Focus

Spondyloarthropathies and their diagnosis

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Key points

- The spondyloarthropathies are a group of related rheumatic conditions sharing a variety of common extra-articular features
- A careful history and examination are usually sufficient to arrive at a diagnosis
- Testing for the presence of HLA B27 is of limited clinical value
- NSAIDs remain the basis of pharmacological treatment

The spondyloarthropathies are a diverse group of rheumatic conditions, important in a number of respects. Firstly, they are common; as a group, they have a prevalence at least as great as that of rheumatoid arthritis. Secondly, there are specific issues in relation to therapy and prognosis that differ significantly from conditions they may be confused with. For instance, they enter into the differential diagnosis of common musculoskeletal presentations such as low back pain or tendonitis. Thirdly, for over a quarter of a century they have provided a tantalising clue as to the interaction of environmental triggers such as infection and genetic predisposition in the aetiopathogenesis of arthritis in particular and chronic autoimmune conditions in general.

While recognised individually prior to this, it was only in 1974 that Moll and Wright first drew attention to the association between ankylosing spondylitis, psoriatic arthritis, Reiter's disease, the intestinal arthropathies and Behçet's syndrome. While the association with Behçet's syndrome is no longer widely recognised, this group has been joined by a diagnosis of undifferentiated spondyloarthropathy while Reiter's syndrome has been redefined as reactive arthritis. Before considering the classical presentations of the individual members of this group, it is useful to think of their clinical features in four dimensions.

CLINICAL FEATURES

Peripheral arthritis syndrome

Lower limb involvement with an asymmetrical distribution is common. The knee and contralateral ankle, for instance, can be involved. Synovitis and effusion may be quite pronounced. Symmetrical, small joint involvement including the MCPJs of the hand is found, and may be difficult to distinguish from rheumatoid arthritis. Involvement of the synovial lining of tendon sheaths is also common, a characteristic presentation being a sausage shaped swelling of entire digits, called dactylitis.

Enthesopathic syndrome

The enthesis is the point of attachment of tendons, ligament or joint capsule to bone. Enthesitis is a distinctive pathological feature of the spondyloarthropathies. It

is most frequently recognised at the heel as plantar fasciitis, but it may be the initial event in the subsequent development of tendonitis or arthritis.

Pelvic and axial syndrome

Sacroilitiis is the cardinal feature of ankylosing spondylitis but also occurs in other members of the group. Just why the sacroiliac joint should be so frequently involved is poorly understood. Whatever the reason, the distinctive features of sacroiliitis allow differentiation from the much more common mechanical low back pain that affects us all. The factors suggestive of inflammatory back pain are:

- onset of back discomfort before the age of 40
- insidious onset
- persistence for at least three months
- associated with significant morning stiffness lasting over an hour
- improvement with exercise.

Extra-articular features

Psoriasis is the most common extra-articular feature, usually as psoriasis vulgaris but pustular psoriasis and guttate psoriasis may also occur. Not infrequently, lesions of psoriasis may be occult and observed only on careful examination of the characteristic sites including the hairline, periumbilical and natal cleft areas. Fingernail pitting may offer an early clue. Lesions very similar to psoriasis can occur acutely in cases of reactive arthritis when they are found on the palms and soles as keratoderma blennorrhagica.

Ocular involvement can vary from the conjunctivitis of reactive arthritis ("Reiter's syndrome") through to acute uveitis. There is a suggestion that uveitis itself may be a manifestation of the spondylo-arthropathies and related to the presence of HLA B27.The association with **inflammatory bowel disease** such as Crohn's disease or ulcerative colitis is well recognised. There is mounting interest in the observation that patients with ankylosing spondylitis may also have occult colitis at a microscopic level.

Cardiac involvement may also occur, and includes manifestations such as heart block and isolated aortic regurgitation.

Urogenital symptoms, while commonly associated with the sexually acquired form of reactive arthritis, may also occur following enteropathic infection suggesting that urethral inflammation may be a result of mechanisms other than direct infection.

<table>
<tr><th colspan="3">EUROPEAN SPONDYLOARTHROPATHY STUDY GROUP PRELIMINARY CRITERIA FOR THE CLASSIFICATION OF SPONDYLOARTHROPATHY</th></tr>
<tr><td>Inflammatory spinal pain</td><td>or

and one or more of the following</td><td>Synovitis: asymmetric predominant in the lower limbs</td></tr>
<tr><td colspan="3">Positive family history
Inflammatory bowel disease
Urethritis, cervicitis or acute diarrhoea within one month before arthritis
Buttock pain alternating between right and left gluteal areas
Enthesopathy
Sacroiliitis</td></tr>
</table>

CLASSIFICATION and DIAGNOSIS

The concept of spondyloarthropathy is based essentially on clinical grounds, but it remains a working concept subject to review. Patients with classical ankylosing spondylitis or psoriatic arthritis with involvement of the axial skeleton undoubtedly fit into the group, but there is some debate as to whether all forms of psoriatic arthritis should be included and patients with isolated oligoarticular asymmetrical arthritis might fit classification criteria in the absence of spondylitis. There are two recognised sets of criteria for the classification of spondylo-arthropathy (see box and table).

