CASE REPORT

Multiple xanthogranulomas in an adult patient: clinical, dermoscopic, reflectance confocal microscopy and histopathological features

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SUMMARY

A case of multiple xanthogranulomas located bilaterally on arms, armpits, legs, trunk, abdomen, scalp, ear and plantar surfaces in a 25-year-old man is reported. The patient was evaluated by polarised light dermoscopy and reflectance confocal microscopy (RCM) that showed peculiar aspects. In particular, dermoscopy showed a homogeneous orange-yellowish hue that is related to the histopathological presence of foamy histiocytes and Touton giant cells; the last typically appeared at RCM as peculiar giant cells surrounded by highly refractive ring due to the cytoplasm rich of lipids. Laboratory investigations, including routine haematological examination, liver and renal function tests, serum lipid and sugar levels were normal. The patient started isotretinoin 20 mg once daily. After 6 months, some lesions flattened, leading to yellowish or hyperpigmented macules, but new lesions raised up.

BACKGROUND

Xanthogranuloma (XG) is a benign non-Langerhans cell histiocytosis that occurs primarily in infancy and childhood and is also called 'juvenile xanthogranuloma'.[1] Around 10% of XGs occur in adulthood, particularly in the third and fourth decades of life.[1 2] Multiple XGs in adults are rare, with <30 cases reported in literature.[1 3] Clinically, XG appears as a red-yellowish, dome-shaped papule, usually asymptomatic,[1 4] which generally regresses spontaneously in children while tend to persist in adults.[5] Histopathologically, XG is characterised by a dense granulomatous dermal infiltrates consisting of foamy cells, multinucleated giant histiocytes with wreath-shaped nuclei and foamy cytoplasm, called 'Touton giant cells', lymphocytes and a few eosinophils mainly at upper dermis.[1] Although generally no treatment is necessary for XG, in case of multiple lesions in adults, the use of systemic isotretinoin has been suggested.[6]

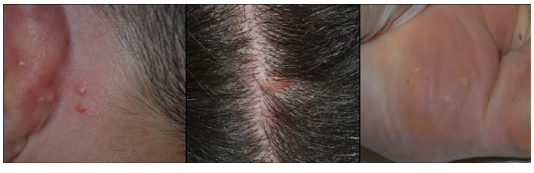

Figure 1 Orange-yellowish papules located on the retroauricular region, the scalp and the plantar surface.

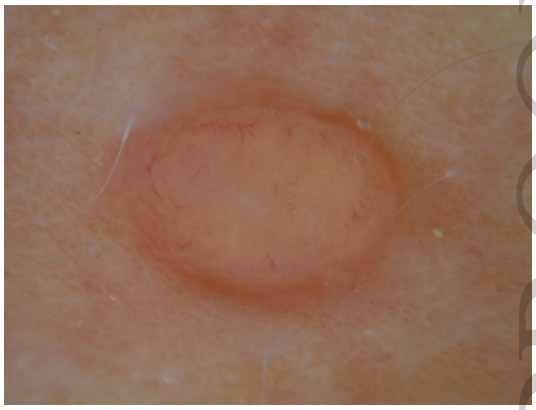

Figure 2 Polarised light dermoscopy of a lesion showing a homogeneous orange-yellowish colour, linear vessels and a thin erythematous border ('setting sun' appearance).

A case of multiple XGs in an adult patient evaluated by polarised light dermoscopy (PLD) and reflectance confocal microscopy (RCM) is reported.

CASE PRESENTATION

A 25-year-old man presented with a 3 months history of several papules and nodules located bilaterally on arms, armpits, legs, trunk, abdomen, scalp, neck and plantar surfaces causing discomfort during deambulation. Skin examination revealed well-demarcated, red to yellow-brown, domed papules and nodules with a shiny surface and of variable sizes (0.2–0.5 cm) (figure 1). The lesions were soft to elastic in consistency. The patient denied any exposure to fatty food, outdoor work or drugs. No other family member was affected.

INVESTIGATIONS

Laboratory investigations, including routine haematological examination, liver and renal function tests, serum lipid and sugar levels were all normal.

PLD (Dermlite hybrid, X10; 3 Gen, San Juan Capistrano, California, USA) revealed the presence of the same features in all lesions, consisting of a homogeneous orange-yellowish colour and linear and/or branched vessels. In some cases, a thin erythematous border was also visible, configuring a 'setting sun' appearance (figure 2).

Handheld RCM (Vivascope 3000, Caliber I.D., Rochester, New York, USA distributed in Europe by Mavig GmbH, Munich, Germany) performed

