

Dermacase

Can you identify this condition?

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A 37-year-old man presented with faintly pink subcutaneous nodules over the extensor surfaces of the proximal interphalangeal joints of his fourth and fifth fingers. These nodules were firm, mobile, and nontender.



The most likely diagnosis is:

1. Dermatomyositis
2. Granuloma annulare
3. Rheumatoid nodules
4. Lichen planus
5. Tophaceous gout

Answer on page 41

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Answer to Dermacase *continued from page 35***3. Rheumatoid nodules**

Rheumatoid nodules are the most common extra-articular manifestation of rheumatoid arthritis (RA); about 25% of adult patients with RA have them.^{1,2} About 90% of patients with RA and subcutaneous nodules test positive for rheumatoid factor, and 40% of all seropositive patients with RA have subcutaneous nodules.³ Rheumatoid nodules are clinical predictors of more severe arthritis, seropositivity, joint erosions, and rheumatoid vasculitis.² The presence of rheumatoid nodules often suggests a need for more aggressive treatment of the underlying RA to prevent sequelae.

Rheumatoid nodules are firm, nontender, and movable within the subcutaneous tissue; however, they could also be attached to underlying structures such as the periosteum, fascia, and tendons.³ The lesions range from 2 mm to >5 cm,⁴ and can enlarge or regress, recur, or persist indefinitely.³ Most subcutaneous nodules are found on bony prominences, extensor surfaces, or adjacent to joints. They are most frequently found on extensor surfaces of the proximal ulna and olecranon, metacarpophalangeal and proximal interphalangeal joints, ischial tuberosities, joints in the foot, and sacrum.⁴ Occasionally, they manifest on the sclera, pinna of ears, heart, vocal cords, lungs, nervous system, abdominal wall, and muscle.^{4,5} Histologically, rheumatoid nodules present as a palisading granulomatous reaction, and mature nodules have a classic three-layer structure.¹

The exact etiology of rheumatoid nodules is unknown. Experts speculate that a series of events beginning with local vascular trauma and pooling of rheumatoid factor immune complexes, followed by activation and mobilization of local monocytes or macrophages; fibrinoid deposition by procoagulants; tissue necrosis by cytotoxins, proteinases and collagenase secretion from macrophages; and chemotactic attraction of macrophages to the necrotic zone is responsible for formation of rheumatoid nodules.^{1,6} This hypothesis is consistent with the clinical findings of higher titres of rheumatoid factor and vasculitis often reported in patients with

RA who have rheumatoid nodules.² Since rheumatoid nodules most commonly arise in areas prone to trauma, a local tissue reaction that creates a focus of granulation tissue might also contribute to initial formation of these lesions.¹

Rheumatoid nodules are not exclusive to RA. Histologically identical nodules are sometimes a feature of systemic lupus erythematosus (SLE), subcutaneous granuloma annulare, necrobiosis lipoidica diabetorum, rheumatic fever, and foreign body granulomas.^{2,3,7,8} Subcutaneous nodules have also been reported in 5% to 10% of children with juvenile rheumatoid arthritis,³ and benign rheumatoid nodules have been described in children and adults with no evidence of RA.^{2,7} Rheumatoid nodulosis syndrome, which presents with numerous rheumatoid nodules, a high titre of rheumatoid factor, but mild or no RA, has been reported in a few patients.^{3,7,8}

Diagnosis

A diagnosis of rheumatoid nodules is made in the clinical context of the disease. Symmetric inflammatory polyarthritis, seropositivity for rheumatoid factor, and other associated symptoms, such as vasculitis, are highly suggestive of RA. Subcutaneous nodules with a history of gout or current podagra could lead to a diagnosis of tophaceous gout. Violaceous papules or nodules (Gottron's papules) with muscle weakness and heliotrope rash are characteristic of dermatomyositis. Lichen planus is a pruritic, papular eruption characterized by its violaceous-purple colour, flat-topped polygonal shape, and sometimes, fine scale. It is most commonly found on the flexor surfaces of the upper extremities (especially wrists), genitalia, and mucous membranes (called Wickham striae in the mouth).

Although biopsies of subcutaneous nodules are occasionally done, they are not useful for diagnosis since many different types of subcutaneous nodules are histologically identical to rheumatoid nodules. Many rheumatoid nodules occur in areas difficult to biopsy, such as over extensor tendons. A complete history and physical examination, focusing on

cutaneous and rheumatologic aspects, and occasionally laboratory testing (eg, rheumatoid factor, serum urate) are sufficient to diagnose rheumatoid nodules.

Management

Rheumatoid nodules typically present asymptotically as a cosmetic complaint. Indications for treatment include areas exposed to repetitive trauma and nodules on weight-bearing prominences that might cause progressive erosions and severe pain, neuropathy, limitation of motion, or deformity, and damage to underlying structures.⁸ Some nodules rupture and lead to deep infections.⁴

There are very few treatment options for rheumatoid nodules. Large nodules can be excised, but they frequently recur within scar tissue, especially if subjected to repetitive trauma. Injecting corticosteroids directly into the lesion sometimes reduces its size. While this procedure is most effective for

deep lesions in the olecranon bursa, nodules on the buttocks and feet tend to ulcerate and are likely to become infected. Once they are infected, surgical excision or drainage is required.⁸ Oral corticosteroids and hydroxychloroquine can also be used,^{3,8} but their effects on rheumatoid nodules vary, as most patients with RA already receive these medications for the chronic condition. Rheumatoid nodules occasionally resolve without medical or surgical intervention. ❁

References

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