The interesting aspects of this case concern the concomitance of unusual features, such as the young age of patient, the recently documented euthyroid state, the summer seasonality and the potential contribution of both diabetic ketoacidosis and neuroleptic drugs in precipitating myxedema coma.

References


Singular coexistence of anti-Hu syndrome, finger clubbing and pseudoscleroderma in small cell lung cancer

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The patient had been in good health until 20 months before our observation, when he started to experience pain in his left hand and cubitus; a tendinitis of the first extensor tendon. This was diagnosed: a surgical decompression was performed, but, once again, the patient showed no improvement. In the mean time, sensory abnormalities showed the feature of stocking-glove paraesthesias with sensory ataxia and wormlike involuntary movements of the outstretched hands and fingers (pseudoathetosis). Skin changes also appeared: first, the fingers and hands became swollen; then the skin gradually became firm, thickened and tightly bound to the underlying subcutaneous tissue (sclerodactyly; Fig. 1). The skin over the cheeks and forehead was tight and shiny with loss of wrinkles and facial expression. There were furrows around the mouth perpendicular to the lips. The lips were thin, and the nose had a pinched appearance. Raynaud’s phenomenon was observed intermittently. The fingers showed typical clubbing. Later, polyarthalgias of the big joints (knees, hips, elbows, shoulders) and manifestations of systemic disease appeared with anorexia, weight loss (10 kg in 8–10 months) and asthenia. The patient was evaluated immunologically to exclude scleroderma. Laboratory test results showed an increase in inflammato-
Sclerodactyly antibodies were normal. Finally, we identified neuropathy markers (CEA, CA19.9; α-FP, PSA, hCG, NSE) revealed a slight increase in the value of NSE (22 ng/ml; conventional units <12.5). Multiple faecal occult blood tests were negative. Metabolic and endocrinological diseases and nutritional deficiencies, potential causes of neuropathy, were excluded; serum minerals, TSH, vitamin B12, folic acid and α1-antitrypsin were normal; HIV and HCV infections were also excluded. The patient had a previous infection with HBV (anti-HBsAb and anti-HBcAb were found). Nailfold capillary microscopy, EEG and a cranial CT scan were all normal. New ENG/EMG tests confirmed the presence of a moderate-grade sensory-motor polyneuropathy, with functional features of both demyelinisation and axonal damage. An X-ray study of the chest showed only a small high-density nodule in the right upper lobe. The patient was submitted to a total-body CT scan: a small area of parenchymal consolidation was confirmed in the anterior segment of the right lung upper lobe (1 cm diameter), with some mediastinal lymphoadenopathy. In the abdominal scans, bilateral adrenal masses were observed; they had a dishomogeneous and hypodense appearance with a diameter of 37x30 mm of the left gland and 15 mm of the right gland. This was interpreted as adenomatous lesions, as further confirmed by an abdominal MRI. The patient was discharged without any specific diagnosis.

Two months later, a bronchoscopy was performed with a macroscopic diagnosis of diffuse chronic bronchitis: the bronchoalveolar lavage of the anterior segmental bronchus of the right upper lobe and the fine-needle aspirate of the sub-carinal nodes did not find neoplastic cells.

In March 2005 the patient was admitted to our department and was still greatly symptomatic with the described sensory abnormalities. TPA (113 UI/l; conventional units <100) and NSE (46 ng/ml) were increased, while chromogranin was not. Twenty-four-hour urinary catecholamines, aldosterone and cortisol were normal; DHEAS and adrenocorticotropic hormone (ACTH) were normal, while calcitonin (22 pg/ml; conventional units <15) and PTH (66 pg/ml; conventional units 7–53) were increased. ACE and anti-Borrelia burgdorferi antibodies were normal. Finally, we obtained the measurement of anti-cerebellum (anti-Hu, anti-Yo, anti-Ri, anti-amphiphysin) and anti-ganglioside antibodies, markers of neurological paraneoplastic disease of the peripheral and central nervous systems. The anti-Hu antibodies levels were high (positive ++).