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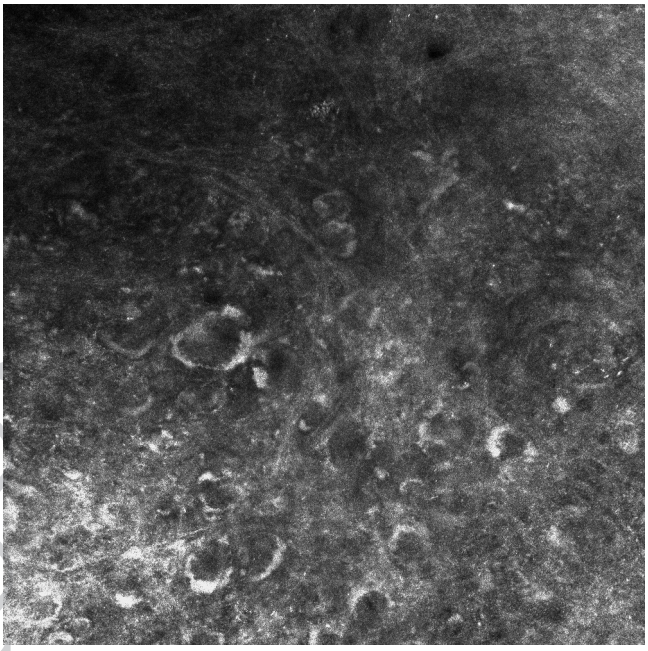

Figure 3 Reflectance confocal microscopy of a lesion showing multiple roundish giant cells characterised by a dark centre surrounded by a highly hyper-refractile peripheral ring in the dermis.

in different lesions showed a normal epidermis with a typical honeycomb pattern, and the constant presence in the dermis of multiple roundish giant cells characterised by a dark centre surrounded by a highly hyper-refractile peripheral ring (figure 3).

Histopathological examination from a skin biopsy of a lesion of the arm showed the presence of a dense granulomatous dermal infiltrate composed of a mixture of foamy histiocytes, multinucleated giant cells (Touton cells), lymphocytes and scattered eosinophils (figure 4).

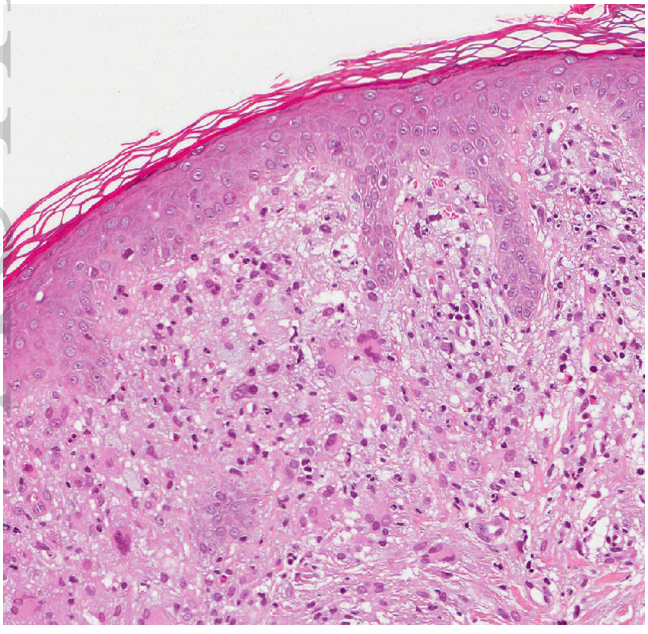

Figure 4 Histopathological examination showing a dense granulomatous dermal infiltrate composed of a mixture of foamy histiocytes, multinucleated giant cells (Touton cells) and lymphocytes.

Based on anamnestic, clinical, instrumental and laboratory data, the diagnosis of multiple XGs was made.

Treatment and outcome

The patient started isotretinoin 20 mg once daily. After 6 months, some lesions flattened, leading to yellowish or hyperpigmented macules, but new lesions raised up. Laboratory tests were repeated and no abnormalities were detected. The patient refused to continue the treatment.

DISCUSSION

Multiple XGs in adult is a rare and poorly understood entity. The differential diagnosis includes a variety of cutaneous lesions, mainly melanocytic nevi, molluscum contagiosum and sebaceous hyperplasia.

PLD and RCM are useful tools for the in vivo diagnosis of XG.[7–12] In our case, the dermoscopy and RCM features were similar to that reported in single lesions of XG.[7–12] The homogeneous orange-yellowish hue seen at dermoscopy is related to the histopathological presence of foamy histiocytes and Touton giant cells; the last typically appear at RCM as peculiar giant cells surrounded by highly refractive ring due to the cytoplasm rich of lipids. The 'setting sun' appearance observed dermoscopically in some lesions results from the presence of a thin erythematous border encircling the homogeneous orange-yellowish hue. This pattern, usually found in early stages of XG, has been also described in other skin lesions with a xanthogranulomatous dermal infiltrate.[13]

Although many articles highlight the benign natural history of adult-onset XG, associations with haematologic malignancies are reported, either before, concurrently or after the development of the cutaneous lesions.[2 3] They include thrombocytosis, chronic lymphocytic leukaemia and monoclonal gammopathy. Thus, it is preferable for patients to undergo a thorough systemic evaluation.

Treatment of multiple XG includes surgical excisions, CO_2 laser and systemic retinoids, especially isotretinoin.[2] In our case, the treatment with systemic isotretinoin was unsatisfactory, so further studies are required to evaluate its effectiveness.

Learning points

- Multiple xanthogranulomas in adult is a rare and poorly understood entity.
- Dermoscopy and reflectance confocal microscopy of xanthogranulomas show peculiar aspects.
- A patient with multiple xanthogranulomas should undergo a thorough systemic evaluation for the possible association with haematologic malignancies.
- Possible treatments of multiple xanthogranulomas include surgical excisions, CO_2 laser and systemic retinoids, especially isotretinoina.

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