Coexistent multiple myeloma and MEN type 1

Sir,

Primary hyperparathyroidism and other endocrine tumours are occasionally associated to constitute well defined syndromes (multiple endocrine neoplasia – MEN – syndromes, type 1 and type 2). Furthermore, recent studies seem to suggest an increased risk of developing a malignant disease in patients with primary hyperparathyroidism.1

We here report the case of a 70 year old white woman who presented with clinical features consistent with acromegaly. The clinical diagnosis was confirmed by the finding of raised serum growth hormone levels in basal conditions (15 ng/ml; normal values lower than 10 ng/ml) that increased following thyrotrophin releasing hormone (TRH) administration.

The patient had undergone surgery for primary hyperparathyroidism due to a parathyroid adenoma in 1979; the diagnosis was confirmed both by the histological examination (biopsy of another gland showing normal appearance) and the occurrence of tetany after parathyroidectomy. During this hospitalization, serum protein electrophoresis showed a low γ-globulin monoclonal peak, this finding being relatively common in patients with primary hyperparathyroidism. However, unlike other reports2 this monoclonal band did not disappear following surgery. Laboratory tests showed an increase of this abnormal peak in the following years. A multiple myeloma was therefore diagnosed in 1988 based on the presence of a serum monoclonal paraprotein and bone marrow aspiration. The patient is therefore undergoing therapeutic courses with both steroidal and chemotherapeutic agents.

Previous papers have reported the rare association between primary hyperparathyroidism and multiple myeloma and between myeloma and acromegaly; on the contrary, to our knowledge, there is no paper describing the association of sporadic MEN type 1 and multiple myeloma in the same subject.

Although we cannot exclude that all three diseases might have associated in our patient by chance, we can also suppose that a common aetiological factor could link them by inducing neoplastic transformation of different cell lines. With regard to this hypothesis it must be stressed that, in animal experiments, the expression of a specific viral oncogene able to transform most cell types, results in the preferential phenotypic development of multiple endocrine tumours or, at least, in proliferative disorders of other neuroendocrine cells.3 Furthermore, a humoral factor that can stimulate parathyroid cell growth in vitro and, possibly, other endocrine cells has been recently discovered in patients with MEN type 1.4

It can also be hypothesized that among the disorders we observed only one constitutes the primary event secondarily inducing the others. Increased serum calcium levels related to hyperparathyroidism might be the primary cause, according to some studies which demonstrate that calcium acts as a mitogenic factor for some cell lines 'in vitro'.5 Alternatively the myelomatous proteins might interfere with polypeptide hormone synthesis, and/or bind their circulating fractions, and/or block their peripheral effects; these events might secondarily stimulate both parathyroid and hypophyseal secretory activity.6

Finally, it can be also speculated that persistently high growth hormone levels could stimulate cellular proliferation: a higher incidence of acute leukaemia has, in fact, been demonstrated in patients treated with synthetic growth hormone for a long time.7

D. Clements,
S. Aslan,
W.E. Wilkins
Department of Medicine,
Princess of Wales Hospital,
Coity Road,
Bridgend CF31 1RQ, UK.

References

Diaphragmatic paralysis: a difficult diagnosis

Sir,

The aetiology of orthopnoea can occasionally represent a formidable diagnostic challenge to the clinician. We report a case of diaphragmatic paralysis which had been misinterpreted as left ventricular insufficiency. In December 1988, a 72 year old man was hospitalized because of dyspnoea resistant to inotropic, diuretic and vasodilating therapy. The dyspnoea typically occurred during the night and after major effort. The patient gave a history of pulmonary tuberculosis treated by left phrenic nerve crush and lower lobe resection in 1949. The physical examination revealed right basal rales, whereas the left base was at the level of the 9th dorsal vertebra. The electrocardiogram showed left anterior hemiblock and non-specific ST-T changes in the precordial leads. On the chest X-ray, some Kerley B lines were evident in the right lower zone. The echocardiogram revealed mild left ventricular (LV) hypertrophy with a LV ejection fraction of 50%. The arterial tension of oxygen and of carbon dioxide were 72 mmHg and 42 mmHg respectively, the pH was 7.39.

The nocturnal dyspnoea was thought to be an angina equivalent, but neither isosorbide dinitrate and verapamil could prevent it nor were perfusion abnormalities shown by a thallium-201 myocardial scintigraphy. As a moderate rise of the blood pressure up to 170/100 mmHg usually paralleled the occurrence of nocturnal dyspnoea, a pathogenetic role of hypertension was supposed. Despite an effective antihypertensive treatment, dyspnoea still occurred during the night and occasionally by day if the patient lay down. After 9 days of hospital stay, a nurse noticed that during sleep the patient's abdomen moved outward and inward during expiration and inspiration respectively. Bilateral diaphragmatic paralysis was considered and was confirmed by measurement of the transdiaphragmatic pressure (Pdi). The patient could generate a maximum Pdi of only 2 cm H_2O versus normal values of 10–20 cm H_2O.

Unfortunately, extensive investigation did not provide an explanation for the right diaphragmatic paralysis. A neuritis seemed the most likely cause. A chest curiass was used at night which completely prevented dyspnoea. After about 15 months of such therapy, the patient remains asymptomatic.

Owing to a peculiar compensatory breathing pattern, the physical examination of a patient with a diaphragmatic paralysis may be misleading as may either the fluoroscopic assessment of diaphragmatic motion or the finding of normal or near normal arterial gas values. The ineffectiveness of cardiologic therapy, the dissociation between severe nocturnal and mild effort dyspnoea, the absence of upper lung blood diversion and cardiomegaly on the chest X-ray all contribute to suggest a noncardiac origin of the dyspnoea. Eventually, a consistent fall of the vital capacity and of the arterial tension of oxygen with passage from the seated to the supine posture is highly suggestive of diaphragmatic paralysis. Awareness of this condition will enable physicians to care for patients who would otherwise undergo useless investigations or ineffective or dangerous therapy.

R. Antonelli Incalzi,
O. Capparella,
A. Gemma,
P.U. Carbonin
Department of Geriatrics of the Catholic University of the Sacred Heart, Largo A. Gemelli, 8-00168, Rome, Italy.

References


'Spread-eagle' position for the rectal examination of the prostate

Sir,

Rectal prostatic examination is an integral part of a urological physical examination in male patients. Generally, for this purpose, the left-lateral (Sims') or knee-elbow position is advised in British texts, and the standing position (with the patient bent over the end of an examination table and his toes pointed inwards) is preferred in American books.

I routinely use the supine position with the knees brought up and gently abducted ('spread-eagle' position).

References

Coexistent multiple myeloma and MEN type 1.

E. Romagnoli, S. Minisola, V. Carnevale, G. Spagna, E. D’Erasmo and G. Mazzuoli

Postgrad Med J 1990 66: 879-880
doi: 10.1136/pgmj.66.780.879

Updated information and services can be found at:
http://pmj.bmj.com/content/66/780/879.citation

These include:

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/