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] and again in 2015.[3]

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.

Erythema marginatum rheumatica epidemiology

In most developed countries, rheumatic fever has become very rare. Erythema marginatum rheumatica occurs in fewer than 6% of cases of rheumatic fever.[4] However, rheumatic fever remains prevalent amongst Aboriginal and Torres Strait Islander communities in Australia,[5] and amongst Māori and Pacific populations in New Zealand.[6]

In the 1990s there appeared to be a resurgence of rheumatic fever in the USA.[7] A large series from Pittsburgh reported erythema marginatum as being uncommon in patients with rheumatic fever.[8]

Erythema marginatum rheumatica symptoms (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.[9] This is detailed below, under 'Further reading & references'.

Characteristically the eruption starts as an erythematous macule. 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, or a serpiginous rash.

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.

Subcutaneous nodules may also appear in rheumatic fever. They are closely associated with carditis. [10]

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. [11]

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. [12]

Associated diseases

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

Erythema marginatum rheumatica treatment and 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.

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