Erythema Marginatum Rheumatica

Erythema marginatum rheumatica is a rash that is associated with acute rheumatic fever. Rheumatic fever is a multisystem disease that occurs after infection with a Lancefield group A streptococcus.

The rash represents one of the major Jones’ criteria for the diagnosis of rheumatic fever.[1] The Jones’ criteria date back to 1944 but were modified in 1992.[2]

The term erythema annulare is sometimes used for erythema marginatum but erythema annulare centrifugum is classified as one of the figurate or gyrate erythemas. It may be due to a hypersensitivity, to malignancy, infection, drugs, or chemicals, or it may be idiopathic. Erythema marginatum is really, by definition, associated with rheumatic fever.

Epidemiology

In developed countries, rheumatic fever has become very rare and this rash occurs in fewer than 5% of cases of rheumatic fever. However, rheumatic fever incidence in New Zealand has failed to decrease since the 1980s and remains one of the highest reported in a developed country, especially amongst Maori children.[3]

In the 1990s there appeared to be a resurgence of rheumatic fever in the USA.[4] A large series from Pittsburgh reported erythema marginatum as being uncommon in patients with rheumatic fever.[5]

Presentation

One of the best descriptions of erythema marginatum was given by Professor Perry of Bristol. It was based on a case series published in Archives of Disease in Childhood.[6] This is detailed below, under 'Further reading & references'.

Characteristically the eruption starts as a solid erythema, little, if at all, raised. It gradually spreads out and, as it does so, the skin in the centre of lesions returns to normal, thus forming the typical spreading marginate or annular eruption. Where the spreading circles of the rash meet they coalesce forming a larger ring. The usual sites of its occurrence are on the front of the abdomen and front and back of the chest. It can develop on the limbs but almost never on the face. It may, and usually does, appear at the onset of an acute attack or relapse of rheumatism but it is frequently present when there are no other signs of active infection and the ESR is normal. Once it has occurred it tends to come and go lasting from one or two days to months or years.

It may appear with subcutaneous nodules which are firm, painless lumps, mainly on the hands, feet, occiput and back.[7] They are usually 0.5 to 2 cm in diameter and often found in crops of about three, appearing two to three weeks after the onset of fever. The rash occurs early in the disease and remains long past the resolution of other symptoms.

Although uncommon, it continues to be reported and a recent case report describes its occurrence in a case of rheumatic chorea.[8]

Differential diagnosis

Consider drug reactions. It may look like urticaria that can also change quite rapidly but in erythema marginatum there is no pruritus.

Erythema marginatum is also recognised as the classical skin finding in hereditary angio-oedema.[9]
Investigations

Diagnosis is based on presence of additional clinical features suggestive of acute rheumatic fever, using modified Jones' criteria.

Evidence of streptococcal infection is the essential criterion and this can be established by:

- Throat swab which grows Group A beta-haemolytic streptococcus.
- Raised or increasing streptococcal antibody titre.

In uncertain cases, skin biopsy may allow early diagnosis.\[10]\]

Associated diseases

It may be associated with carditis, arthritis, fever and Sydenham's chorea.

Management

There is no specific management of the rash but rheumatic fever must be treated as described in the Rheumatic Fever article. If the diagnosis is suspected, it is wise to start a full course of penicillin as for rheumatic fever.

Complications

There are no specific complications of the rash but complications such as cardiac disease and Sydenham's chorea may occur as a result of the rheumatic fever.

Prognosis

As for rheumatic fever.

Prevention

As for rheumatic fever.

Further reading & references

1. Jones TD; Diagnosis of rheumatic fever. JAMA 1944; 126: 481-85
7. Erythema marginatum; Medical Pictures

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