

Erythema Multiforme

Erythema Multiforme (EM) is an acute, self limiting, inflammatory skin eruption. The rash is made of spots that are red welts, sometimes with purple or blistered areas in the center. It often also affects the mouth, eyes and other moist surfaces. Erythema multiforme has been so named because of the “multiple forms” it appears in- there is a large degree of variety in its clinical presentation. This variation has led to EM being divided into two overlapping subgroups (EM minor and Stevens-Johnson syndrome). These are different faces of the same disease.

EM is relatively common problem for a dermatologist. Half the cases are in young people (under 20). It is rare both under the age of 3 and over the age of 50. Males are slightly more affected than females and there is no racial predilection. One third of EM sufferers will suffer from a recurrence of the disease. Seasonal epidemics are common.

Erythema multiforme minor: EM minor as the name suggests is the less severe of the two types and accounts for 80% of EM. The rash appears over a few days, however in some patients several crops follow each other during one attack. There may be minor burning or itch. It is most intense over the backs of the hands and feet. There are usually round “bull’s eye” target shaped rings on the palms. In severe cases some blisters may be present both on the skin and lips. . The rash lasts for 1 to 2 weeks and then recedes leaving residual brown pigmentation.

The main cause of EM minor is the herpes virus, either as a cold sore, genital herpes or as a hidden infection. Other cases are due to other bacterial or viral infections or reactions to medications. If the infection returns at a later date, or the medication taken again, the rash will soon follow.

Stevens Johnson syndrome: In Stevens Johnson syndrome (SJS) the degree of damage is greater than in EM minor. In 5% to 15% of the cases it results in death, although this number has probably come down with modern treatments. There are some large blood blisters, along with the red rash. The lips eyes and other moist surfaces are more severely affected and occasionally the internal organs. Pain interferes with eating and going to the bathroom.

SJS often appears following symptoms of fever, malaise, cough and sore throat for 1 to 14 days. These are usually (incorrectly) blamed on a virus or infection. When the rash suddenly appears it is often initially blamed (incorrectly) on whatever was given for the “virus”. Because it is rare, it is usually a dermatologist who makes the correct diagnosis. A skin biopsy is taken to confirm it.

Medications are still thought to be the main cause of EM, despite the fact that they have often been falsely accused. However many drugs are trigger factors and a strong causative link has been found between antibiotics (especially sulfa drugs and penicillin’s), NSAID’s (drugs like Advil, Motrin and Naprosyn) and anti seizure medications (especially Dilantin and Phenobarbital).

How and why EM develops is incompletely understood. It appears to be caused by an immune system malfunction. Severe forms of SJS are definitely more common in those with AIDS and other conditions that lead to a weakened immune system. If the same medication that caused the SJS is taken again in the future, the eruption returns. The second time it often shows up in hours rather than days.

Treatment

Treatment of Erythema multiforme begins with identification and removal of the trigger factor, however that is not always possible. EM minor is typically asymptomatic and therefore needs no treatment, as the lesions will clear up by themselves within 2-4 weeks. In herpes virus induced EM minor, acyclovir or Valtrex pills will help, but only if started in the first few days. If the EM keeps recurring, a continuous low dose of acyclovir or Valtrex will prevent it. Systemic therapy to treat SJS used to be limited to supportive measures, such as placement in a burn unit. We now know that if caught early, intravenous Cytoxan, pooled gamma globulin or oral Neoral are dramatically helpful. An older treatment, oral steroids (such as prednisone) also worked. It should not be used because when given to with patients with severe mouth and throat sores it causes them to succumb more readily to fatal respiratory infections. For a fully developed case of SJS, transfer to a burn unit still may be needed.