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Identify the triad

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1. Case description

A 46 year old man presented with a 15 year history of generalized excessive sweating and palpitations. He also had progressive redness of both eyes along with blurred vision. Clinical examination revealed clubbing, soft tissue swelling involving all digits (Fig. 1A) and asymptomatic multiple raised firm skin coloured nodules and coalescing plaques of around 5 × 5 cm over both shins (Fig. 1B). A single depigmented patch suggestive of focal vitiligo was also noted over the neck. Ocular examination revealed conjunctival chemosis (right > left), proptosis and visual acuity of 6/12 of right eye (Fig. 1C). Histopathological examination from the plaques demonstrated the primary pathology in dermis showing loosening of collagen which was confirmed to be secondary to mucin deposition. (Fig. 1D, E).

2. What is the diagnosis?

2.1. Diagnosis

Thyroid function tests were deranged. The clinical picture is consistent with Graves' disease (GD). GD is an autoimmune hyperthyroid disorder, this patient had classical triad of thyroid associated orbitopathy (TAO), thyroid dermopathy in form of pretibial myxedema and thyroid acropachy (clubbing, soft tissue swelling of digits with periosteal new bone formation), known as Diamond triad. It is also termed as EMO syndrome (exophthalmos, myxedema, osteoarthropathy) [1]. These features usually appear in the above mentioned chronological sequence. In GD, autoantibodies are directed against the thyroid stimulating hormone receptor (TSHR) found most commonly in the thyroid gland, fibroblasts and adipocytes. These antibodies stimulate fibroblasts leading to excessive dermal deposition of glycosaminoglycan's. Dermopathy is often restricted to dependent areas of lower limbs and has concomitant disturbance of lymphatic drainage giving rise to non-pitting edema [2]. The above patient also had vitiligo, which as per a recent metanalysis is associated with autoimmune thyroid disorders in around 14.3% cases [3]. Management options of GD include antithyroid drugs (thionamides), radioactive iodine and surgery. Various local therapies like potent topical steroids, intralesional steroid, local compression and UVA1(ultraviolet A) phototherapy have been used for treatment of pretibial myxedema. Severe cases require systemic treatment with oral steroids, octreotide, intravenous immunoglobulins and plasmapheresis.

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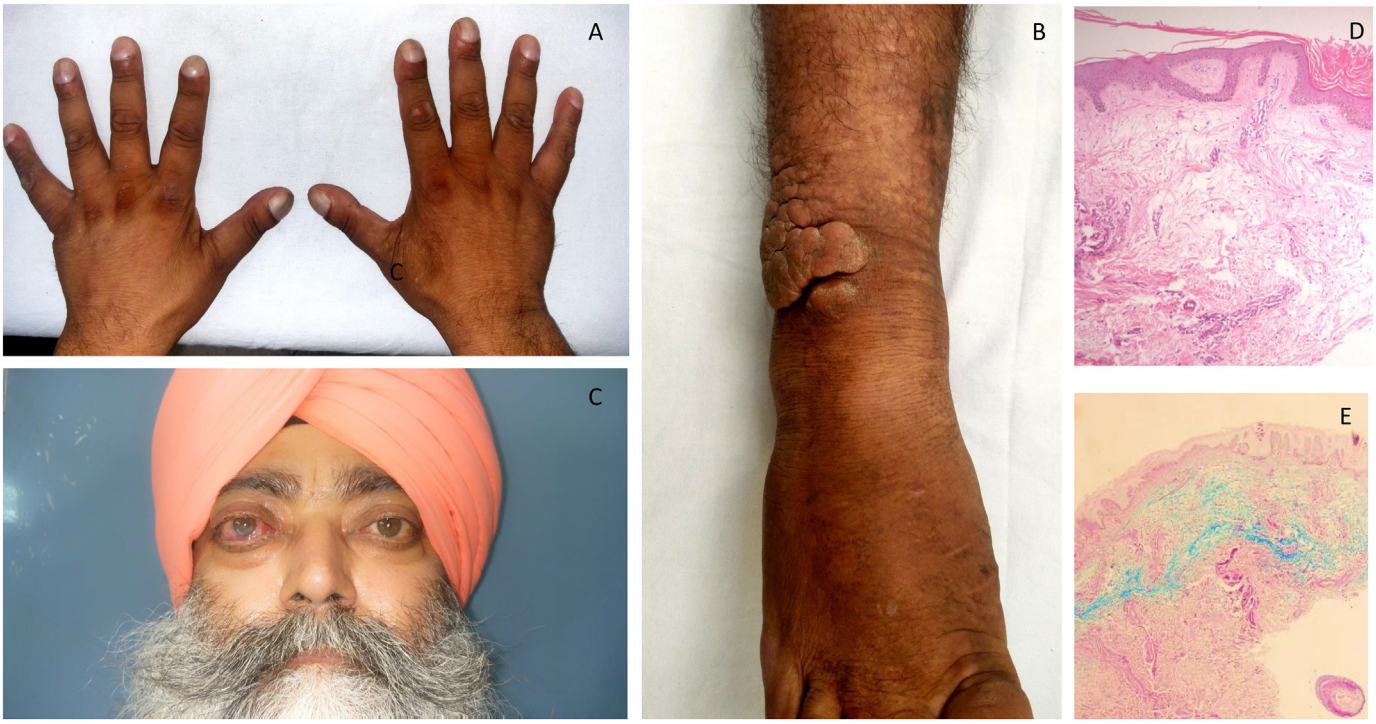

Fig. 1. Panel A- Soft tissue swelling and clubbing involving all the digits. Panel B- Asymptomatic raised skin coloured plaques over right shin. Panel C- Conjunctival chemosis and proptosis (right eye > left eye). Panel D- Loosening of collagen in the dermis (Hematoxylin-Eosin, 10×). Panel E- Bluish stained mucin within the dermis (Periodic acid Schiff, 10×).

Conflict of interest

None.

Sources of support

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