RHEUMATOLOGY AND REHABILITATION

Edited by
Hedley Berry, Eric Hamilton
and
John Goodwill

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DIAGNOSIS AND MANAGEMENT

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PREFACE

This book represents the course followed by the Undergraduates at King's College Hospital in Rheumatology and Rehabilitation. In view of the present varied state of union between these specialties it has been felt necessary to present the book in two separate halves. There are numerous centres which for practical reasons find this the easiest way to teach the two disciplines and hence we felt it reasonable to offer a combined book. Whilst other text books cover rheumatology, few have ventured into the minefield of rehabilitation at an undergraduate level. We hope we have gone some way towards offering more information to undergraduates and more senior practitioners by presenting the book in the way we have done. We also hope that the book will be of interest to the many nurses, occupational therapists, physiotherapists, speech therapists, social workers and others who are closely involved in rehabilitation.

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PART ONE:

RHEUMATOLOGY

INTRODUCTION

Hedley Berry

Problems presenting to the Rheumatic clinics are wide and varied. Many patients in such clinics do not suffer from joint disease, but suffer instead from disease of the associated tissues. In this book we consider first the commonest causes of arthritis, namely osteoarthritis and rheumatoid arthritis. Then we turn to the less common causes of arthritis; these include the arthritides in which the rheumatoid factor test is negative (namely the sero-negative arthropathies), diseases associated with infection in the joints (this has become increasingly common of late) and systematic lupus erythematosus (a disease in which the joint pains are associated with a facial rash which makes the patient look a little 'wolf-like'). This latter disease shows *par excellence* the importance of multisystem involvement in conjunction with arthropathies. Polyarteritis nodosa is similar in this respect in that multisystem involvement is often seen without very much in the way of arthropathy present. Gout and pseudogout are then considered in chapter 7: whilst they are both the cause of arthritis of crystal origin, the crystals are different, namely urate and pyrophosphate. Polymyalgia rheumatica and giant cell arteritis are considered next. These two entities appear to be part of an overall disease spectrum and we follow this with discussion of a much rarer condition, progressive systemic sclerosis or scleroderma, a disease where the skin is primarily affected with multisystem involvement. Polymyositis and dermatomyositis also appear to be part of an overall disease spectrum. The outlook of these conditions again appears to have improved as a result of therapy.

Whilst arthritis particularly affects older people, the young also experience an arthropathy and a chapter on Juvenile Chronic Polyarthritis shows the wide range in nature of this affliction. In addition to arthritis other, perhaps even more common, problems present to our clinics including low back pain, which we believe merits a substantial chapter, the painful shoulder, neck pain, soft tissue lesions and sports medicine.

A separate chapter on drug therapy seemed appropriate given the wide number of drugs available and the difficulty in making therapeutic decisions and we hope we have recognised the importance of radiology in diagnosis in the last chapter of this section of the book. We have assumed that readers either already have a knowledge of the anatomy or physiology of joints or will readily be able to find this material elsewhere and so we have not deemed to include an introductory chapter covering these areas.

1 OSTEOARTHRITIS

Sara Rae

Osteoarthritis (OA) is the term which describes degenerative disease affecting diarthrodial joints. The suffix –itis denotes a form of inflammatory process and as this is present at least intermittently during the OA disease process it will be retained here. OA is common in the whole animal kingdom (unlike rheumatoid arthritis, (RA) which occurs in man only). It is found in a wide range of vertebrates from small mammals such as mice to domestic pets such as the alsatian as well as valuable animals such as racehorses.

Incidence

OA is an extremely common disorder in man. Epidemiological studies based on X-ray findings demonstrate that it is present in weight-bearing joints from the end of the second decade of life. Ninety per cent of the over 50 year old population are affected.