A new total-body CT scan was performed: it showed the aforementioned nodule in the anterior segment of the right lung upper lobe, with a slight increase of its diameter. Several mediastinal lymph nodes were found; the adrenal lesions were also confirmed. We decided to perform a US-guided biopsy of the right adrenal gland: the histologic examination disclosed the presence of a malignant neuroendocrine tumour, i.e., small cell cancer. A metastatic origin was suspected.

A new bronchoscopy was performed, but it was not possible to perform a transbronchial biopsy of the lesion, because it was too small and much too peripheral to be reached with the fibroscope. Nevertheless, a fine-needle aspirate of the sub-carinal nodes was obtained, disclosing malignant cells (small cell cancer type). A bone scintigraphy was negative.

Thus, it was possible to establish a diagnosis of SCLC with mediastinal nodes and bilateral adrenal metastasis with paraneoplastic peripheral sensory-motor polyneuropathy (anti-Hu syndrome) and paraneoplastic pseudoscleroderma. The patient was therefore transferred to the oncologic clinic of our hospital where he underwent a polychemotherapy regimen. He died seven months later.

Paraneoplastic neurological syndromes are observed in fewer than 1% of malignancies. The neurologic disorders usually appear before the cancer has been identified; the tumour is found months and even a few years after the appearance of the neurologic syndrome. It is thought that most or all paraneoplastic neurological disorders are immune-mediated. The mechanism entails ectopic expression by a tumour of an antigen that normally is expressed exclusively in the nervous system (the onconeural antigens). The immune attack controls the growth of the cancer, and may in a few instances obliturate it. Paraneoplastic neurologic disorders are usually severe, disabling and sometimes lethal [1–4].
Many neurological paraneoplastic disorders have been described: anti-Hu paraneoplastic neuropathy is an uncommon immune-mediated disorder that manifests as a sensory neuropathy. The target antigens are a family of RNA-binding proteins (HuD, HuC and Hel-N1) that in normal tissues are only expressed by neurons. SCLC may express the same proteins, triggering an immune response in some patients (antibodies and cytotoxic T cells that cross-react with the Hu proteins of the dorsal root ganglia, with neuronal destruction) [5, 6].

Anti-Hu antibodies can also be found in the absence of neurological symptoms. Anti-Hu-positive patients, with or without clinical symptoms, seem to show a better prognosis [7, 8].

A variety of associations between scleroderma and malignancies have been described: lung cancer, melanoma, epithelial skin cancer, leukaemia, carcinomas of the breast, uterus, liver, stomach, oesophagus, colon, ovary, kidney, prostate and urothelium. Furthermore, some reports of scleroderma-like skin lesions have been published in patients with malignancies. Pseudoscleroderma, or pseudosclerosis, is a rare paraneoplastic syndrome, in which sclerotic skin lesions resembling systemic scleroderma occur. The putative mechanism underlying the spectrum of paraneoplastic syndromes includes the release of humoral mediators (such as serotonin in SCLC), growth factors, cytokines and antibodies by malignant cells [9, 10].

Further, as has been described in reflex sympathetic dystrophy, mutatis mutandis, there might be pathogenetic links between the cutaneous changes and the peripheral nervous system involvement that might contribute significantly to the onset and development of the cutaneous lesions.

The intriguing aspect of our clinical case is the association between the paraneoplastic neurological anti-Hu syndrome, finger clubbing and the scleroderma-like cutaneous findings (sclerodactyly and facial involvement). To our knowledge this is the first finding of such an association in the literature.

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References


Subdural haematoma in a patient with minimal risk factors

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The use of head computed tomography (CT scan) to evaluate patients with minor head injury continues to be a controversial issue. While all head-injury patients presenting with a Glasgow Coma Scale (GCS) less than 13 undergo scanning, the decision to scan patients with a GCS of 13–15 remains varied. In this group the yield of finding an abnormality on head CT scan is generally low at 0.7%–3.7%, but the risk of “missing” such a significant finding can be catastrophic [1]. For this reason many physicians feel that a head CT scan should be required for all patients presenting with minor head injury. However, due to increasing costs and nationwide variability in practice, others feel that a “CT head rule” should be developed to provide standardised guidelines and decrease unnecessary imaging studies. All attempts thus far to identify the most accurate clinical variables of patients with minor head injury to predict a positive finding on head CT scan have not been successful. Design variability, data analysis and debate over an acceptable sensitivity level have