CASE REPORT

Confluent and reticulated papillomatosis: Case series of 3 patients from Kedah, Malaysia and literature review

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SUMMARY
Confluent and reticulated papillomatosis (CRP) was first described in 1927 by Gougerot and further characterised by Carteaud. It is relatively rare, and the exact pathophysiology was not well known. Over the years, multiple treatment modalities were proposed.

We report our experience with three cases of CRP which showed complete clearance with tetracycline group of antibiotics.

INTRODUCTION
Confluent and reticulated papillomatosis (CRP) of Gougerot and Carteaud is a relatively rare dermatosis of unknown aetiology. It is characterized by persistent papules and plaques that are confluent in the center and reticulated at the periphery, typically distributed around the neck, interscapular region, infra-mammary area, and the abdomen. There are numerous therapeutic options available with variable results.

CASE REPORT
We report 3 cases of CRP that were alleviated by Tetracycline group of antibiotics.

Case 1 was a 21-year-old Malay female who had multiple asymptomatic brownish papules and plaques on the intermammary region, abdomen, neck and back (interscapular region) of 2-years duration (Figure 1).

Case 2 was a 17-year-old Malay girl who had non-pruritic, painless brownish papules and mildly scaly plaques on the abdomen, neck and back of 5 years' duration, in which they are confluent in the centre and reticulated at the periphery of the lesions.

Case 3 was a 17-year-old Chinese male patient who had asymptomatic non-velvety brownish papules and plaques on the abdomen of 3 year's duration.

DM screening and thyroid function test were normal for all 3 patients, only the first patient was overweight with BMI 27, 2nd and 3rd patients' BMI were < 25. Potassium hydroxide staining of the scale and PAS stain were negative in all 3 cases. Biopsy specimens from their cutaneous lesions revealed hyperkeratosis, papillomatosis and acanthosis, in keeping with the diagnosis of CRP (Figure 2).

Multiple courses of topical and systemic antifungal were tried before giving tetracycline, however lesions were not improved. Case 1 patient was then treated with oral minocycline 100 mg daily for 8 weeks; 2nd and 3rd patients were given oral Doxycycline 100mg 2 times per day for 12 weeks (as minocycline was out of supply at that moment). All their lesions were completely cleared with no relapse observed throughout the follow up until now. The patients' demographic, duration and treatment of the disease are summarized in Table I.

DISCUSSION
Confluent and reticulated papillomatosis is not only rare, but also poorly recognized by physicians. Frequently, the diagnosis is delayed and thus the treatment. Clinically, the eruption is most often confused with Tinea versicolor, acanthosis nigricans and Darier’s disease. Acanthosis nigricans is usually associated with increased body habitus or a history of insulin resistance; however these comorbidities may co-exist with CRP. If pruritus is present, prurigo pigmentosa has to be considered, with a skin biopsy showing lymphocytic infiltrate and multiple necrotic keratinocytes. Darier’s disease is a differential diagnosis of CRP without pigmented changes, classically over seborrhoeic areas, with palmoplantar pits and nail changes.

CRP occurs predominantly in young adults, primarily in their teens, with no particular sex predilection. Commonly they presented with cosmetically displeasing brown scaling patches and affecting anterior aspect of the torso, lesions may extend superoinferiorly in a rhomboidal fashion to involve the anterior neck; lower abdomen, and pubic region. Posteriorly, CRP tends to start off congregating over the interscapular area and extend similarly in a rhomboidal pattern cephalocaudally involving the shoulders, up to the nape of neck and down to the natal cleft. Sometimes, lesions can be seen in the upper limbs including the antecubital fossae and lower limbs including the popliteal fossae. It is characterized by persistent papules and plaques that are confluent in the center and reticulated at the periphery. Most of the patients were asymptomatic, some may complaint of itchiness. CRP has a chronic course, with mean duration of skin eruption at presentation around 3 years; however spontaneous resolution and subsequent recurrence can also occur with or without treatment.

The proposed diagnostic criteria by Davis et al ¹ are: i) clinical findings of scaling brown macules and patches, some

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Retinulated and papillomatous; (ii) location on the upper trunk and neck; (iii) fungal staining of scale negative for spores and hyphae; (iv) lack of response to antifungals; and (v) excellent response to minocycline. All 3 patients have fulfilled the criteria for the diagnosis of CRP.

Most common histopathological findings are hyperkeratosis, papillomatosis and acanthosis. The dermis may contain perivascular lymphocytic infiltrates, mild dilatation of superficial dermal blood vessels, beading of elastic fibers and hypermelanosis of basal layer.2

The pathophysiology is not precisely known, but two prominent theories have been proposed - abnormal host response to fungi/bacteria and abnormal keratinization. Other explanations include photosensitivity, endocrinological imbalance and genetic factors. Recent reports suggested that bacterial superantigens could induce cell adhesion molecule expression in keratinocytes through the induction of various cytokines.3,5

Various treatment modalities such as antibiotics, antifungal agents, selenium sulphide, vitamin A derivatives, salicylic acid and vitamin D derivatives have been proposed. However, responses to those modalities have been unsatisfactory and inconsistent. Recently, treatment with various antibiotics especially minocycline and other macrolides has been reported to be highly effective in CRP patients.3 The good response may be related to anti-inflammatory (most probably attributed to inhibiting neutrophil migration and subsequent reactive oxygen species release and inhibit matrix metalloproteinases) rather than antimicrobial effects alone.

CONCLUSION

The diagnosis of CRP requires a high index of suspicion. Complete clearance with tetracycline group of antibiotics in our case series further support antibiotic as the treatment of choice for CRP. Empirical treatment with tetracycline may be a clue to diagnosis.

REFERENCES


Table I: Summary of the patient’s sex, age, duration, treatment and follow up duration before the lesions were cleared

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex / Age (year)</th>
<th>BMI</th>
<th>Duration (year)</th>
<th>Treatment</th>
<th>Dose (mg/day) / Duration (week)</th>
<th>Follow-up (month)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Female / 21</td>
<td>27.2</td>
<td>2</td>
<td>Minocycline</td>
<td>100 / 8</td>
<td>6</td>
</tr>
<tr>
<td>2</td>
<td>Female / 17</td>
<td>22.5</td>
<td>5</td>
<td>Doxycycline</td>
<td>200 / 12</td>
<td>7</td>
</tr>
<tr>
<td>3</td>
<td>Male / 17</td>
<td>23.3</td>
<td>3</td>
<td>Doxycycline</td>
<td>200 / 12</td>
<td>5</td>
</tr>
</tbody>
</table>

Fig 1a & 1b: Pre and post treatment (Case 1).

Fig. 2: HPE revealed hyperkeratosis, papillomatosis and acanthosis.