Considering its prevalence, the percentage of patients presenting with symptoms of the disorder is relatively small, although the numbers themselves are quite significant. Those patients under 40 years of age presenting with symptoms are mainly men. Over 50 years the predominance is among women and this persists until the end of life. Population studies have failed to demonstrate any race which is resistant to OA, but OA of the hip is less common in the Indians and Chinese-people who commonly adopt a squatting posture. Climate appears to have no effect either – the incidence in Jamaica is the same as in the UK.

Aetiology

OA can be divided into two categories – Primary and Secondary.

Primary OA

In primary OA, no aetiologic agent can be identified; it can affect single joints or it can be generalised. The latter usually affects post menopausal females.

Secondary OA

A variety of causes, listed in Table 1.1 can produce secondary OA. Trauma usually predisposes to localised disease. For instance, miners develop OA in spine and knees, lorry drivers in shoulders, pneumatic tool workers in shoulders, elbows and hands. Footballers develop OA of knees and ankles, ballet dancers OA of feet (especially hallux rigidus) and boxers develop OA of upper limb joints. The more generalised disorders such as acromegaly, chondrocalcinosis (see chapter 7) and the rare, alkaptonuria (cartilage deposition of homogentisic acid due to an inborn error of metabolism), and Wilson's disease (deposition of copper due to deficiency of caeruloplasmin) affect cartilage metabolism and so tend to cause generalised OA. This is true also if the very rare Kashin Beck disease (ingestion of *Fusarium sporotrichella* in parts of China and the Far East), which has a toxic effect on cartilage. It is probably also true of obesity, as obese patients demonstrate OA changes in non-weight-bearing joints such as the acromio-clavicular and sterno-clavicular joints.

Table 1.1: Aetiology of Secondary Osteoarthritis

Trauma	Fractures Occupational Sporting
Endocrine	Acromegaly Diabetes mellitus
Metabolic	Chondrocalcinosis Alkaptonuria Wilson's Disease
Dietary	Obesity Kashin Beck Disease
Inflammatory	Any inflammatory or septic arthritis
Congenital	Hip dysplasia Slipped capital epiphysis Perthe's Disease Lax ligaments
Neuropathic	Charcot's joints Diabetes mellitus Syringomyelia Syphilis
Haematological	Sickle cell Disease Haemophilia

An inflammatory arthritis can result finally in OA by causing a toxic effect on cartilage. Congenital disease produces secondary OA by altering the articular surfaces so that abnormal strain is placed on cartilage. Neuropathic processes produce the same result by predisposing to frequent trauma and weakening muscle power, which reduces stability.

Caissons disease, an occupational hazard in divers, and also sickle cell disease give rise to OA of femoral and humeral heads after aseptic necrosis has occurred. Haemophilia, in which frequent haemarthroses are present, leads to iron deposition in cartilage and hence cartilaginous derangement and OA.

Pathology

The pathological changes of OA are basically the same whatever the aetiological agent. First there is a change in the cartilage itself. As this is the primary event the underlying biochemical process should be explained.

Cartilage comprises three layers – an outer layer of densely packed collagen fibres arranged parallel to the bony surface. A middle layer of more loosely arranged fibres surrounding water-retaining proteoglycan molecules. This layer becomes rearranged under stress to form the main shock-absorbing mechanism. The final layer lining the subchondral bone comprises collagen fibres arranged perpendicular to the bone surface which have the effect of 'cementing' the cartilage onto the bone.

It is not known in which layer the defect begins, but the chondrocytes stop producing adequate proteoglycan molecules. As these are the mainstay of structure, the framework described above alters. This alteration stimulates chondrocytes to multiply, excess proteoglycans are produced which are distributed randomly. They attract water into sites of high collagen content and this disrupts structure further. The softened cartilage then flakes off during movement of the joint.

The detritus can give rise to synovitis which may become chronic but which is more often accompanied by a simple effusion and only minimal synovial inflammation.

