

Can Necrotizing Fasciitis be the First Symptom of Hyperimmunoglobulin E Syndrome?

Sir,

Necrotizing fasciitis is a disease characterized by destruction of the skin, subcutaneous fat and fascia with or without inflammation of the muscle tissue.¹ Hyperimmunoglobulin E syndrome (HIES) is a rare, multisystem disorder that is characterized by cutaneous and sinopulmonary infections, dermatitis, and elevated serum IgE levels.²

A 34 days of age female infant presented to our hospital with erythema, swelling and discharge from the mammary region. The patient's medical history included frequent manipulation of the breast by the mother. Lesions between 1 and 10 cm in diameter with purulent discharge, destruction of skin, subcutaneous fat, and fascia were noted. *Staphylococcus aureus* was isolated from swab of wound exudate. Treatment consisted of antibiotics, debridement of all dead and dying tissue, followed by daily dressing of the wound. The patient underwent placement of split thickness skin graft. The lesions markedly recovered two months after discharge. Immunological studies revealed all normal values whereas Ig E level was 75.3 IU/ml (Range:15-32 IU/ml).

In outpatient follow up, she was treated multiple times for recurrent wheezing and itchy dermatitis particularly noticeable on face, scalp and neck. At 11 months of age, she was rehospitalized for pneumonia. Immunological studies were repeated where Ig E level was 13000 IU/ml with marked peripheral eosinophilia. The patient was diagnosed as hyperimmunoglobulin E syndrome.

Necrotizing fasciitis is characterized by a bacterial infection of the skin, subcutaneous fat, and superficial and deep fascia, with rapid dissemination and signs of systemic toxicity.³ Breast hypertrophy may occur as a result of elevated circulating maternal endogenous steroid hormones in late gestation. It may be associated with discharge from the nipples known as "witch's milk." Repeated manipulation of the breast can exacerbate the condition and may cause mastitis and abscess formation.⁴ In some rural areas, there is an interesting opinion that breast hypertrophy and witch's milk can be harmful to the baby and breast massage is to be applied.

The classic triad of HIES also known as Job's syndrome (OMIM #243700) includes recurrent cutaneous and sinopulmonary infections, and elevated serum IgE levels. Serum IgE levels in HIES are elevated over ten times the normal limit but there is no correlation between the IgE levels and the severity of the disease.⁵ The patient initially had slightly elevated IgE level in the newborn period. The first clinical finding was necrotizing fasciitis with satellite lesions. From this period onwards, respiratory or cutaneous complaints were continuous.

Mastitis may be initiating event for necrotizing fasciitis. The families in rural areas should be informed about harmful effects of breast massage in newborn babies. We consider that underlying immunodeficiency played role in development of skin lesions in our patient. Normal or slightly elevated levels of immunoglobulin E do not rule out HIES in neonates and Job's syndrome should be in mind as part of the differential diagnosis when immunodeficiencies are considered.

Although HIES is a rare condition, progressive infections in early infancy should raise a suspicion, which deserves further evaluation for detecting the syndrome.

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