Treatment was started with DDS, 50 mg per os daily. It was well tolerated but led to only partial control of the lesions and pruritus. Complementary systemic steroid therapy with betamethasone, 1.5 mg per os daily, led to complete disappearance of the symptoms within few days. Good control was maintained after reduction of the dose of betamethasone to 0.5 mg on alternate days, but attempts to discontinue the steroid were unsuccessful because of the rapid recurrence of blisters and pruritus. Since January 1978 the child has been on a gluten-free diet (which according to his mother he observes strictly). He is presently receiving 50 mg of DDS daily and 0.5 mg of betamethasone every second day. Occasionally 2-4 bullae have appeared, usually when the betamethasone dose is reduced to 0.25 mg every other day.

DISCUSSION

On histological examination of this case of blistering disease a subepidermal bulla was seen, as characteristic of BP and DH, but the negative IF tests performed on skin and serum led to exclusion of these potential diagnoses and a diagnosis of CBDC was therefore considered, although jejunal biopsy showed villous atrophy as in DH and functional tests showed the presence of intestinal malabsorption. Now some authors (6) believe that after a certain length of time most cases of CBDC evolve into true cases of DH, with the typical IF findings; in the present case the finding of villous atrophy of the present case the finding of villous atrophy of the able. However, because previously positive IF tests were observed in all the 22 cases of DH (15 children and 7 adults) seen in our Department (4), because DDS alone failed to control the disease and because after 7 months of gluten-free diet it was impossible to reduce the dosage of the drugs, this case was classified as CBDC.

ACKNOWLEDGEMENT

This study was partly supported by Fondazione 'Pro Ricerca Dermatologica', Rome, Italy.

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Multiple Glomus Tumours: A Report of a Family in Denmark

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Received September 22, 1978

Abstract. In a family affected by multiple glomus tumours, one of the members had approximately 500 tumours spread over the entire skin. The histopathological similarity to cavernous haemangioma is emphasized. We consider that multiple glomus tumour is a more common skin disease than is assumed today.

Multiple glomus tumour (MGT) is a benign, dominant, hereditary skin disease. The tumours develop from specialized arterio-venous anastomoses, the glomus bodies, which are particularly numerous on the distal parts of the extremities. Only 21 families suffering from MGT (3, 4, 5) have been reported since Touraine (6) first described the disease in 1936. The clinical picture is characterized by painless, small bluish, partly compressible tumours, often somewhere between 10 and 20 in number, on the extremities. We have studied a family affected by MGT, where one of the members had approxi-

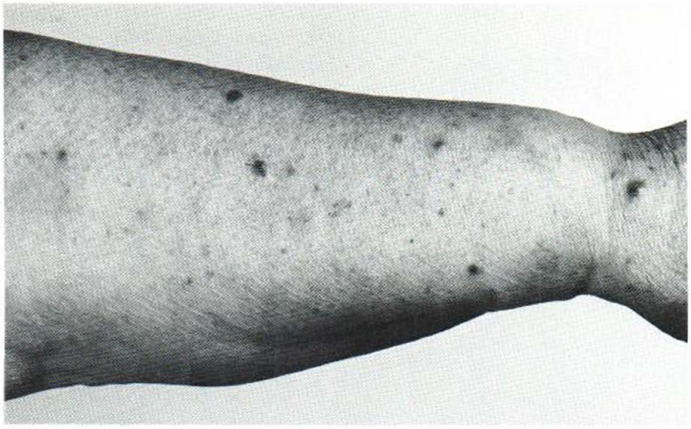


Fig. 1. Multiple glomus tumours on the ulnar aspect of the left lower arm.

mately 500 tumours dispersed over the entire surface of the skin.

FAMILY REPORT

The proband was a 71-year-old woman, who ever since puberty had developed numerous tumours in the skin. The first tumours appeared on the extremities (Fig. 1). In more recent years they also developed over the trunk and on the face. The tumours are quite painless, but a few around the joints cause some discomfort. They vary in appearance. The largest are up to 20 mm in diameter, elevated, compressible to a certain extent, and the vivid blue colour partly disappears on diascopy. The smaller tumours lie deeper in the epidermis, are of a more solid consistence, are only slightly elevated, and have a paler, bluish hue, which also only partly disappears on diascopy. No haemorrhage has ever issued from the tumours, and none occurs in the mucosa of the mouth, nose, pharynx, vagina or rectum. She has never suffered from haemoptyses, haematuria, or blood in the stools. All the laboratory tests proved normal and there is no anaemia or bleeding abnormality. Otherwise, she has always been healthy.