As cartilage is worn away the underlying bone becomes exposed and osteoblasts become activated and remodel the subchondral bone to a fine dense consistency similar to ivory, a process called eburnation. At the margin of the joint this process is more haphazard and results in osteophyte formation; overgrowths of bone projecting into the joint space or into adjacent tendons and ligaments.

Eburnated bone is less pliant than ordinary bone and microfractures tend to develop with stress. These allow passage of synovial fluid into deeper cortical regions and cysts are therefore formed.

Clinical Presentation

Information as to diagnosis, joints affected and possible aetiology can be gained from the moment the patient walks into the consulting room. A careful history must be taken with the following points borne in mind.

The prominent presenting symptom is pain. Typically early in the disease this is brought on by movement and relieved by rest. Later the pain becomes more persistent and avoidance of movement and loss of function of the joint results in muscle wasting. This decreases the stability of the joint and further use will lead to deformity.

Nocturnal pain is not a problem unless the hip, knee or cervical spine are involved. Stiffness on rising in the morning and in getting up out of a chair should only last a few minutes, and the patient declares that this can be fairly easily 'worked off'.

Careful enquiries should be made as to the patient's occupation; prolonged standing at work can aggravate pain in weight-bearing joints, fine manual movements may aggravate OA of the hands, and heavy vehicle driving can cause deterioration in OA of shoulders and cervical spine. Similarly past and present sporting activities should be assessed. Past medical history should be taken to exclude diabetes and acromegaly as treatable causes of secondary OA. Any past conditions associated with hypercalcaemia (e.g. sarcoidosis, milk alkali syndrome) should suggest chondrocalcinosis and past trauma should be noted.

As for family history, the presence of Heberden's nodes in female relatives should be discovered (these are a genetic marker for primary generalised osteoarthritis), as should any history of hip problems in the family. The patient may also be aware of abnormal joint laxity in the family giving rise to the phenomenon of 'double jointedness'.

Social enquiries can assess how the loss of function of the joint affects the patient's life. Upper limb OA usually does not interfere with lifestyle, unless occupationally, but OA of weight-bearing joints can lead to serious loss of mobility and daily activities such as ease of getting in and out of a bath, tying shoelaces, climbing stairs and getting on and off buses should be established.

Examination

The traditional principles of observation, palpation, percussion and even auscultation apply as well to examination of joints as to any other system.

Observation of gait can be assessed initially, a limp is obvious with early OA knee, but only becomes apparent with longstanding OA hip. The joint itself will be swollen and there will be associated muscle atrophy. Palpation will reveal that the swelling is due to bony enlargement and perhaps a small effusion. The boggy sensation typical of rheumatoid synovitis is absent. The joint will neither be hot nor reddened.

The movements of the joint will be limited and measurement of degree of flexion and extension should be taken. If this is not possible, overall loss of joint function can be assessed as a percentage of normal range. Crepitus of a joint can be audible at the knee.

Distribution

The Hand

Heberden's nodes (named after William Heberden 1710-1801, physician to King George III) affect the distal interphalangeal joints (see Fig. 1.1). Initially the pea sized swellings are cystic, but then become bony. Bouchard's nodes (named after Charles Jacques Bouchard 1837-1915, Professor of Medicine and General Pathology in Paris) affect the proximal interphalangeal joints, but neither the metacarpophalangeal joints nor the wrist joints are involved (as distinct from RA). The thumb trapezometacarpal joint is often involved and leads to difficulty using the thumb and the appearance of a square hand. A semi-objective measurement of hand function is grip strength.

Hip Joints

This is usually affected by OA secondary to a congenital abnormality. Hip pain is usually in the groin and is often referred to the medial aspect of the knee. It can particularly disturb the patient at night. The affected hip is held slightly flexed and adducted. The latter gives rise to apparent shortening. If flattening of the femoral head has occurred, shortening may be present also. There is usually wasting of the glutei.

Movements are limited in flexion, abduction and especially in early OA in internal rotation. Crepitus can be heard (using a stethoscope) as