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Coexistence of Ankylosing Spondylosis with Benign Joint Hypermobility Syndrome. Are they Complementary to Each Other?

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Abstract

Objective: **To present cases of co-existence of pathologically opposite conditions ankylosing spondylitis (AS) and benign joint hypermobility syndrome (BJHS).**

Design: **Clinical findings of 25 cases of ankylosing spondylitis with overlapping clinical picture of benign joint hypermobility syndrome were evaluated.**

Results: **There were 23 males and 2 females with the mean age of 25.92 ± 7.54 years (age range 17-41 years). The mean duration of the disease was 6.25 ± 6.72 years. The mean Beighton's score for joint hypermobility syndrome was 6.36 ± 1.32. Schobers test was positive in 13 patients and 23 patients were positive for HLA-B27. ESR was elevated in 24 patients and 68% patients were with grade II sacroilitis. Cardiovascular, ophthalmologic complications were also observed in some patients. Ligament and menisci injuries were observed in 3 patients. It was observed that many of the patients had contracures of major joints along with spinal deformity and simultaneously they had hypermobility in other major and smaller joints.**

Conclusion: **Coexistence of these two opposite disorders causes confusion and thus delay in the diagnosis and management, leading to further disability. Hypermobility can be advantageous for a stiff spine and for major joints. By the care of the joints and spine with proper exercises programme further deformities and soft tissue injuries can be prevented.**

Key Words: **Ankylosing Spondylitis, Benign joint Hypermobility Syndrome, Exercise, Rehabilitation**

Introduction

Ankylosing spondylitis (AS) is an inflammatory disorder of unknown cause, primarily affecting axial skeleton, peripheral joints and extra articular structures. It affects 0.1% to 0.2% of the population, commonly affecting males, 90% of the patients are positive for HLA-B27 but it has not got any diagnostic importance. Common complications observed in AS are restricted range of motion of spine and joints, contractures, ankylosis, kyphosis, osteoporosis, fractures of vertebra and atlanto axial subluxations. Some of the extra articular manifestations are pulmonary fibrosis, aortic regurgitation, endocarditis and iridocyclitis.

Benign joint hypermobility syndrome (BJHS) is a complex of acute recurrent or recalcitrant wide spread soft tissue lesions of traumatic origin. It is one of the most commonly misdiagnosed entities in the musculoskeletal pain syndromes. Kirk et al observed that it has distinct pathology with the presence of rheumatic symptoms with generalized joint laxity in the absence of any demonstrable systemic rheumatic disease.[1] Females and children are most affected and it is most common in Asians (6-8%). Soft tissue injuries, dislocations, easy fatigue, touch pain;

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anxiety, depression, varicose veins and hernias are observed in this condition. Most common extra articular manifestation is mitral valve prolapse (7%). Brighton's diagnostic criterion is used for the diagnosis of BJHS.

We observed coexistence of both the conditions in 25 patients as both the conditions are opposite to each other in manifestations and complications. It becomes important to understand the management and prognosis of such patients. In the present article we report unique features of both the conditions in patients who fulfill the diagnostic criteria for AS and BJHS.

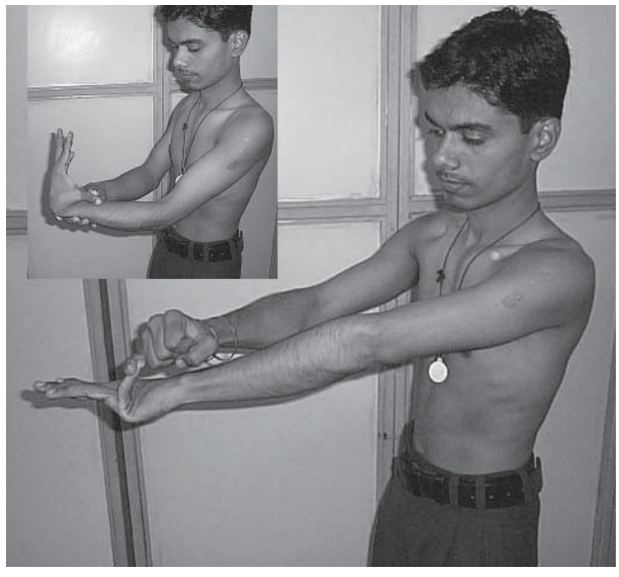

Fig 1: A patient with AS who was treated as tuberculosis of hip joint with hypermoblity.

Fig 2: Patient of AS with hypermobility in elbow joint.

