RHEUMATOLOGY IN PRIMARY CARE

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CHAPTER - 11

ARTHRITIS IN CHILDREN

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Musculoskeletal pain affects about 15% children. Various conditions - local and systemic, acute and chronic, benign and malignant - are associated with musculoskeletal pain (Table 11.1) and a correct diagnosis is essential for appropriate management. It is important to remember that all that pains is not arthritis and all arthritides are not painful.

Table 11.1 Causes of Arthropathy in Children (ARTHRITIDES)

Avascular necrosis (femur, scaphoid etc) and other orthopedic conditions e.g. SCFE
Reactive - secondary to infections, notably those of GI tract
Trauma
Hematologic diseases - hemophilia, leukemia, sickle cell disease
Rickets and other metabolic diseases (e.g. scurvy)
Infections - bacterial and tuberculous
Tumors - benign and malignant
Idiopathic - JIA
Drugs - e.g. pyrizinamide
Ehler Danlos and other hypermobility syndromes
Systemic connective tissue disorders e.g. systemic lupus, vasculitis

CLINICAL MUSCULOSKELETAL EXAMINATION

A detailed history and systematic clinical examination are essential as in any other rheumatology case. Examination of children may be difficult in clinical practice due to various reasons. A simplified validated method of musculoskeletal examination for school-age children (Pediatric Gait, Arms, Legs and Spine pGALS screen) takes barely two minutes to perform and can be easily employed in routine clinical practice.

The components of pGALS screen are as follows:

1. Start with 3 screening questions to enquire about
   a) pain and stiffness in joints or back
   b) difficulty in dressing oneself and
   c) difficulty in going up and down stairs.
2. Gait: Observe the child walking, ask the child to walk on tip-toes and heels.
3. Arms: Moving hands in different directions, making a fist, touching fingertips with thumb, squeezing metatarsophalangeal joints.
4. Legs: Bending and straightening knees, passive flexion and extension of hip, feel for knee effusion.
5. Spine: Lateral flexion of cervical spine, bending forwards to touch toes, observe spine from side and behind.

Approach to a child with arthritis should answer following questions:

1. Is the involvement articular on non-articular ? Articular disorders are characterized by pain, swelling, joint line tenderness and limitation of active as well as passive movements. Pain only during active movement and local tenderness are features of non-articular disorders.
2. Is the involvement inflammatory or non-inflammatory ? Pain during physical activity and improvement with rest imply non-inflammatory or mechanical pain. Inflammatory pain is associated with morning stiffness, gelling (pain after a period of inactivity), diffuse joint swelling and tenderness. Fever, fatigue, anorexia, weight loss and other systemic features also indicate inflammatory disease. ESR and CRP are raised in inflammatory diseases. Examination of synovial fluid indicates inflammation.
3. Is the involvement acute or chronic ? A joint swells within hours after trauma or a bleeding diathesis. Acute arthritis (days to weeks) is a feature of infections, rheumatic fever, neoplasia, connective tissue diseases and mechanical joint pains. Chronic arthritis (> 6 weeks) is a feature of Juvenile Idiopathic Arthritis (JIA) and chronic infections such as tuberculosis.
4. How many joints are affected ? Classification of JIA is based on number of joints involved (mono - single joint, oligo - 4 or less joints, poly - 5 or more joints). Acute monoarthritis is seen in infective arthritis, reactive

arthritis, JIA, neoplastic diseases, trauma and mechanical disorders. Chronic monoarthritis may be tuberculous, septic or reactive in a sick child. Acute polyarthritis with fever is a feature of viral arthritis, rheumatic fever, infectious endocarditis and other conditions. Chronic polyarthritis is a feature of various forms of JIA, mechanical problems and metabolic disorders.

