Recurrence monoarthritis with tender erythematous nodules in a 28-year-old man: a diagnostic dilemma

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ABSTRACT: Cutaneous polyarteritis nodosa (CPAN) is a rare form of vasculitis that involves small and medium-sized arteries of the dermis and subcutaneous tissue. CPAN should be considered a separate disease entity and distinguished from systemic polyarteritis nodosa (PAN) as the clinical course and management of these conditions differ from each other. While PAN is a vasculitis of medium-sized muscular arteries involving the liver, kidneys, heart, lungs, gastrointestinal tract and musculoskeletal system, and is potentially life-threatening, cutaneous PAN is a localized cutaneous vascular disorder characterized by necrotizing arteritis of medium-sized vessels in the dermis and subcutaneous tissue which usually runs a chronic but benign course. Patients with CPAN usually present with fever, myalgias, arthralgias, neuralgias, and neuropathies, but are usually normotensive and lack life-threatening organ involvement. Moreover, arthritis and weight loss are seldom seen in CPAN, and have rarely been reported in the past in medical literature. We have reported here, a rare case of cutaneous polyarteritis nodosa (CPAN) which presented with vasculitic plaques and recurrent monoarthritis of the right knee joint, accompanied by significant weight loss.

KEYWORDS: Recurrent monoarthritis, painful erythematous nodules, weight loss, polyarteritis nodosa, skin biopsy

We report here our first case of CPAN which initially posed a diagnostic dilemma. The patient was a 28-year-old man who presented with recurrent right knee monoarthritis, vasculitic plaques and painful erythematous nodules over his left lower limb, which was accompanied by weight loss. The diagnosis of CPAN was finally established on skin biopsy.

Case Report

A 28-year-old male presented with complaints of multiple episodes of recurrent monoarthritis of his right knee since the past 4 years. On each occasion, his right knee got badly swollen and was acutely warm and tender. He also complained that, in the past he had developed “fluid” in the right knee joint during two such episodes. There was also history of significant loss of weight of 7 kgs over the past year, despite a good appetite.

On examination, the right knee joint appeared to be warm and acutely tender to touch and the range of movements was significantly limited. However, he gave no history of early morning stiffness and his other joints were normal on examination.

His previous case reports revealed that synovial fluid examination done a year ago did not culture any organisms nor did it show presence of any crystals, and the synovial tissue biopsy done then was also normal.

Assuming then that his recurrent monoarthritis could be due to seronegative spondyloarthropathy, he was started on sulphasalazine, to which he responded.

He was symptom-free for about 8 months while on sulphasalazine when he now developed both, right knee and left wrist arthritis. However, this was not accompanied by any early morning joint stiffness, red eyes or urethral discharge.

Two months later, the patient presented with a three day history of painful erythematous lesions over the lateral malleolar region of his left leg. On examination, he was found to have vasculitic plaques and tender erythematous nodules over the area (fig.1). On further questioning, he revealed that he had had such skin lesions many a time in the past. However, as these skin lesions healed within 1 week without any residual scarring, he did not seek any treatment nor did he report the same to his physician.

There was no history of fever, pustular skin lesions, night sweats or chronic cough. There was also no history of abdominal pain, diarrhoea, melena, numbness in limbs, angina, nasal discharge, oral ulcers or photosensitivity. He did not have any past history suggestive of pulmonary tuberculosis or ischaemic heart
Polyarteritis Nodosa (PAN) is defined as a systemic necrotizing vasculitis affecting medium-sized arteries in the kidneys, liver, heart, skin and gastrointestinal tract. In contrast, CPAN is a vasculitis of the small and medium-sized arteries which is more limited in nature with its findings confined to the skin, musculoskeletal, and nervous systems. The first case of PAN was described in 1866 and was originally named periarteritis nodosa due to the nodular protuberances along medium-sized vessels. In the early 1900s, it was discovered that there were inflammatory cells in all levels of the vessel and the name was subsequently changed to 'polyarteritis nodosa.' PAN was also the first 'vasculitis' to be described. The first case of CPAN was described in 1931. Until that time it was unclear whether CPAN, a benign and less severe form of vasculitis, was a distinct entity. In 1980, investigators confirmed that CPAN was a separate disorder, distinct from systemic PAN.

According to the ACR 1990 criteria for classification of PAN, a patient with vasculitis is said to have PAN if at least 3 of the 10 criteria are present. However, CPAN does not have any specific diagnostic criteria. The distinguishing characteristics are the lack of systemic involvement and its benign, yet relapsing, long-term course. CPAN affects adults as well as children. The reported age of patients with CPAN has ranged from 5-68 years. Presenting symptoms in adults include fever, myalgias, arthralgias, neuralgias, and neuropathies, but patients are usually normotensive and the lack of life-threatening organ involvement helps distinguish this condition from systemic PAN. Typically, the disease is limited to the skin, muscles, and joints. Mononeuritis is sometimes seen and can present as decreased reflexes and paresthesias. Myalgias occur with physical activity in 100% of those affected.

