

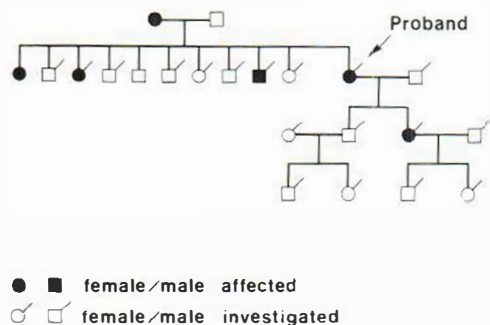


Fig. 3. Family pedigree.

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.

REFERENCES

1. Bean, W. B.: *Vascular Spiders and Related Lesions of the Skin*, 178 pp. Blackwell, Oxford, 1958.
2. McEvoy, B. F., Waldman, P. M. & Tye, M. J.: Multiple haematomatous glomus tumours of the skin. *Arch Dermatol* 104: 188, 1971.
3. Pepper, M. C., Laubenheimer, R. & Cripps, J.: Multiple glomus tumours. *J Cutan Pathol* 4: 244, 1977.
4. Rycroft, R. J. G., Menter, M. A., Sharvill, D. E. &

Wells, R.S.: Hereditary multiple glomus tumours. *Trans St John's Hosp Dermatol Soc* 61: 70, 1975.

5. Schnyder, U. W.: Über Glomustumoren. *Dermatologica* 131: 83, 1965.

6. Touraine, A., Renault, S. & Renault, P.: Tumeurs glomiques multiples du tronc et des membres. *Bull Soc Fr Dermatol et Syph* 43: 736, 1936.

Scleredema of Buschke with IgA Deficiency

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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 only 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).

Table 1. The values of immunoglobulins and C3 in the patient's serum and the range of normal values

IgG=2 200 mg/100 ml	Normal 600-2 000 mg/100 ml
IgM=130 mg/100 ml	Normal 50-200 mg/100 ml
IgA=not measured	Normal 150-300 mg/100 ml
IgD=2.5 mg/100 ml	Normal 1-10 mg/100 ml
IgE=1 500 IU/ml	Normal 350-1 750 IU/ml
C3=120 mg/100 ml	Normal 90-200 mg/100 ml

Immunological disturbances are not reported in the literature as having any association with scleredema. Recently, we were afforded an opportunity to observe a female patient with scleredema of Buschke with IgA deficiency.

CASE REPORT

A female patient aged 63, complaining of a 3-month history of tightness and hardening of the skin of the upper part of the body which started a month after an episode of tonsillitis with high temperature (40°C). On examination she had a non-pitting, woody hardness of the skin of the face, neck, shoulders and upper part of the trunk, making it very difficult for her to turn her head and causing discomfort when breathing hard.

Laboratory investigations revealed normal complete blood counts, elevated ESR (50/90), anti O titre 560 U Todd, ANA negative, LE cells negative, RA test negative. Electrophoresis of serum proteins revealed a normal situation and plasma total proteins were normal. Immunoelectrophoresis revealed the absence of IgA precipitation line. Quantitative estimation of serum immunoglobulins and C3 by the radial immunodiffusion technique gave the results listed in Table 1. Chest X-ray examination was negative and cultures from the pharyngeal cavity revealed the presence of *b*-haemolytic streptococcus.

Histological examination of samples taken from the affected skin showed normal epidermis. The dermis was thick, with swollen collagen bundles. Staining for acid mucopolysaccharides gave positive results.

Therapy was commenced with 600000 IU of Benzathine penicillin G (Penadur; L. A. Wyeth, USA) every 4 days for 5 months, where upon lesions cleared up completely.

DISCUSSION

The etiology of scleredema is still unknown. Among the etiological factors, streptococcal infections have been implicated in the pathogenesis of the disorder. In the case presented, an episode of acute tonsillitis is mentioned 20 days before the onset of

the disease. Whether streptococcus, in the case presented, was the trigger factor in the appearance of the scleredema is difficult to say. Another point to be explained is the complete absence of IgA and the association with the disease. As far as we could judge, the patient was well, apart from this disorder and no other diseases such as autoimmune phenomena are mentioned in her medical record. The normal values of IgG, IgM, IgD, IgE and C3 indicate that the patient had a selective IgA deficiency which in fact represents an isolated immunoglobulin deficiency.

Deficiency of IgA occurs with an incidence of one in 700 normal populations (1, 7). In several studies on selective IgA deficiency it was intimated that this defect may be benign (5, 9) or associated with other diseases (10). Whether the deficiency of IgA is in any way associated with scleredema of Buschke is difficult to determine, as this is the first published report and we are not aware that any of the cases reported in the past had IgA deficiency. It would be of great value if, in future cases of this disorder, search is made for the presence of IgA or other immunoglobulin deficiencies and, if found, a follow-up study to determine its association with the disease entity would be valuable.

REFERENCES

1. Bachman, R.: Studies on the serum γ A-globulin level. III. The frequency of A- γ A globulinemia. *Scand J Clin Lab Invest* 17: 316, 1968.
2. Cohn, A. B., Wheler, C. E. & Brigaman, A. R.: Scleredema adultorum of Buschke and diabetes mellitus. *Arch Dermatol* 101: 27, 1970.
3. Curtis, A. C. & Shulak, B. M.: Scleredema adultorum. *Arch Dermatol* 92: 526, 1965.
4. Fleischmajer, R., Faludi, G. & Krol, S.: Scleredema and diabetes mellitus. *Arch Dermatol* 94: 84, 1970.
5. Goldberg, L. S., Barnet, E. V. & Fudenberg, H. H.: Selective absence of IgA: A family study. *J Lab Clin Med* 72: 204 a, 1968.
6. Greenberg, I. M., Geppert, C., Worthen, H. C. & Good, R. A.: Scleredema adultorum in children. *Pediatrics* 32: 1044, 1963.
7. Hobbs, J. R.: Immune imbalance in dysgamma-globulinaemia type IV. *Lancet* i: 110, 1968.
8. Leinwand, I.: Generalised scleredema. Report with autopsy findings. *Ann Int Med* 34: 226, 1951.
9. Rockey, J. H., Hanson, L. A., Heremans, J. F. & Kunkel, H. G.: Beta-2A-aglobulinemia in two healthy men. *J Lab Clin Med* 63: 205, 1964.
10. West, C. D., Hong, R. & Holland, N. H.: Immunoglobulin levels from the newborn period to adulthood and in immunoglobulin deficiency states. *J Clin Invest* 41: 2054, 1962.