While both sets of criteria have been put forward as classification criteria they are nonetheless useful in diagnosis. Regrettably, neither are particularly sensitive in early disease when differentiation from rheumatoid arthritis is important.

Ankylosing spondylitis

The typical patient with ankylosing spondylitis is a young man presenting in his 20s with a history of the insidious onset of inflammatory back pain over some months. There is a male predominance, perhaps not quite as much as has been considered in the past; the male to female ratio being about 3:1. The predictive value of the history of inflammatory back pain is low, given the low prevalence of ankylosing spondylitis (0.1 per cent) and the false positive rate of 10-15 per cent in common mechanical back pain.

Physical examination often reveals a stiff, flattened lumbar spine with range of movement reduced in all directions. Radiology in the early stages may not be helpful. In patients with a high level of pretest probability (eg, a family history), the rapid response to the institution of NSAID treatment strongly supports the diagnosis. While no NSAID has been adequately demonstrated to be better than any other, clinical experience suggests agents such as indomethacin may have particular value in treatment. Early referral to physiotherapy for range of movement and back protection exercises is of prime importance.

It is well known that HLA B27 is found in most, if not all, patients with classical ankylosing spondylitis. Seven per cent of the New Zealand population have this gene, and given the low prevalence of ankylosing spondylitis, this test has no positive predictive value in a patient with back pain alone.

TABLE: AMOR CRITERIA FOR THE CLASSIFICATION OF SPONDYLOARTHROPATHIES

	A. Past or current clinical manifestations	Pts
1	Back pain at night and/or back stiffness in the morning	1
2	Asymmetric oligoarthritis	2
3	Gluteal pain without other details Alternating gluteal pain	1 or 2
4	Sausage like digit or toe	2
5	Heel pain or other enthesopathy	2
6	Iritis	2
7	Non-gonococcal urethritis or cervicitis within 1 month before the onset of arthritis	1
8	Diarrhoea within 1 month before onset of arthritis	1
9	Past or current psoriasis and/or balanitis and/or inflammatory bowel disease	2
	B. Roentgenographic changes	

Reactive arthritis

Reiter's syndrome was described in 1916, following an outbreak of *Shigella* dysentery. The triad of conjunctivitis, urethritis and asymmetrical lower limb oligoarthritis was also recognised to occur after

10	Sacroiliitis (stage 2 or more if unilateral)	3
C. Predisposing genetic factors		
11	Presence of the HLA B27 antigen and/or positive family history for ankylosing spondylitis	2
D. Responsiveness to treatment		
12	Improvement within 48 hours after initiation of a non-steriodal anti-inflammatory drug	1
*Patients with a total score of 6 points or more are classified as having a spondyloarthropathy		

non-gonococcal urethritis. *Chlamydia* has become most closely associated with this form of reactive arthritis, also known as SARA (sexually acquired reactive arthritis). Reactive arthritis may go on to involve the sacroiliac joints, the entheses, and demonstrate characteristic mucocutaneous lesions such as circinate balanitis and keratoderma blennorrhagica.

First line treatment is again the use of NSAIDs. Classically, the joint symptoms will settle spontaneously. However, some patients develop a chronic arthritis while others follow a remitting and relapsing course. The issue of antibiotic treatment for reactive arthritis following a definite infective trigger has not as yet been satisfactorily resolved.

Psoriatic arthritis

Psoriatic arthritis occurs perhaps most frequently in people in their third and fourth decades. It is defined as an inflammatory arthritis associated with psoriasis in the absence of a positive rheumatoid factor. It may present as a peripheral arthritis alone or as an axial arthritis involving the sacroiliac joints and spine. Involvement of the distal interphalangeal joints of the hand along with fingernail changes is a characteristic presentation, but symmetrical small joint arthritis can often be indistinguishable from rheumatoid arthritis. An asymmetrical oligoarticular involvement may present with the classical "sausage" digits (dactylitis).

The least common presentation is that of arthritis mutilans. The arthritis of psoriasis may precede the development of cutaneous lesions and there is no relationship between the intensity of the skin lesion and the development or pattern of arthritis. First-line treatment again involves the use of NSAIDs, but the peripheral arthritis may respond to second line agents in a manner similar to rheumatoid arthritis.

Inflammatory bowel disease

Ulcerative colitis and Crohn’s disease are of relatively low prevalence, and arthritis