BENIGN RHEUMATOID NODULES OF CHILDHOOD*

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The nodules associated with rheumatoid arthritis and rheumatic fever appear with other signs of active rheumatic disease. Rheumatoid nodule-like lesions irrelevant to rheumatoid disease occasionally occur in children who are well and have no complaints associated with rheumatoid diseases. Laboratory tests are normal. Children with benign rheumatoid nodule are not at increased risk for rheumatic disease. No therapy or prophylaxis is required. We present a two-year-old girl with a subcutaneous nodule on the right pretibial region who was diagnosed with clinical and histological findings. *Key words: benign rheumatoid nodule.*

The rheumatoid nodule is the most characteristic histopathological lesion of rheumatoid arthritis[1]. Nodules occur particularly over extensor surfaces and the metacarpophalangeal and interphalangeal joints. They may also involve pericardial, pleural, peritoneal, and endocardial tissues, lungs, gastrointestinal system, central nervous system and kidneys. Subcutaneous rheumatoid nodules are found in about 25 percent of rheumatoid arthritis cases. These nodules are generally accompanied by positive test result for rheumatoid factor and symptoms of rheumatoid arthritis[2-6]. Subcutaneous nodules with the histopathological features of rheumatoid nodules can occur in both rheumatic and non-rheumatic diseases[1,7-10].

The terms benign rheumatoid nodule and pseudorheumatoid nodule are used for nodules localized in the subcutis that mimic rheumatoid nodules histologically but develop in the absence of rheumatoid arthritis or systemic disease. These nodules have also been considered a subcutaneous variant of granuloma annulare. The subsequent development of rheumatoid arthritis occurs infrequently in adults but rarely in children[11].

In this report, we describe a benign rheumatoid nodule in a two-year-old girl without rheumatoid disease.

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Case Report

A two-year-old girl was admitted to hospital because of a two-month enduration on her right pretibial region. There was no pain or limitation of motion. There was no history of trauma in this area. Physical examination was normal. Complete blood count, sedimentation rate, ASO, C-reactive protein (C-RP), latex, antinuclear antibodies (ANA), anti-DNA and immunglobulin values were in normal limits. A provisional differential diagnosis of soft tissue tumor/leiomyoma was considered. The lesion was totally excised without skin. Biopsy specimen was 2.5 x 1.5 x 1 cm. It was fixed in 10 percent formalin and tissue sections were stained with hematoxylin-eosin, Masson's trichrome and alcian blue. Microscopic examination showed that the central fibrinoid necrosis was surrounded by radially oriented mononuclear cells, and a marginal zone of vascular connective tissue was seen around these cells (Fig. 1). The fibrinoid necrosis was stained red-blue with Masson's trichrome and was stained dark blue in some areas with alcian blue (Fig. 2). Special stains were negative for fungi and mycobacteria. Benign rheumatoid nodule was diagnosed with clinical and histological findings. The patilent was healthy and without complaint after one year.

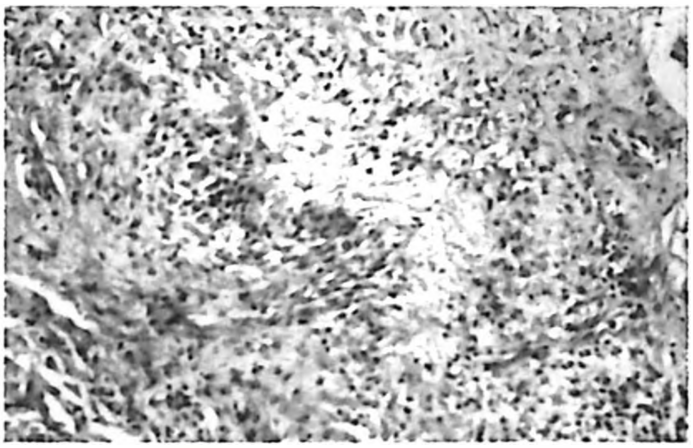

Fig. 2: Fibrinoid necrosis staining red-blue (Masson's trichrome x 80).

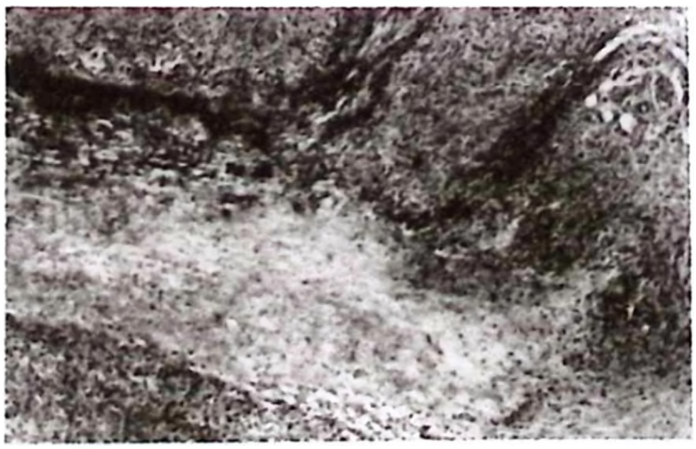

Fig. 1: Rheumatoid nodule (hematoxylin-eosin x 80).

Discussion

The etiology of benign rheumatoid nodule is unknown. Single or multiple lesions may be present over various sites, including pretibial areas, dorsa of the feet, hands, scalp and elbows. They may appear over pressure points or after trauma as do true rheumatoid nodules.

Affected children are well and have no associated rheumatic complaints. Laboratory tests of active rheumatic disease are normal. Tests for rheumatoid factor and ANA are negative[9,12]. "Hidden" rheumatoid factor has been found with higher titers than in controls. The nodular lesions may recur, but recurrences eventually cease, although it could be after months or years[8,11].

Subcutaneous nodules are characterized by foci of fibrinoid necrosis surrounded by palisading histiocytes and fibroblasts and a marginal zone of vascular connective tissue usually having chronic inflammatory cells[1]. Patterson[13] studied more closely the histology of subcutaneous granuloma annulare and rheumatoid nodules. He was able to differentiate these lesions by alcian blue and Masson's trichrome staining. Alcian blue, which stains mucin in degenerated collagen, was the most helpful diagnostic histochemical method. Patterson suggested that positive alcian blue staining shows the benign nature of the disease. He demonstrated that rheumatoid nodules stain homogeneous red with Masson's trichrome, whereas the central zone of granuloma annulare stains blue-red[13]. A similar staining pattern was demonstrated in our case.

Rheumatoid nodules can occur in association with rheumatoid arthritis, juvenile rheumatoid arthritis, rheumatic fever and systemic lupus erythematosus. Single or multiple subcutaneous nodules are a major criterion for the diagnosis of rheumatic fever; however, they are very infrequent. They are most commonly observed with severe carditis. Histologically, central, homogeneous fibrinoid necrosis and palisading of histiocytes usually do not develop well in rheumatic fever nodules, and fibrosis is minimal or absent[1,11,12,14]. The clinical and histopathological features and the age of our patient were not compatible with rheumatic fever.

Other subcutaneous nodules that might be considered in the clinical differential diagnosis include erythema nodosum, sarcoid, bone and soft tissue tumors, mycobacterial or fungal infections and the occasional nodules of scleroderma and periarteritis nodosa[8,11]. The clinical features associated with these diseases are usually sufficient to distinguish them.

Children with benign rheumatoid nodule do not have increased risk for rheumatic disease. No therapy or prophylaxis is required[12].

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