The half-yellow man

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Introduction
Diffuse normolipaemic plane xanthoma is a rare form of xanthoma usually associated with an underlying haematological or inflammatory condition, and may present many years prior to the onset of systemic illness. We present a case of this uncommon condition occurring in association with a monoclonal gammopathy of unknown significance.

Case study
A 74-year-old Caucasian male presented with diffuse yellow-orange skin discolouration, which had progressed over the preceding few months. There was no medical history of note, and he was not taking any medication. There was no family history of dyslipidaemia or premature cardiovascular disease. On examination, the patient was found to be clinically well, but had diffuse mildly raised yellow-orange skin discoloration of his face, neck, arms, chest and back (Figure 1). *Corresponding author, emails: bmerwitz@hotmail.com, brad.merwitz@gmail.com

Figure 1: Xanthomata, featuring extensive involvement of the chest, upper limbs and face

Evidence of necrobiosis or Touton cells, in keeping with diffuse normolipaemic plane xanthomatosis (DNPX) (Figure 2). The erythrocyte sedimentation rate was 76 mm/hour and an immunoglobulin G kappa monoclonal band was demonstrated on serum protein electrophoresis. The urine was negative using the Bence-Jones protein test. Both the chest radiograph and an abdominal ultrasound were unremarkable. The electrocardiogram showed a normal tracing. A bone marrow aspirate and trephine biopsy demonstrated a variably cellular bone marrow with < 10% plasma cells, suggestive of a monoclonal gammopathy of unknown significance (MGUS).

These findings are consistent with a diagnosis of DNPX, associated with MGUS. The patient was started on atorvastatin 20 mg daily for his mild hyperlipidaemia, and did not require specific therapy for the skin lesions. There was some resolution of the skin lesions on statin therapy, and he continues to follow-up routinely at our facility.

Discussion
Also called generalised planar xanthomatosis, DNPX is a rare form of xanthoma which occurs as a macular yellow-orange or yellow-brown skin discoloration, and is most commonly seen in a symmetrical distribution over the upper torso and neck, though rarely as extensively as in our patient. These xanthomata were originally thought to occur idiosyncratically because of
their association with normal serum lipid levels. However, many of these patients subsequently developed paraproteinaemia or dysglobulinaemia. In addition to MGUS, multiple myeloma, leukaemia, adult T-cell lymphoma, cryoglobulinaemia, rheumatoid arthritis, Takayasu’s arteritis and eosinophilic granulomatosis have all been described as being associated with the condition. The skin lesions may precede the haematological or systemic disorder by many years. The cause of the xanthomatous deposition within the skin is not well understood, but a number of theories have been proposed. According to the most plausible theory, the monoclonal immunoglobulin binds with apolipoprotein B-100 on the LDL particles at a site remote from the LDL receptor-binding site. This does not result in a change in affinity for the LDL receptor, but in an increased affinity for the acetyl LDL receptor, enhancing macrophage uptake and esterification, and resulting in cutaneous xanthomata with normolipaemia.

Conclusion
This case illustrates the occurrence of DNPX in association with a monoclonal gammopathy of unknown significance. Although a rare form of xanthomatosis, this condition often heralds an underlying haematological or systemic condition, and highlights the need to search for such conditions.

References

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