Spondyloarthropathies

Dee Chapter 74
Apley
Mark Miller
OKU8
JAAOS

Topics

AS
DISH
Other spondyloarthropathies

Ankylosing spondylitis

Prototype disease of seronegative spondyloarthropathies – family of arthritic conditions not associated with positive rheumatoid factor serology

Other similar conditions include reactive arthritis (Reiter's syndrome), spondylitic forms of psoriatic arthritis and IBD

Epidemiology

0.2% prevalence in Western populations
much less common in Asians and Africans
age of onset 15-25 yrs
M>>F - 5-10:1
Familial aggregation
Association with HLAB27
- General population 4-8%
- AS patients 90-95%, 1st degree relatives 50%
- Japanese population 1%

Cause

Immunological disorder
Cause unknown

Pathogenesis and pathology

Areas of inflammation

1. Synovitis of diarthrodial joints
   o Sacroiliac joints
   o Vertebral facet joints
   o Costovertebral joints
   o Peripheral joints (30%) – hips, shoulders

2. Inflammation at fibro-osseus junctions of syndesmotic joints and tendons
   o Intervertebral discs
   o SI ligaments
   o Symphysis pubis
   o Manubrium sterni
   o Bony insertions of large tendons

3. Extraskeletal manifestations
   o Acute anterior uveitis 25%
Valvular disease, carditis, pulmonary fibrosis are rare and late

**Pathological course**
1. inflammatory reaction with round cell infiltration, granulation tissue and bony erosion
2. replacement of granulation tissue with fibrosis
3. ossification of fibrous tissue with resultant joint ankylosis
4. spinal deformity due to limitation of motion at ankylosed levels and increased motion at non-ankylosed levels

**Clinical**

**Clinical presentation and natural history**
Starts insidiously in teenage/young adult years as backache and stiffness
Worse in the morning and after inactivity
Gradual ↑ to continuous pain
10% start as an asymmetrical large peripheral joint arthritis
**family history**
spine disease usually more severe if patient also has hip pathology
Progressive loss of posture and mobility
Clinical problems – fractures, kyphotic deformity, AA instability, large joint disease

**Physical**
Initially diffuse pain and tenderness over back, SIJ or occasionally single large joint
Then:-
Loss of lumbar lordosis, Increased thoracic kyphosis, Forward thrust of neck, Chin-on-chest deformity
Upright posture maintained by hips and knees slightly flexed (later may be fixed deformities)
↓ spinal ROM in all directions, extension affected the earliest and most severe; “wall test”
Decreased lengthening on forward flexion – Shober test
Decreased chest expansion (normal 7cm)

**Xrays**
Sacroiliac joints
- first skeletal change that occurs in AS
- early cardinal sign is erosion of SIJ with patchy osteopenia, usually on sacral side
- then superficial erosions + subchondral sclerosis
- finally bony ankylosis
- better evaluated with CT

Spine
- order of skeletal changes is
  - Thoracolumbar & lumbosacral → midlumbar, thoracic, cervical
- earliest change is **squaring of vertebra** (loss of anterior concavity of ant VB)
- relative sclerosis at corners and relative osteopenic midsection
- then **bridging marginal syndesmophytes** between adjacent vertebrae *(bamboo spine)*
  - ossification of annulus
  - vertically oriented marginal syndesmophytes classical for seroneg spondyloarthropathies
- kyphosis
- **canal stenosis** – better evaluated with MRI
- **Cspine**
  - AA subluxation
  - Occipitoatlantal destruction and painful deformity
  - Subaxial # deformity & instability
  - Fixed cx kyphotic deformity
- **Peripheral joints**
  - erosive arthritis and progressive bony ankylosis

**Bone scan**
Presence of activity in SI joints and spinal column may help with diagnosis
Good screening tool to identify occult fractures

**DEXA Scan**
- Osteoporosis – better to measure this with DEXA hip measurements or quantitative CT; DEXA spine measurements may be falsely elevated

