cigarettes daily for 60 years. His only medical history was an episode six years previously of congestive cardiac failure, attributed to myocarditis, which had been treated with corticosteroids for three months. He was taking maintenance digoxin for chronic atrial flutter and mild diuretics. He looked quite well but he had a clinically malignant lymph node 2 cm in diameter in the left supraclavicular fossa. A left Horner's syndrome was present. Movement of the left shoulder was restricted by pain. The fingers were not clubbed. The radial pulse was 75/min and the blood pressure 110/80 mm Hg. There was no evidence of cardiac failure and no cardiac murmurs. Air entry was diminished at the left apex but the chest was otherwise clear to auscultation. The liver and spleen were impalpable. There was no motor or sensory deficit in the extremities. Chest x-ray examination showed a left apical mass with erosion of the second and third ribs posteriorly and lateral borders of adjacent vertebral bodies (figure). Haemoglobin was 13.5 g/dl, white blood count $9.4 \times 10^9/l$ ($9400/mm^3$) with normal differential, platelets $170 \times 10^9/l$ ($170\,000/mm^3$). Serum concentration of creatinine was 237 $\mu mol/l$ (2.7 mg/100 ml); albumin 38 g/l; globulin 31 g/l, with normal electrophoretic pattern; calcium 2.5 mmol/l (10 mg/100 ml), and urate 0.4 mmol/l (6.7 mg/100 ml).

A biopsy specimen of a left supraclavicular lymph node showed complete replacement by immature plasma cells, which by fluorescent antibody studies were positive for IgG. A bone marrow biopsy specimen showed small colonial proliferations of atypical plasma cells which included some binucleate types consistent with myeloma. A needle biopsy specimen of the left apical mass showed sheets of neoplastic plasma cells. Serum IgG concentration was 11.42 g/l, IgA 3.59 g/l, IgM 0.42 g/l, and IgD 0.04 g/l. Free lambda chains were detectable in the serum by immunoelectrophoresis in small amounts and in the urine in large quantities (4.5 g/l). Skeletal survey showed vertebral collapse at T10, L4, and L5. Skull x-ray examination showed a few radiolucencies, one of which was considered consistent with myeloma. The patient was treated with radiation to the left apical mass and intermittent chemotherapy with combined melphalan and prednisone.

Apical mass (myeloma) left lung with erosion of second and third ribs posteriorly and lateral borders of vertebral bodies.

Comment

Pancoast's syndrome¹ of apical lung opacity, erosion of ribs, pain in the shoulder and arm with wasting of the hand muscles, a drooping eyelid, and contracted pupil is most often due to bronchial carcinoma.²

Our patient had no wasting of the hand muscles, which is the least frequent part of the syndrome.³ The case emphasises the need for biopsy before starting to treat Pancoast's syndrome. In contrast to the generally poor prognosis with this syndrome, patients with extramedullary plasmacytoma have a much more favourable outlook. Our patient's disease probably represented stage III of extramedullary plasmacytoma according to the classification of Wiltshaw.⁴ Metastasis in this type of myeloma is more often to lymph nodes than to bone, which may be affected at sites not normally containing bone marrow. Other notable features of metastatic extramedullary plasmacytoma are a relative infrequency of bone marrow disease (21%); absence of hypercalcaemia, systemic amyloidosis, and paraplegia; and rarity of death due to renal disease.

¹ Pancoast, H, *Journal of the American Medical Association*, 1924, 83, 1407.

² Hall, W, *American Journal of Roentgenology*, 1940, 44, 838.

³ Herbut, P A, and Watson, T S, *Archives of Pathology*, 1946, 42, 88.

⁴ Wiltshaw, E, *Medicine*, 1976, 55, 217.

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Effects of naftidrofuryl on microcirculatory cold sensitivity in Raynaud's phenomenon

The treatment of Raynaud's phenomenon (RP) is unsatisfactory. That may be owing to insufficient knowledge of the underlying mechanisms in the individual patient and difficulty in assessing the effect of treatment on them. The effect of one of the newest vasoactive agents, naftidrofuryl, was investigated in a double-blind, crossover study with newly developed skin microcirculatory cold sensitivity tests.