5. Which joints are involved? Axial involvement (spine, hips, shoulders, etc) is observed in juvenile ankylosing spondylitis. These joints are rarely affected in systemic diseases such as systemic lupus erythematosus (SLE). Distal interphalangeal joint involvement is typical of psoriatic arthritis.
6. What is the sequence of joint involvement ? An additive involvement implies reactive arthritis; migratory involvement is seen in acute rheumatic fever and intermittent arthritis in SLE or sickle cell disease.
7. Is the involvement symmetric or asymmetric? Inflammatory connective tissue diseases such as SLE, polyarticular JIA and viral arthritis have symmetric joint involvement. Joint involvement is asymmetric in oligoarticular JIA and septic arthritis.
8. Are there any extra-articular features? Various features related to eyes, mouth (Fig. 11.1), skin (Fig. 11.2), hair, nails, genitalia, as well as many systemic features (diarrhea, pleural effusion, renal failure, seizures, etc) may be observed in a child with arthritis. One must look for them carefully. A detailed description of these features is beyond the scope of this chapter.

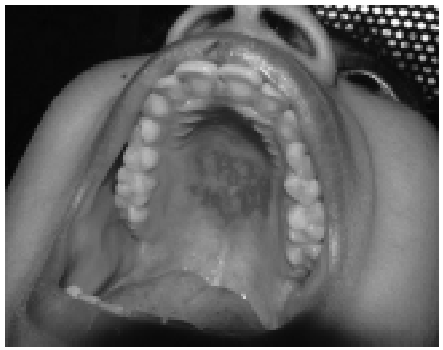

Fig. 11.1. SLE mucositis on hard palate.

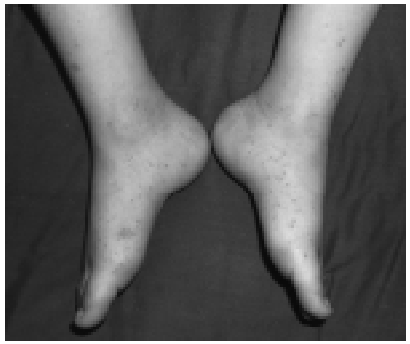

Fig. 11.2 Rash in Henoch Scholein purpura.

NONARTICULAR MUSCULOSKELETAL PAIN IN CHILDHOOD

Mechanical joint pain is common in children due to trauma, sports injuries (tendonitis, sprains), and overuse. Pain in these conditions appears after activity and is relieved by rest. Fractures are also common and must be ruled out in cases of acute pain. Some peculiar conditions affecting children are described below:

1. Growing pains: While well recognized and defined by Peterson's criteria, the precise cause of growing pains is unknown and probably multifactorial. The child (3-8 years) usually complains of evening and night pains which are poorly localized and often relieved by gentle massage. The child always wakes up well the next morning. Clinical examination and laboratory tests are normal. Reassurance is essential and paracetamol may be used for control of pain. Pain will subside as the child grows older.
2. Osgood-Shlatter disease: This is seen in athletically active adolescent males as inflammation of the quadriceps tendon at its attachment to anterior tibial tubercle (Fig. 9.3). There is point tenderness over bilateral tibial tubercles. Knee joint examination is normal. Rest, cold packs and anti-inflammatory drugs are helpful.
3. Legg-Calve-Perthes (LCP) disease: LCP disease (more common in boys aged 4-6 years) is due to avascular necrosis of the femoral head. It starts with limping and pain develops later in course of disease. X-Ray shows flattening of femoral head. MRI may identify the disorder in early stages of the disease before routine X-rays. Medical management with bisphosphonates has been described. Orthopedic management consists of traction and casting followed by physiotherapy.
4. Slipped capital femoral epiphysis (SCFE): Growing end of femur slips off from the shaft due to an injury to the growth plate. SCFE is more common in overweight boys of 10-15 years age and may be associated with hypothyroidism. The onset may be sudden or gradual with progressive pain and stiffness of the involved hip. SCFE is evident on X-Ray of hip (frogs-leg view). Management is surgical.
5. Back pain: Acute back pain is often a sinister complaint in children and signifies infection or malignancy. Chronic back pain is common in children due to mechanical causes such as wrong posture, heavy school bags and sports injuries. Scoliosis (lateral curvature - one shoulder appears higher than the other), kyphosis (forward bending),

spondylolisthesis (slippage of one vertebra over another) are common causes of back pain in children. Stress fracture of pars interarticularis (part between superior and inferior articular processes) or spondylolysis is common in sports that require backward bending and rotation of spine. Scheuermann's ('Shoy-eh-ah-man') disease is due abnormal growth of vertebra (usually lower thoracic) leading to anterior wedging and kyphosis.