**Labs**
- ESR high in active disease
- HLAB27 90-95% but not good screening test as normal population 4-8%

**Tri:**
- Mechanical LBP, spondylolisthesis – type of pain different
- DISH – usually older men, ESR normal, SIJs normal
- JRA (not associated with HLAB27)
- Seronegative spondyloarthropathies
  - Reiters, Psoriatic, GI, Whipples, Behcet may be indistinguishable from AS

**Treatment**

**Principles**

1. **General measures**
   - physiotherapy, stretching, exercise
   - avoidance of immobilisation
   - avoid pillows which would worsen neck kyphosis
   - avoid contact sports

2. **NSAIDS + Calcium Vit D**

3. **Treatment of complications**
   a. Spine fractures  ORIF
   b. Hyperkyphosis  kiv op in severe
   c. Spinal cord compression  from AA subluxation or OPLL – treat accordingly
   d. L/S N root compression  tx as reqd

**Operative treatment in AS**

Degenerative arthritis and ankylosis of hip

- **THR - See AR section**
Fractures

- **Difficulty in management**
- **Marked osteoporosis**
  - Long ankylosed lever arms cause motion and stress concentration at # site
  - Even undisplaced # have been seen to displace completely
  - Conservatively tx # may get late *epidural hematoma* due to continuing micromotion at # site

**Neurologic compromise common**

- **Cervical spine fractures**
  - High morb and mort both with surgical and nonsurgical tx
  - Halo vest may be used, reasonably high union rates by 12-18 weeks
  - Partial and progressive neurological deficits warrant urgent surgery
  - Surgery
    - Laminectomy for *epidural hematoma* (can develop weeks after injury)
    - Fixation difficult due bone quality, need to consider long fusions
    - Need to consider postop halo vest

- **Thoracic and lumbar fractures**
  - Consider rigid internal fixation

**AA-instability**

Causes are multifactorial
  - Still lower cspine ↑ stress at the AA level
  - Inflammation at AA region leading to loss of transverse and alar ligament integrity

May have AA subluxation with Cord compression
May also autostabilise

**Management**

- Pre-any GA Cspine film need to be taken to document AA stability
- If **unstable + cord compression** → stabilisation

**Cervical spine kyphotic deformity**

- **Clinical problems**
  - Field of vision restricted
  - Skin care problems
  - Even opening mouth may be obstructed (chin-on-chest deformity)

- **Chin-brow angle**
  - Angle formed by line from brow to chin to vertical is measured with hips and knees extended – neck is either in fixed or neutral position
  - Most reliable measure of degree of trunk deformity
• **Acute onset of cervical flexion deformity**
  Presumed to have fracture until proven otherwise
  Usually shear #s at the base of the neck
  Gradual ↑ in deformity during the day, return to normal position in the morning after the night
  CT + recon required to demonstrate C spine clearly
  Treatment of choice
  • Halo vest with slow correction and healing # over 12 months
  • Care not to hastily overcorrect deformity and result in neurologic compromise

• **Fixed severe cervical painless deformity**
  May be candidates for resection-extension osteotomy
  • Age
  • Condition
  • Feasibility of surgery
  • Other risk factors
  Usually performed at C7T1 level
  • Canal relatively wide
  • C8 root quite flexible and also the least important UL root
  • Vertebral A/V usually pass anterior to the TP, not within

  Rough operative procedure
  • Done under LA
  • Halo vest applied first
  • Dissection down under LA, wedge cut according to chinbrow angle
  • Short acting GA, spine broken and wedge closed (closed osteoclasis)
  • Patient awoken and asked to move all 4 limbs
  • The resected bone applied to facilitate fusion

  Thoracic spine kyphotic deformity
  If severe may be treated with lumbar osteotomy **at or below L2**
  Second most common level for corrective osteotomy
  Must determine level of conus to prevent neurologic injury

**Surgical options**
  • Classic osteotomy described by Smith-Peterson
    • Post element removal with **opening wedge of anterior column**
    • problems
      o Superior mesenteric artery syndrome
      o Aortic disruption or vascular injury 1%
      o Neurologic injury
    • Mortality 10%
    • Can correct up to 75°