Methods and results

Twenty normal controls and 14 patients with RP (typical skin colour changes by mild cooling) were studied. Six of the 14 patients had primary RP (Raynaud's disease for more than three years without associated disease), five had systemic sclerosis (proved by biopsy), two had rheumatoid arthritis (RA), and one had systemic lupus erythematosus (SLE). Informed consent was obtained from all. Under strictly standardised conditions in a temperature-controlled room ($18^{\circ}\text{C} \pm 0.5$, 45-50% humidity) the basal blood flow in the skin fold between the index finger and the thumb was measured from the clearance of radioactivity¹ after non-invasive labelling with xenon-133 gas. The skin microcirculatory response to localised cooling, not eliciting systemic vasoconstriction, was assessed from the basal flow changes caused by touching the labelled skin (area $2\text{ cm} \times 2\text{ mm}$) with a cooled (5°C), water-filled metal cylinder (the local cooling test).¹ Cold-induced systemic skin vasoconstriction was assessed from the basal flow changes of the right hand caused by immersion of the left hand in water of room temperature (the contralateral hand cooling test).² Results from the same individuals on different days varied by less than 15%.

Randomly the patients took on days one to seven either 200 mg of naftidrofuryl or a placebo thrice daily by mouth, and vice versa on days eight to 14. The tests were done initially and on days seven and 14. A paired *t* test was used when comparing each patient's initial results with those on day seven of each agent, and Student's *t* test when comparing the results in the controls with those in the patients. All patients completed the trial. Their initial results and those from day seven of placebo were equal to the normal controls. Their mean ($\pm$ SD) low basal flow of $4.4 \pm 1.5\text{ ml/min/100 g}$ increased significantly ($P < 0.05$) on naftidrofuryl to $6.6 \pm 2.8\text{ ml/min/100 g}$ and reached in most patients the normal mean of $6.9 \pm 1.4\text{ ml/min/100 g}$. Both cooling tests improved significantly during naftidrofuryl treatment (table). In most patients the results of the contralateral hand cooling test reached normal, whereas for many that was not so with the local cooling

Mean ($\pm$ SD) basal skin flow changes (% plus or minus) during local and contralateral hand cooling tests before and during naftidrofuryl treatment

Test	Before treatment	During treatment	P value
Local cooling	-21.5 $\pm$ 26.9	-9.0 $\pm$ 18.5	P = 0.001
Normal range	+ 7.5 $\pm$ 19.6		
Contralateral hand cooling	-49.2 $\pm$ 19.5	-25.4 $\pm$ 12.1	P < 0.001
Normal range	-23.4 $\pm$ 10.4		

test. Individual cooling test results that were within the normal range before the treatment were not much changed by naftidrofuryl. Five patients had epigastric discomfort while taking naftidrofuryl. This was the only side effect.

Comment

The mechanisms causing RP can be divided into systemic microvascular reflexes and local mechanisms (local hyperreactivity of the blood vessel walls and increased local blood viscosity)—the relative importance of each differing in individual patients—and an already decreased basal flow when such mechanisms are normal.² Earlier studies^{3,4} suggest that naftidrofuryl helps in various peripheral vascular disorders without causing hypotension and flushing. It enhances the tissues' oxidative potentials after conditions of raised anaerobic tissue metabolism.⁵ This study shows that naftidrofuryl has a beneficial effect on the basal skin flow and both the reflex and the local mechanisms underlying RP. Its effect on the microvascular reflexes was most pronounced, indicating that a better clinical effect might be expected in patients in whom these are impaired. A controlled clinical trial is therefore warranted in patients selected on these grounds. They will be found mostly among patients with RD, RA, and possibly SLE.²

I thank Lipha Pharmaceuticals Ltd for placebo capsules.

¹ Haavik Nilsen, K, *Microvascular Research*, 1978, **15**, 251.

² Haavik Nilsen, K, *Scandinavian Journal of Rheumatology*, 1978, in press.

³ Forrest, H, *Scottish Medical Journal*, 1978, **23**, 45.

⁴ Judge, T G, *Scottish Medical Journal*, 1978, **23**, 43.

⁵ Shaw, S W J, and Johnson, R H, *Acta Neurologica Scandinavica*, 1975, **52**, 231.

