

A 'benign' condition masquerading as arthritis

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Case History

9 year old boy presented with a 3 week history of:

- Swelling of major lower limb joints
- Progression was additive (right ankle followed by left ankle and then left knee)
- No preceding fever/ trauma
- No GI or GU symptoms/ contact with tuberculosis

General Examination

- Afebrile, well looking child
- Significant node left axilla, central group
- No BCG scar



➤ **Anthropometry**

Height: 127 cm (between 25th and 50th centile)

Weight: 21 kg (on the 10th centile)

Arm span: 129cm

Arm span : Height = 1.01

➤ Systemic examination (CVS/RS/Abdomen) - normal

Musculoskeletal examination

- Swelling of both ankles and left knee
- Positive patellar tap s/o knee effusion (left side)
- No other signs of inflammation i.e. warmth/
tenderness
- Significant generalised hypermobility noted

Differential diagnosis at admission

Inflammatory arthritis probably

- Oligo articular JRA
- Reactive arthritis
- TB synovitis left knee

Investigations

- Counts: Normal
- Inflammatory markers (ESR, CRP): Normal
- ASO, ANA, RA factor, anti CCP: Negative
- Urine and stool culture: Sterile
- ECG & ECHO: Normal
- Mantoux : 20x20 mm positive

Distracters

Strongly positive Mantoux

hence further investigations for TB were done

- CXR: Normal
- FNAC of axillary node: Reactive changes
- Aspiration & synovial biopsy of left knee planned

MRI done prior to synovial biopsy revealed a

partial ACL tear > 50%, large joint effusion

MRI left knee



Partial tear involving > 50% of the anteromedial bundle of the ACL just above its tibial insertion with interstitial tear involving the entire stretch of the ACL fibres. However no complete discontinuity is noted and the ligament is parallel to the Blumensatts line

*D R Pai, P J Strouse. MRI of the Pediatric knee.
AJR: 196, May 2011*

Course in hospital

No significant response to a trial of NSAID
(Naproxen)

Review of history and clinical findings done

In view of....

- Absence of constitutional symptoms
- History suggestive of mechanical joint pathology
- Weight bearing joint involvement
- No rise in inflammatory markers
- Hypermobility with a Beighton score of 9/9
- ACL tear in the absence of trauma

G D Myer, K R Ford, M V Paterno, T G Nick et al. The effects of generalised joint laxity on risk of anterior cruciate ligament injury in young female athletes. Am J Sports Med. 2008 June; 36(6): 1073 - 1080

R Ramesh, O Von Arx, T Azzopardi, P J Schranz. The risk of anterior cruciate ligament rupture with generalised joint laxity. J Bone Joint Surg [Br] 2005; 87 - B: 800 - 3

Revised diagnosis

Hypermobile joint syndrome masquerading as arthritis

RM Lewkonia, BM Ansell. Articular hypermobility simulating chronic rheumatic disease. Arch Dis in Child, 1983, 58, 988 - 992

H A Bird, V Wright. Joint hypermobility mimicking pauci - articular juvenile polyarthritis. BMJ August 1978

Discussion

Hypermobile joint syndrome

- Connective tissue disorder, AD
- It is commonly seen in children (prevalence 8 – 30%), Asians, female gender
- Earlier nomenclature - BHJS
- Not considered to be benign anymore

N. Adib, K. Davies, R. Grahame, P. Woo et al. Joint hypermobility syndrome in childhood. A not so benign multisystem disorder? Rheumatology 2005; 44:744 – 750

Pathology

- Abnormality in collagen leading to loss of tensile strength and increased fragility
- Defect in matrix protein **tenascin x** probably due to a gene deletion
- Ratio of type 1 to type 3 collagen is decreased

Types

Syndromic

- Marfan & EDS
- Osteogenesis Imperfecta I, IV
- Down
- Stickler
- Some Skeletal dysplasias

Non syndromic

- Hypermobile joint syndrome

Discussion contd.

- Unclear why some cases are symptomatic while others are asymptomatic
- Only if symptomatic the diagnosis of HMS can be entertained
- Early diagnosis necessary to prevent complications
- Beighton and Brighton scoring systems are used in the diagnosis

Table 4: The 1998 Brighton criteria for a diagnosis of Benign Joint Hypermobility Syndrome[8].

Major Criteria:

- 1. Beighton Score of $\geq 4/9$**
 - 2. Arthralgia for > 3 months in > 4 joints**
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Minor Criteria:

- 1. Beighton score of 1–3**
 - 2. Arthralgia in 1–3 joints**
 - 3. History of joint dislocation**
 - 4. Soft tissue lesions >3**
 - 5. Marfan-like habitus**
 - 6. Skin striae, hyperextensibility or scarring**
 - 7. Eye signs, lid laxity**
 - 8. History of varicose veins, hernia, visceral prolapse**
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For a diagnosis to be made either both of the major criteria must be present or 1 major and 2 minor or 4 minor.

Complications

Acute/ Traumatic

- Meniscal tears
- Joint dislocation/ subluxation
- Traumatic arthritis
- Sprain
- Bruising
- Fractures

Chronic/ Non traumatic

- Soft tissue rheumatism
- Chondromalacia
- Fibromyalgia
- TM joint dysfunction
- Nerve compression disorders
- Flat foot
- Osteoarthritis
- CDH

Management

Physiotherapy – Mainstay

- Splints for joints undergoing excessive movements
- Stretching techniques for muscles
- Maintain mid range of movements during exercises
- Minimizing trauma

Leslie N Russek. Examination and treatment of a patient with hypermobility syndrome. Phys Ther. 2000; 80:386 - 398

At follow up

- No significant response to NSAID therapy
- Developed tenosynovitis of tendons around both ankle joints (documented by MRI)
- Physiotherapy has been initiated
- Long term outcome unclear

Key messages

- HMS is certainly **not a benign condition**
- It can masquerade as arthritis
- Diagnosis is one of exclusion
- Specific treatment is not available
- Physiotherapy is the mainstay of treatment

Take home message

- All children with musculoskeletal problems, including arthritis, should be screened for joint hypermobility
- Early diagnosis is essential - to retard progression to osteoarthritis and to avoid unnecessary therapy