  • New technique of pedicle subtraction osteotomy
    • Posterior closing wedges reduce length of posterior column
      o Take off posterior elements and wedge of post aspect of VB
      o Can correct up to 45°
      o Centered over L3 with PSIF 2-3 levels above and below
      o Advantage of more stability, **not opening anterior column** and bone to bone healing
    • Fixation with Luque, sublaminar wires and postop cast/bracing x 1 year
    • Reduced risk of vascular injury, superior mesenteric artery syndrome and paraplegia
DISH

Basics

Diffuse idiopathic skeletal hyperostosis
Forrestier’s disease
Originally mixed up with AS but now definitions are distinct
Etiology unknown
- No association with HLAB27
- May be associated with HLA B8
- DM, hyperuricaemia, dyslipidaemia + 15-25% of all men and women >50 years
- no racial differences

Clinical
Most are asymptomatic and discovered incidentally
Usually presenting with chronic mild mid/low back pain, spinal stiffness

May get extraskeletal manifestations
Also have symptoms from hyperostosis at tendons and ligaments – esp. Achilles tendon
Occasional dysphagia from cervical osteophytes
ESR, CRP, RA, ANA HLAB27 normal

Spinal manifestations

Thoracic and lumbar spine
Most frequent area of occurrence, especially lower T vertebrae
Lumbar occurs, slightly less often than T
- May be associated with normal DDD of L4-S1
- Combination with DISH may result in significant spinal stenosis that may warrant operation
Screen with CXR; if suspecting DISH should then do T spine AP/lat

Syndesmophytes
- Nonmarginal horizontally oriented syndesmophytes causing extraarticular ankylosis – ossification of ALL originating in Sharpey’s fibres at point of attachment of ligament to waist of each vertebra
- Diagnostic feature is non-marginal bridging osteophytes at 4 contiguous levels
- More prominent on R side, (May be because on L side aorta prevents growth – in situ inversus, there are more prominent syndesmophytes on L side!)

3 diagnostic criteria required

1. Flowing ossification anterolateral aspect of at least 4 contiguous vertebrae
2. No DDD
   a. no significant DDD changes
   b. preserved disc height
   c. no marginal sclerosis
   d. no vacuum phenomenon
3. No other ossifying problem
   a. no facet joint ankylosis
   b. no SI erosion, sclerosis or intraarticular osseus fusion
Usually no osteoporosis like in AS, BMD may in fact be high for age
Also no flattening of anterior aspect of vertebral body as seen in AS
Facets and disc spaces are preserved

Bone scan may be hot in involved areas making the △△ of mets a possibility

**Cervical spine**
Not as common as T and L spine
Anterior syndesmophytes may cause dysphagia and warrant possible excision
Multiple level cervical stenosis
- with normal lordosis: laminectomy or laminaplasty is possible
- kyphosis must go anterior decompression + stabilisation

**Spine trauma**
High risk of fracture and instability even from minor trauma
DISH with pain → # must be aggressively sought for and excluded
Risk of neurologic compromise
Treat with ORIF
- consider fusing longer (may not lose that much motion cause ROM may have already been ↓ preop anyway),
- or ant/post procedures