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Plasma lipoproteins in renal transplant patients

Changes in plasma lipoprotein patterns in chronic renal failure and after renal transplantation were reported by us,¹ and our observations have been confirmed by others.² We have re-examined the plasma lipids in 17 of our original patients four years after our initial investigation.

Patients, methods, and results

Of our 32 original patients in 1971, 17 were available for study in 1975, exactly four years later. Six of the remainder had died, five had rejected their allograft and been returned to haemodialysis, and four were unavailable for study. Out of the six who died, two had myocardial infarcts, two had renal failure due to rejection, one had haemorrhage from a duodenal ulcer, and one had a subarachnoid haemorrhage. The group consisted of seven men and 10 women with an average age of 41 years (21-56). Each patient was seen and examined after a 12-hour fast in a renal transplant follow-up clinic. Venous blood samples were analysed by semiautomated techniques

for cholesterol and triglycerides and lipoprotein patterns electrophoretically according to techniques previously described.¹

The comparative body weights, prednisolone dose per day, and plasma cholesterol and triglyceride concentrations of the 17 patients studied in 1971 and 1975 are shown in the table. During this follow-up period the dose of corticosteroids had been gradually reduced so that in the second study period the average ($\pm$ SD) dose (11 $\pm$ 1.1 mg/day) was virtually half that used in the initial period (21 $\pm$ 2.3 mg/day). Concomitantly the plasma cholesterol and triglyceride concentrations had significantly fallen and were within the normal range. Furthermore, the lipoprotein patterns had returned to normal in 11 cases, only two remaining in class IIa and four in class IV. There had been no significant change in glomerular filtration rate over this period (63.5 $\pm$ 6.1 ml/min and 73.9 $\pm$ 7.6 ml/min).

Comparative mean ($\pm$ SD) plasma triglyceride and cholesterol concentrations, glomerular filtration rates, body weights, and prednisolone dose/day in 1971 and 1975 in renal transplant patients

	1971 (n = 32)	1975 (n = 17)
Triglycerides (mmol/l)	2.4 $\pm$ 0.2	1.4 $\pm$ 0.1*
Cholesterol (mmol/l)	6.5 $\pm$ 0.3	5.2 $\pm$ 0.2*
GFR (ml/min)	63.5 $\pm$ 6.1	73.9 $\pm$ 7.6
Weight (kg)	61.4 $\pm$ 2.6	60.2 $\pm$ 2.7
Prednisolone (mg/day)	21 $\pm$ 2.3	11 $\pm$ 1.1*

*P < 0.001 (paired t test).

Conversion: SI to traditional units—Triglyceride: 1 mmol/l $\approx$ 86 mg/100 ml. Cholesterol: 1 mmol/l $\approx$ 39 mg/100 ml.

Comment

Vascular disease is the commonest cause of death in patients with chronic renal failure on haemodialysis. Cardiac and vascular accidents account for over 30% of all deaths in patients with renal transplants, taking second place only to infections.³ Our initial study in 1971¹ was prompted by a similar clinical observation. Like other authors, we found substantial hypertriglyceridaemia in the non-nephrotic renal failure group, whereas in the transplanted individuals taking an average prednisolone dose of 21 mg/day cholesterol, triglyceride, and non-esterified fatty acid levels were significantly raised. Insulin concentrations remained normal throughout. Postulating that these raised values could well be related to the corticosteroid treatment, this further study was undertaken four years later when the corticosteroid dose in the original patients had been reduced from a mean of 21 mg to 11 mg/day. A pronounced improvement was observed. Most patients had normal concentrations of cholesterol and triglycerides, and lipoprotein patterns had returned to normal.

¹ Ghosh, P, *et al*, *Transplantation*, 1973, **15**, 521.

² Casaretto, A, *et al*, *Lancet*, 1974, **1**, 481.

³ Gurland, H J, *et al*, *Proceedings of the 13th European Dialysis and Transplantation Association*, ed B H B Robinson, vol 13, p 38. London, Pitman Medical, 1976.

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Prenatal diagnosis of a galactosaemia heterozygote by fetal blood enzyme assay

Although dietary exclusion of galactose greatly improves the prognosis of galactosaemia, in all six pregnancies referred to us where the mother had given birth to an affected child the parents requested diagnostic amniocentesis so that they could choose to have only unaffected children. We describe such a mother who refused to accept the risk of having another affected child but who presented at 20