A case of Autoimmune Progesterone Dermatitis seen in an adolescent gynaecology clinic

Dann P, O'Mahony F
Royal Stoke University Hospital, United Kingdom

Introduction

Autoimmune Progesterone Dermatitis is a rare dermatological condition, usually presenting with cutaneous or mucosal lesions that manifest in a cyclical manner in relation to a woman's menstrual cycle - specifically in the luteal phase in response to elevated progesterone levels. The diagnosis of this condition is often delayed due to a number of different factors, but when diagnosed, the therapeutic options available have proven to be successful in symptom control. Case reports may need to be obtained before a diagnosis is confirmed and appropriate treatment initiated. There is a lack of literature describing this condition, especially written from a gynaecological perspective, which ultimately is the specialty that initiates treatment and arranges follow up for these patients.

Case report

A 13 year old female initially presented to the paediatric emergency department with a non-blanching ecchymotic rash localised to her left shoulder, measuring 9.5 x 4cm. There were no associated systemic symptoms, suggestive of other autoimmune conditions and she was clinically well in herself. After numerous presentations, the history became clearer and outpatient paediatric and dermatology appointments led to further investigation and sought specialist opinions. This rash occurred in a cyclical manner, just prior to her monthly menstrual cycle and gradually faded spontaneously within a few days of her menses starting. All investigations, predominantly blood tests, were normal, with extensive autoimmune screens and extensive clotting profiles revealing no abnormality. A punch biopsy taken at a time when the rash was present showed florid interstitial dermal oedema, not directing towards a specific diagnosis, but raising the suspicion of artefact dermatitis, despite the patient denying any traumatic skin interference.

Focusing more on the history of this rash manifesting at the time of the menstrual cycle, the possible diagnosis of autoimmune progesterone dermatitis was considered. Intradermal injections of progesterone (Gestone 0.1ml [50mg/ml]) administered into the flexor aspect of both forearms provoked an erythematous local reaction on the same day, but at the time of medical review the following day, the erythema had subsided, leaving only a degree of induration present at the injection sites.

These findings were considered a possible positive result, opposed to a diagnostic result but tailored the hormonal treatment trialled. A combined oral contraceptive pill (Levest) was commenced. Subsequent follow up revealed that the rash no longer developed at the time of menstruation, as it had done previously. The adolescent gynaecology team were involved once treatment was initiated, but not until the patient was aged 16 years. The main objectives within this clinic were to monitor the response to this treatment, provide counselling on the use of a combined contraceptive pill and advise to avoid exogenous progesterone in the future. After 18 months follow up, this patient was discharged from the specialist clinic but with the advice to return if her contraceptive needs changed or the symptoms recurred.

Discussion

This case is similar to other reported cases of autoimmune progesterone dermatitis, in which skin eruption manifests in a cyclical nature in relation to a female’s menstrual cycle. The clinical presentation of this rash corresponds with the increasing levels of progesterone during the luteal phase of the menstrual cycle and noting this correlation, hormonal treatments that suppress ovulation and consequently inhibit this hormonal stimulus by decreasing endogenous progesterone levels, have proven to be effective in some cases. However, the role of typical dermatological treatments such as antihistamines and topical steroids are limited, particularly when used as monotherapy, as the underlying mechanisms are not addressed. Once it is noticed that the skin lesions are refractory to such treatments and in conjunction with a cyclical aggravation of symptoms, hormonal treatments are essential in management. The combined contraceptive pill has been used frequently as the initial therapy and reported successful in some cases, yet due to it containing progesterone its success is not consistently evident. Gondatrophin-releasing hormone (GnRH) agonists have also been used and with reported success, but as with Tamoxifen, carry a higher side-effect profile, predominantly oestrogen deficiency.

A lack of research about this condition, means the exact pathogenesis is not completely understood, with most of the evidence comprising case studies, which report varying success of different treatment modalities trialled and conflicting reports of development of a rash only after exogenous oestrogen. Further research is required to further develop: understanding of the pathogenesis of this condition, its behaviour in pregnancy and at other times of hormonal imbalance and ultimately the optimal way of managing this condition. Unlike other cases which have not been identified until adulthood, in the case described, the diagnosis was made relatively quickly in adolescence. However from the time of presentation, the diagnosis was not considered for 8months, treatment not initiated until 1year later and gynaecology follow up with hormonal counselling not until 2years after presentation; this highlighting an aspect of patient care which can be improved.

Conclusion

- It is essential to develop a differential diagnosis with regards to specific features of the presenting history, which will then direct appropriate investigations and lead to prompt recognition of the condition.
- Involvement of different specialties optimised this patient’s care with regards to diagnostic investigations, treatment and follow up; however, the lack of an immediate diagnosis lead to numerous presentations and ultimately a delay in informed treatment.

References