**Extraspinal manifestations**

<table>
<thead>
<tr>
<th>Shoulder</th>
<th>Elbow</th>
<th>Wrist and hand</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptomatic bony hyperostosis with ↓ ROM or pain</td>
<td>Olecranon spurs</td>
<td>DRUJ</td>
</tr>
<tr>
<td>• deltoid tubercle</td>
<td>Distal medial humeral hyperostosis</td>
<td>Interosseus membrane</td>
</tr>
<tr>
<td>• greater/lesser tubs</td>
<td>Irregular radioulnar joint</td>
<td>Periarticular hyperostosis in hands</td>
</tr>
<tr>
<td>• inferior glenoid</td>
<td></td>
<td>Arrowheading of P1 tufts</td>
</tr>
<tr>
<td>• clavicle</td>
<td></td>
<td>Possible ↑ finger 1° OA</td>
</tr>
<tr>
<td>• attachment of CC ligaments</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Again, no ↑ shoulder 1° OA</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Pelvis</th>
<th>Hip</th>
<th>Knee</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enthesophytes involving, iliac wing, ischiatic tuberosity</td>
<td>Variable</td>
<td>Prominent tibial spine</td>
</tr>
<tr>
<td>Ossification of sacrotuberous and iliolumbar ligaments</td>
<td>If present may have findings that</td>
<td>Ossification of quadriceps</td>
</tr>
<tr>
<td>Periarticular (not intraarticular) osteophytes hip, SIJ &amp; symphysis pubis</td>
<td>may or may not be related to DISH</td>
<td>Patellar hyperostosis</td>
</tr>
<tr>
<td></td>
<td>• periarticular bone proliferation with intact joint space</td>
<td>Don’t seem to have an ↑ in knee OA</td>
</tr>
<tr>
<td></td>
<td>• hyperostosis with narrow jt space</td>
<td></td>
</tr>
<tr>
<td></td>
<td>• frank ON</td>
<td></td>
</tr>
<tr>
<td></td>
<td>May need THR - ↑ risk of HO</td>
<td></td>
</tr>
</tbody>
</table>

| Foot & ankle                |                                 |                                  |
|-----------------------------|---------------------------------|                                  |
| 70% have F&A manifestations spur formation from any bone, or ligament/tendon attachments | |                                  |
| • calcaneal spurs 75%       |                                 |                                  |
| • TA or plantar fascia calcification |                                 |                                  |
• Heel pain in 25%
• Enthesophytes of other foot bones

Treatment of DISH
Exclude other causes
Symptomatic treatment after diagnosis
Aggressive investigation to rule out # in acute pain/neuro deficit
ORIF of spine KIV front and back
Lumbar spinal decompression if required

THR in DISH patients
Definitely at increased risk of HO (40-50% vs 10-20% in some studies)
HO may not really cause significant ↓ROM or disability to warrant prophylaxis
Studies on prophylaxis
• Preoperative irradiation
• Anti-vitamin K drugs (e.g. warfarin) significantly ↓ HO
• Aspirin, subcut heparin, LWM heparin not investigated
Other spondyloarthropathies

Spondylitis associated with Inflammatory bowel disease

Both Crohn's disease and UC are associated with sacroiliitis and spondylitis – similar clinically and radiographically to AS – occur in 15-60% of pt's with IBD
Not affected by colectomy in these patients

Spondylitis related to Psoriatic arthropathy

Common seronegative spondyloarthropathy
In psoriatic arthritis, spinal involvement is seen in up to 70% of patients
  - Asymmetric involvement of small joints of hands and feet
  - Psoriatic tend to have bigger syndesmophytes
  - Psoriatic tends also to present more with RA type spine involvement
    - Bony erosions
    - Subluxations
    - AAS
    - Pannus of odontoid
  - Neurologic deficits
  - Treatment is like that of RA

Reiter’s syndrome

  - Peripheral polyarthritis (large and small joints of lower limbs) with urethritis and conjunctivitis
  - Post-infectious reactive arthritis
  - 3% spinal involvement – less severe and extensive than AS
  - Xray changes – juxtaarticular osteoporosis, joint space narrowing, erosive changes
  - Late complications – Cardiac complications, AV block, pulmonary fibrosis

Neuropathic spondyloarthropathy

Pathogenesis
Occurs secondary to alteration of pain sensation in affected joint
Absence of protective sensory motor reflexes causes excessive loads and range of motion – joint destruction

Associations
Tabes dorsalis
  - Affects primarily dorsal columns resulting in loss of proprioception
  - Mainly involving lumbar spine
Syringomyelia – mainly cervical spine
DM
SCI

Pathology / Xray changes
Progressive spinal deformity and joint destruction
  - Advanced spondylosis
  - Osteophytes
  - Facet hypertrophy
  - Vertebral body sclerosis
Possibly leading to cord impingement
Differential diagnosis
Severe OA, DISH, infection, tumour, Paget’s disease

Treatment
Spinal stabilisation possibly with bracing initially
- Intractable pain and progressive weakness/neurology are indications for surgery
- Usually combined anterior and posterior spinal fusion