Consequently, CPAN is considered to be a distinct clinical entity characterized by a chronic prolonged course but an overall good prognosis.

The characteristic cutaneous manifestations of CPAN are vasculitic plaques and tender erythematous nodules usually 0.5cm in diameter located on the foot, ankle, and lower part of legs. The lesions turn bluish as they heal and tend to recur in the same location. This is in sharp contrast to the bullae seen in classic PAN. Nodules are unusual in systemic PAN and are thus a useful distinguishing feature. Lower limb ulceration is seen in up to 50% of patients with CPAN.

The diagnosis of both PAN and CPAN is made via skin biopsy. While necrotizing arteritis with neutrophilic infiltration of small and medium-sized arteries is seen in the reticular dermis and subcutaneous tissue in CPAN, necrotizing vasculitis of medium-sized muscular arteries involving the liver, kidneys, heart and gastrointestinal tract is usually the hallmark of systemic PAN.

Arriving at the diagnosis....
Our patient presented with recurrent right knee monoarthritis (and left wrist arthritis, on one occasion) over a 4-year period, which is a rather uncommon presentation for CPAN. This was accompanied by a significant weight loss of 7 kgs, the underlying cause of which could not be determined inspite of a detailed clinical examination and investigations.

The unusual feature in our patient was the recurrence of monoarthritis requiring sulphasalazine. While arthralgias and non-destructive arthritis have been reported in CPAN, an association with recurrent arthritis has rarely been reported in medical literature. So far in only one review, seronegative arthritis was reported. One study reported a patient with CPAN and severe arthritis which resulted in progressive joint destruction requiring arthroplasty. The skin lesion in our patient healed within 1 week without any scarring. Hence, other conditions mimicking the skin lesion such as metastatic tumours and atypical infections were ruled out. His ANCA was negative which made Wegener’s granulomatosis unlikely. Consequently, the differential diagnosis was narrowed down to PAN, sarcoidosis and CPAN.

The diagnosis of both PAN and CPAN is definitively made only on skin biopsy. Hence, at times, a group of patients remain undiagnosed. In our patient too, despite extensive investigations, the diagnosis remained elusive until a skin biopsy was performed. The skin biopsy finally clinched the diagnosis of CPAN when it demonstrated arteritis involving medium-sized vessels in the dermis and subcutis, which was consistent with CPAN.

Consequently, we would like to emphasize that when recurrent monoarthritis in a patient cannot be explained by the more common causes of monoarthritis such as trauma, infection, crystal-induced synovitis, non-inflammatory arthritis and tumours, a differential diagnosis of CPAN should be considered if the patient has a medical history of skin lesions. Moreover, the patient should be regularly followed up for the development of new skin lesions, for timely biopsy, in order to reach a definitive diagnosis of CPAN.

Treatment for CPAN is primarily with systemic corticosteroids. It is paramount to watch for the development of systemic symptoms of PAN as a patient continues to be treated for CPAN, since there have been cases of patients presenting with only cutaneous lesions and not progressing to the systemic form of PAN until 18-19 years later. Remissions can range from months to almost 4 years. Treatment is usually:

1. Suppression of the inflammatory response with the use of non-steroidal anti-inflammatory drugs
2. Modulation of the underlying immune mechanism involved in the vasculitis with the use of corticosteroids
3. Adjunctive agents
4. Colchicine

This case highlights the importance of every clinical symptom and sign in arriving at the diagnosis of CPAN. In our patient, the clinical features were those of cutaneous PAN except for the presence of recurrent monoarthritis and significant weight loss. However, the cause of the weight loss could not be determined inspite of detailed investigations. He was treated only with glucocorticoids without any cyclophosphamide as he did not have any critical organ involvement. Moreover, he will continue to be closely monitored for systemic complications, which could develop years later.

Conclusions

PAN and CPAN are two rare vasculitides that are manageable and treatable if the signs and symptoms are recognized by the astute clinician. Since clinical findings are highly variable, it is often necessary to individualize therapy. Hence, the treating physician should be well-aware of the wide clinical spectrum of vasculitides, for not all patients have cutaneous findings and not all have systemic components as well. Patients with CPAN may initially present with only joint pain, so an awareness of the various cutaneous manifestations is important to enable early diagnosis and prompt management of this condition.

We have reported here, a rare case of cutaneous polyarteritis nodosa (CPAN) which presented with vasculitic plaques and recurrent monoarthritis of the right knee joint, accompanied by significant weight loss.

REFERENCES