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PHOTO QUIZ

A young woman with facial oedema

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CASE REPORT

A 23-year-old woman of Surinam-Hindu origin presented to our clinic with a two-week history of fatigue, headache, constipation and joint pain. Since four days she noticed a swollen face. She had no medical history, was not on any medication and the family history was unremarkable. Physical examination showed a moderately ill woman with a blood pressure of 130/60 mmHg, a pulse of 105 beats/min and a temperature of 37.8 °C. Her face was pale with periorbital oedema and oedema of the upper lip. Her thyroid gland was not enlarged.

Figure 1. *Before treatment*

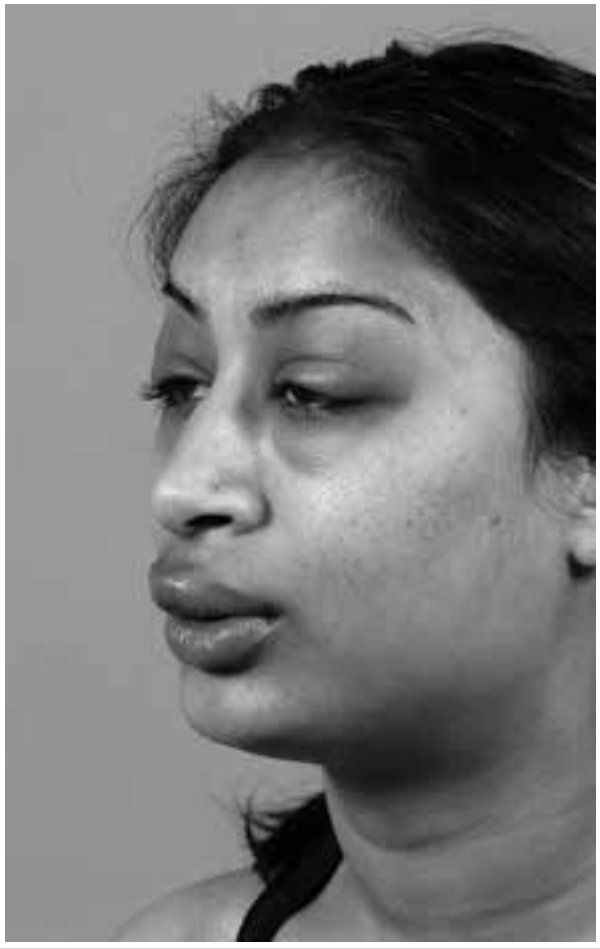

Figure 2. *After treatment*

WHAT IS YOUR DIAGNOSIS?

See page 208 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 205)

A YOUNG WOMAN WITH FACIAL OEDEMA

DIAGNOSIS/DISCUSSION

Laboratory results showed an elevated level of thyroid-stimulating hormone (TSH, 65 mIU/l), a reduced level of free thyroxine concentration (9.1 pmol/l) and thyroid peroxidise antibodies (TPO-Ab) were present. The presence of TPO-Ab supports that an autoimmune thyroid disease (Hashimoto's disease) is the cause for the diagnosed hypothyroidism. Hashimoto's disease is the most common cause of hypothyroidism in iodine-sufficient regions. It is caused by cell- and antibody-mediated destruction of thyroid tissue.[1]

Hypothyroidism is a common disease and has well-known signs and symptoms. It can affect all organ systems. In our case the most remarkable signs were the cutaneous manifestations: the pale skin, periorbital oedema and oedema of the upper lip. This is called myxoedema. The term myxoedema, formerly used as a synonym for hypothyroidism, refers to the symptoms of the skin and subcutaneous tissues in patients with a severely hypothyroid state.[2] Myxoedema is rarely seen today. A possible explanation could be the fact that patients now seek medical attention in a much earlier phase of their disease.

Hypothyroidism causes an increased deposition of glycosaminoglycans, especially hyaluronic acid, in the dermis. The increased deposition causes mucinous oedema which is responsible for the thickened features and a puffy appearance. The main cause of the depositions of glycosaminoglycans is local expression of the TSH receptor.[3,4] Myxoedema is a non-pitting oedema and is apparent around the eyes, on the dorsa of the hands and feet and in the supraclavicular fossae. It can also cause thickening of the tongue and the pharyngeal and laryngeal mucous membranes.[2] A histologically similar deposit may occur in patients with Graves' disease, usually over the pretibial area.[3] The skin pallor is very common and results from both peripheral vasoconstriction and increased deposition of water and mucopolysaccharides in the dermis, which alters the refraction of light.[5]

We treated our patient with oral administration of synthetic thyroxine, levothyroxine 50 mcg daily to reach an euthyroid state. This can easily be accomplished in almost all patients and appropriate treatment reverses the clinical manifestations of hypothyroidism.

After four weeks our patient still had some complaints of constipation, weight gain and fatigue but overall she felt rather well. The facial swelling had completely disappeared. Her laboratory results still showed an elevated TSH level and a reduced free thyroxine concentration. We increased the dose of levothyroxine. A few months later she reached the euthyroid state and had no more complaints. Hashimoto's disease is a permanent condition, and lifelong therapy is necessary.

In conclusion autoimmune hypothyroidism was diagnosed. Myxoedema as seen in this case is rarely seen today but it is a typically presentation of hypothyroidism. The term should be reserved for description of the physical signs.[2]

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