Histological examination showed vascular tumours lying below the normal epidermis in the reticular dermis, each consisting of large irregular cavities, bordered by a layer of flattened endothelium, and surrounded by a few layers of uniform, cubic glomus cells (Fig. 2). There were no elastic fibres in the vascular walls, and the tumour was not encapsulated.

The other affected members of the family are shown in Fig. 3. The daughter of the proband is now 42 years old. She has developed 20 tumours on the legs since the age of 12 years. A brother, 62 years old, has a similar number of tumours on the arms, but had never noticed these until pointed out at the examination. A sister, 78 years of age, has 16 tumours, which have developed on the arms, since the age of 20 years. The diagnosis was verified histologically in all the affected members of the family. The fourth generation of the family are all children, and at the time of examination they had no tumours. An older sister, now dead, and the deceased mother of the proband are both said to have had a dozen bluish tumours on the arms.

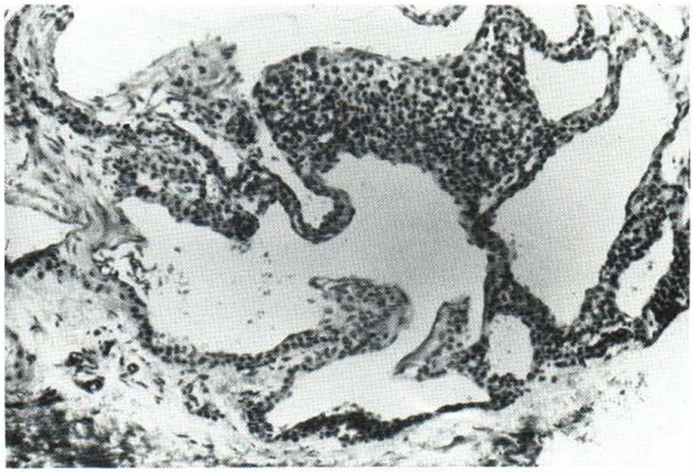
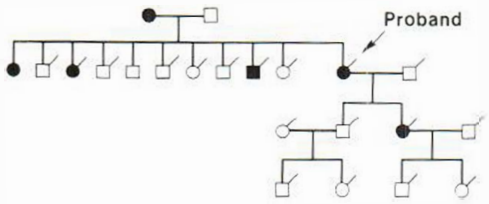


Fig. 2. Large irregular spaces lined by a flattened endothelium and two or more layers of cuboidal glomus cells in the reticular dermis.



● ■ female/male affected
♂ ♂ female/male investigated

Fig. 3. Family pedigree.

None of the married members of the family were blood related.

DISCUSSION

The present case differs from earlier reported families with MGT with respect to the large number of tumours. In a single previous article (2), a 70-year-old man was reported to have 400 glomus tumours and thrombocytopenia. There were no other cases of MGT in that family. In our proband the histological picture was characteristic, but the initial histological diagnosis was cavernous haemangioma, and this is, as reported earlier, a common error (4). As the clinical differential diagnosis is that of blue rubber bleb nevus (multiple cavernous haemangioma) (1), there is a considerable risk of a diagnostic error (4). The other members of the present family had MGT with a characteristic number and localization, though these had not even been noticed by one member of the family, even though the "blue spots" had often been discussed amongst themselves.

This is the first reported case of hereditary MGT in Scandinavia, but we consider that MGT is more common than hitherto to assumed and that the diagnosis should be considered in all cases of bluish tumours, often discrete, particularly when appearing on the extremities.

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Scleredema of Buschke with IgA Deficiency

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Received July 12, 1978

Abstract. We describe here a case of Scleredema of Buschke in a female patient aged 63 with IgA deficiency. The disorder appeared after an acute episode of tonsillitis, followed by non-pitting, woody hardness of the skin of the face, neck, shoulders and upper part of the trunk. The disorder resolved after 5 months of penicillin treatment.

Key words: Scleredema; IgA deficiency; Streptococcal infection

Scleredema of Buschke is a rare disorder of unknown etiology (3) which occurs at all ages. According to an investigation by Greenberg et al. (6), 29% of cases appear before the age of 10, 22% in the age range 10 to 20 and 49% over the age of 20 (6). The condition is characterized by non-pitting edema and induration of the skin, usually begins on the face and spreads rapidly to the neck and trunk. The disorder frequently appears after an infectious episode and clears up by itself after some months or years. Generally the disease is benign and as far as we know it has ony resulted in one death (8).

The etiology of scleredema is unknown, but obstruction of the lymphatic ducts by inflammation, streptococcal hypersensitivity, disorders of the peripheral nervous system, and pituitary function of estrogens have been blamed for the disorder. Some persistent cases are reported to be associated with severe diabetes mellitus, but neither the skin nor the diabetes responds to anti-diabetic treatment (2, 4).