6. Nursemaid's elbow: A child of 2-3 year age holds an arm bent and refuses to use it. The injury is due to pulling of extended arm leading to subluxation of radial head. Reduction of radial head resolves the problem. Repeated subluxations seen in joint laxity syndromes may lead to permanent damage.

7. Little leaguer's shoulder: This is a common condition in throwing athletes between 11 and 16 years of age. Inflammation of growth plate leads to pain, tenderness and sometimes weakness in the affected shoulder. The condition is managed with rest and physiotherapy. Rotator cuff injuries can also occur in children.

8. Juvenile fibromyalgia and chronic pain syndromes: Generalized pain of gradual onset is often seen in adolescent girls. There may be an initial insult such as infection but usually there is no obvious trigger. Fatigue, poor sleep and low mood usually accompany pain. This is a disease of central sensitization with multiple tender points. Irritable bowel, poor memory, tension headache and restless legs may accompany the condition. The pain may increase with anxiety, stress, overwork and weather changes. A multidisciplinary approach with family support is essential for management of this condition.

9. Juvenile hypermobility: Ligament laxity (Fig. 11.3) can lead to diffuse pain as well as localized pain such as anterior knee pain and back pain. Recurrent sprains and joint dislocations are also common in these children. A cause-effect relationship may not always be established between hypermobility and pain. Early recognition, physiotherapy, occupational guidance and psychological support are components of successful management of this condition. Hypermobility can be a part of a rare and more serious condition such as Marfan's syndrome, Ehlers-Danlos syndrome and osteogenesis imperfecta.

Fig. 11.3. Hypermobile elbow joint.

JUVENILE IDIOPATHIC ARTHRITIS (JIA)

JIA is an umbrella term that refers to a group of disorders with chronic (> 6 weeks) inflammatory arthritis in children less than 16 years old. It is the most common rheumatological disease of childhood leading to significant disability. JIA is presently classified on the basis of onset, progression, number of joints involved as well as associated clinical and laboratory features. Morning stiffness and joint swelling are predominant features. Pain may or may not be severe. A child with JIA will rarely look sick or have fever except in the systemic onset variety. Identification of the correct subtype is essential for appropriate management and prognostic information. The diagnosis of JIA is essentially clinical. A careful clinical examination is, therefore, mandatory. Laboratory tests help to rule out other conditions rather than prove JIA. Tests such as anti nuclear antibodies (ANA) and rheumatoid factor (RF) merely help to stratify or subclassify the disease. Leukemia, SLE and other inflammatory connective tissue diseases, reactive arthritis, septic arthritis, tuberculous arthritis, HIV disease, fractures and bone tumors need to be considered in differential diagnosis of JIA. A short description of subtypes of JIA follows:

1. Oligoarticular JIA: Almost half of the cases of JIA are oligoarticular (4 or less joints during initial 6 months of disease) - knees (Fig. 11.4), ankles and wrists are most commonly involved. Hips are almost never affected. Local growth disturbances are common sequelae. This form is more common in preschool girls. Laboratory investigations are normal except frequent positivity of anti-nuclear antibodies. More joints

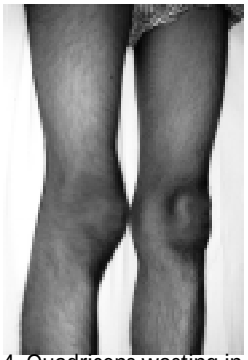

Fig. 11.4. Quadriceps wasting in chronic knee arthritis

can get involved beyond the first six months in about one third cases (extended oligoarthritis). Asymptomatic anterior uveitis is a common association which may lead to visual loss ('painless blind eye'). A regular ophthalmic checkup (3 monthly in ANA positive cases) with slit lamp examination is essential in all these cases.

