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LETTERS TO THE EDITOR

Superior Vena Cava Syndrome: An Important Differential Diagnosis in Patients with Facial Edema

Sir,

Facial edema is a frequent symptom in dermatological patients. At first glance, facial edema is often mistaken for classic angioedema, but may also be related to the administration of angiotensin-converting enzyme inhibitors (1), the Melkersson-Rosenthal syndrome (2), dermatomyositis and viral or bacterial diseases (e.g. erysipelas). A further but rarer differential diagnosis is Morbus Morbihan, i.e. persistent facial edema which, for example, occurs in patients with acne, rosacea or after erysipelas (3). Furthermore, patients with kidney diseases often exhibit facial swelling, especially of the eyelids (4).

In this paper we describe three patients initially examined by dermatologists because of their most visible symptom – acute or chronic facial swelling. In all three patients, the examination of facial edema resulted in detection of a superior vena cava syndrome caused by malignant tumours.

Clinically, this diagnosis should be suspected by the presence of associated findings such as dilatation of the two external jugular veins and increasing symptoms when the patient is in a horizontal position. Rapid detection of this condition is essential, otherwise acute life-threatening complications can be expected.

CASE REPORTS

Case 1

Within the course of 7 weeks, a 65-year-old man developed swelling and flushing of the face and upper extremities which increased overnight when he lay in a horizontal position. He also complained of headaches and the formation of dilated skin vessels on the upper trunk (Fig. 1). Computed tomography of the chest some weeks previously had not shown any pathological findings. A physical examination disclosed swelling of the face and hands and dilatation of the superficial veins of the chest and of both external jugular veins. Repeated contrast-medium-enhanced computed tomography showed compression of the superior vena cava caused by a tumour in the upper mediastinum.

Histopathological examination revealed a low differentiated adenocarcinoma. The patient underwent a stent implantation into the superior vena cava and radiation treatment. Just a few days after stent implantation, the clinical symptoms improved dramatically.

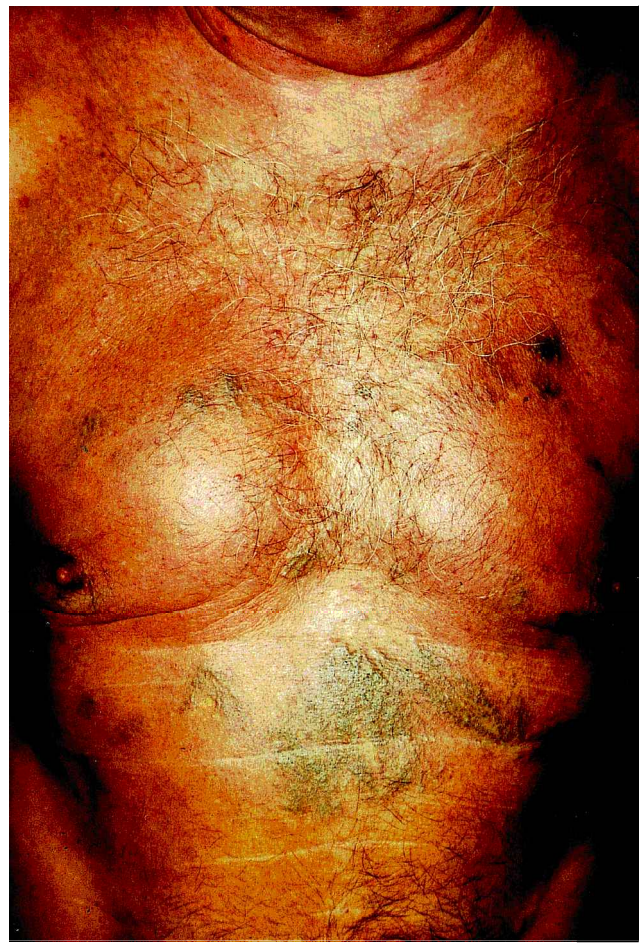

Fig. 1. Dilated superficial veins on the upper trunk in patient 1.

Case 2

A 50-year-old man was admitted with edema of the face and neck, flushing and a dilated external jugular vein on the right side, which had developed within the course of 6 months (Fig. 2). The symptoms increased when the patient was lying down or doing hard physical work. Six months previously, thrombosis of the right internal jugular vein, the subclavian vein and the superior vena cava had been diagnosed. At that time computed tomography of the chest did not reveal any underlying cause. Laboratory tests revealed an activated protein C (APC) resistance and Paget-von Schroetter syndrome (thrombosis of the axillary or subclavian veins following muscular effort) was suspected owing to the hard physical exertion which the patient had to perform daily in his job as a welder. A stent was implanted in the cranial superior vena cava to improve the clinical symptoms. After stent implantation all clinical findings showed regression. Three months later, further computed tomography of the chest showed a tumour growing in the upper mediastinum. The histopathological diagnosis was embryonal rhabdomyosarcoma. Because of compression of the aorta and the esophagus as well as severe heart disease, the tumour was found to be inoperable. At present, the patient is undergoing chemotherapy.

Case 3

A 50-year-old woman had noticed swelling of her face, especially of the eyelids, for the first time several weeks previously. In addition, she had developed dyspnea and a cough as well as pain and weakness of the upper arms. An increase in weight and tension of the mammae were further symptoms. Physical examination disclosed periorbital edema, swelling of the mammae and dilated superficial veins on the chest. Auscultation of the lung revealed that the respiratory sound of the right lung was reduced. Computed tomography of the chest was performed immediately, disclosing a tumour in the upper mediastinum with narrowing of the right pulmonary artery, the right main bronchus and the superior vena cava. Histopathological examination showed a parvicellular bronchial carcinoma. Computed tomography of the brain and the abdomen showed metastasis of the brain, the liver and the suprarenal gland. Following initial radiotherapy of the chest, the tumour shrank and all symptoms disappeared. Furthermore, the patient underwent chemotherapy with carboplatin/etoposide and radiotherapy of the whole brain.

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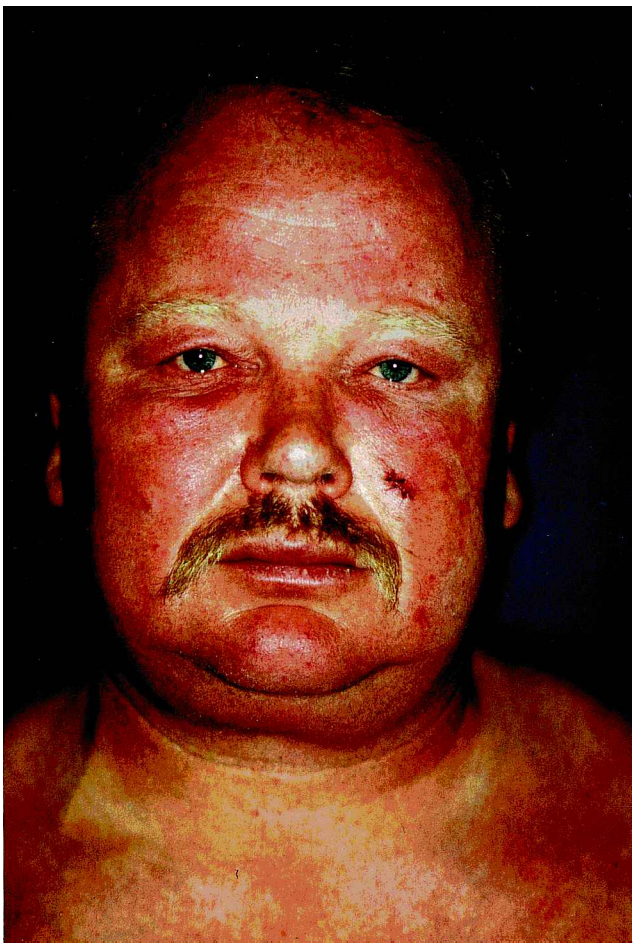

Fig. 2. Flush and swelling of the face in patient 2. Dilatation of the right external jugular vein. A sutured incision from skin biopsy can be seen on the left cheek. The patient agreed that the photograph could be published without masking.

DISCUSSION

Superior vena cava syndrome often shows gradual development with increasing swelling of the face and upper extremities. A further and very characteristic clinical feature is extension of the external jugular veins and increasing symptoms when the patient is in a horizontal position. If the syndrome develops slowly, dilated superficial veins of the chest can also be seen. Dyspnea, acute swelling, flushing and cyanosis of the face and neck indicate the immediate onset of superior vena cava syndrome (5).

Numerous different diseases may cause superior vena cava syndrome. In up to 85% of cases, a primary lung carcinoma is the underlying cause (10). In our first case, a low differentiated adenocarcinoma of the lung could be found, and in the third case a parvicellular bronchial carcinoma was the underlying cause of the superior vena cava syndrome. Only in the second case was a rare tumour such as embryonal rhabdomyosarcoma revealed by histopathological examination.

In addition, lymphomas, invasive thymomas, metastatic lymph nodes or fibrosing mediastinitis can lead to compression of the superior cava vein. Other causes might be, stenosis after multiple central venous catheterization, peritoneovenous shunts or cardiac pacemakers (6), or benign tumours such as retrosternal goiters (7). Rare causes described in the literature have been intrathoracic plasmocytoma (8), Behcet's syndrome (9), syphilitic aneurysm of the ascending aorta (10) and superior vena cava syndrome in association with infectious diseases such as Klebsiella pneumoniae pneumonia (11). In another case, superior vena cava syndrome was the main symptom of a mediastinal amelanotic melanoma (12).

The onset of superior vena cava syndrome demands immediate therapy. Depending on the underlying diagnosis, treatment may include surgical reconstruction of the superior vena cava (13), radiation treatment (14) or percutaneous stenting (15).

In conclusion, the three cases presented here clearly demonstrate that, in patients with facial edema, superior vena cava syndrome should always be taken into consideration.

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