2. Polyarticular JIA: This subtype begins with symmetric arthritis of 5 or more joints in both upper and lower limbs. RF positive children are usually girls in late childhood and may have rapidly progressive and erosive disease with rheumatoid nodules. The disease is like adult rheumatoid arthritis. Eye involvement is uncommon. Rheumatoid factor negative variant of polyarticular JIA affects younger children. RF negative disease may involve large joints in asymmetric pattern. Active disease may persist for most childhood and remain so as the child goes into adulthood. Eye involvement and growth retardation is possible but rare.
3. Systemic onset JIA (SOJIA): SOJIA is an auto inflammatory syndrome with prominent systemic features such as fever and rash. Daily spiking (quotidian) fever disappears at least once each day. Fever is associated with salmon colored, evanescent, macular rash on trunk and extremities. Lymphadenopathy, organomegaly and serositis are other features. Child looks normal when afebrile. Anemia, leucocytosis, thrombocytosis and mild elevation of liver enzymes are common associations. Raised ESR, CRP and ferritin levels are always found in SOJIA. Macrophage activation and amyloidosis are some of the serious complications of SOJIA that can be life-threatening.
4. Psoriatic arthritis (PsA): PsA is characterized by skin psoriasis, arthritis of distal interphalangeal joints, dactylitis (swelling of entire finger) and nail pitting. History of psoriasis in first degree relative is sufficient to confirm PsA in absence of skin lesions. Arthritis may precede skin lesions by many years.
5. Enthesitis-related arthritis:Most common forms of enthesitis (Chapter 6) are plantar fasciitis, Achilles tendonitis and peripatella pain. This form is more common in boys older than 6 years and presents with arthritis of joints of lower limbs and enthesitis. Spinal involvement is characterized by pain and stiffness. HLA B27 antigen is positive in half of the cases and may be a precursor of adult ankylosing spondylitis. Eye involvement can occur in these patients but is usually symptomatic.
6. Undifferentiated arthritis: This includes children whose arthritis does not fit into any of the above types or those with arthritis fitting into more than one type.

Management of JIA aims at preserving joint function and improving quality of life. Early diagnosis and referral, aggressive pediatric rheumatology management, parent education and physical therapy are integral parts of dealing with these children. Initial treatment includes nonsteroidal anti-inflammatory drugs (e.g. Naproxen) with or without simple analgesics such as paracetamol. Methotrexate (10 mg/sqm per week) and sulphasalazine (30-50 mg/kg per day) are disease modifying drugs and should be introduced early in the course of disease. Glucocorticoids used as topical (eye), intra-articular, low-dose oral and intravenous rescue dose play a crucial role in management. Newer biologic agents are expensive but offer good control in those nonresponsive to first line drugs. Non-compliance to drugs and school absenteeism are peculiar problems in children. Monitoring for disease activity and drug effects are essential. Regular exercise and healthy lifestyle are equally important.

ACUTE RHEUMATIC FEVER

Acute rheumatic fever (ARF) is an immunologically mediated disease subsequent to upper respiratory infection with group-A beta haemolytic streptococci. The prevalence of ARF in India has declined significantly over last 30 years. It is often overdiagnosed without careful application of modified Jones criteria. Carditis (Echocardiography), migratory polyarthritis (inflammatory - lasts for 4 weeks or so), chorea (often a late manifestation), erythema marginatum, and subcutaneous nodules are major criteria for diagnosis of ARF. Minor criteria include fever, arthralgia, elevated ESR/CRP and PR prolongation on ECG. Evidence of preceding streptococcal infection is essential for diagnosis. Throat culture is positive in about 11% cases. A four-fold rise in ASO titre (320 Todd units) in association with other features of ARF may be considered useful for diagnosis. Treatment includes rest, anti-inflammatory drugs (aspirin, glucocorticoids), penicillin (or other antibiotic if sensitive) and other symptomatic therapy. Long term penicillin for prevention of recurrent streptococcal infection is indicated only in correctly diagnosed cases of ARF.

Post streptococcal reactive arthritis (PSRA) does not fulfill criteria for ARF. The arthritis is additive rather than migratory and does not respond well to salicylates. It is recommended that penicillin prophylaxis be given for one year and discontinued if there is no evidence of cardiac involvement.