Materials & Methods

25 consecutive patients who fulfilled the diagnostic criteria for both AS and BJHS were studied. These patients were either referred from the rheumatology clinic or had directly attended the out patient department of PMR between April 2001 and March 2004 with various musculoskeletal complaints. Before attending our hospital few patients were already diagnosed cases of ankylosing spondylitis and were on treatment. In these cases some of them were earlier treated as tuberculosis of hip, juvenile rheumatoid arthritis, rickets and gout before the diagnosis of either AS or BJHS. All the patients had features consistent with those of ankylosing spondylitis with backache, stiffness in the spine, joint pain with effusions, contractures and radiological findings of sacroilitis. Hypermobility syndrome was based on Beightons score of > 4/9 (1 point for each of the following: hyperextension of the each fifth finger > 90^{o}, opposition of each thumb to the flexor aspect of the forearm (Fig-1), hyperextension of elbow and knee > 10^{o} (Fig-2) and forward trunk flexion placing hands flat as floor with knees extended; maximum of 9 points).

All patients satisfied the 1984 modified New York criteria for ankylosing spondylitis and 1998 Brighton's criteria for the diagnosis of the joint hypermobility syndrome (Table-1). After the clinical diagnoses the routine investigations for blood (Hemogram, ESR), X-ray of sacroiliac joints, tissue typing for HLA-B27 and echocardiography were done. Ophthalmologic examination was also done in all patients. After confirmation of the diagnoses drugs like NSAIDs, methotrexate and sulphasalazine depending on the

Table-1: Brighton's diagnostic criteria for the diagnosis of benign joint hypermobility syndrome:

Major criteria:

- Beighton's score $\geq$ 4/9
- Arthralgia for > 3 months in more than 4 joints

Minor criteria:

- Beighton's score 1-3
- Arthralgia in 1-3 joints
- History of joint dislocation
- Soft tissue lesions > 3
- Marfanoids habitus
- Skin striae, hyperestensibility or scarring
- Eye signs, lid laxity
- History of varicose veins, hernia, visceral prolapse.

For the diagnosis:

- 2 Major criteria
- 1 Major + 2 Minor criteria
- 4 Minor criteria

Table-2: Suggested management plans for these kinds of patients:

Number	Plan		
1	Proper diagnosis		
2	Drugs: NSAIDs, Methotrexate, Sulphasalazine		
3	Exercises	**Do's**	**Don'ts**
		1. Range of motion exercises in the possible range, not beyond normal range	1. Don't put overstrain on joints and muscles
		2. Stretching exercises in the contractured joints.	2. Not to put weight on joints in a single posture
		3. Isometric strengthening exercises	3. Not to overstretch the injured joints
		4. Deep breathing exercises	
		5. Encourage non strenuous play like swimming and dancing	
4	Prevention of soft tissue injuries, Joint protection measures		
5	ADL advice		
6	Counseling / Reassurance: For sex, muscle pain, anxiety, depression		
7	Education regarding the disease and the prognosis		
8	Surgery: THR, TKR for ankylosed joints if indicated		

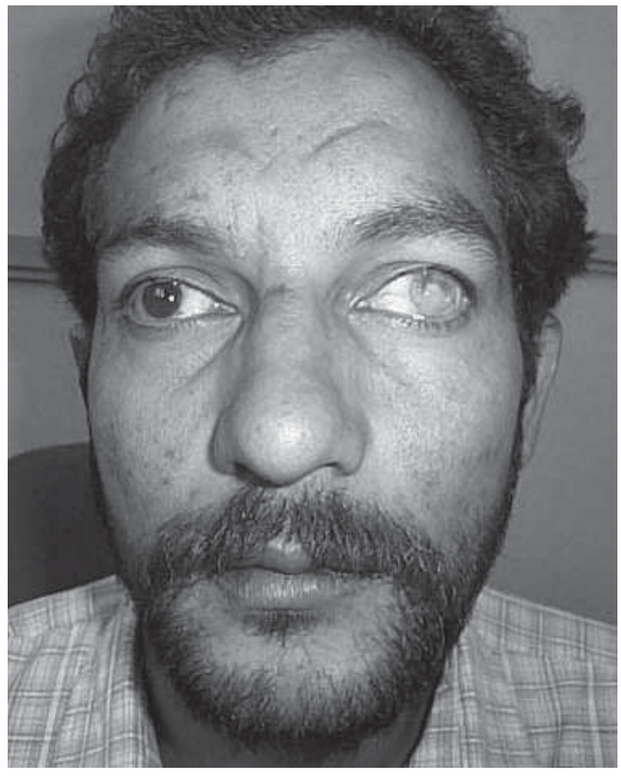

Fig 3: Patient with opaque cornea on left eye.

severity of the disease, exercises for joints and spine according to the deformity or injury (Table-2), activities of daily living modifications and postural care was advised.

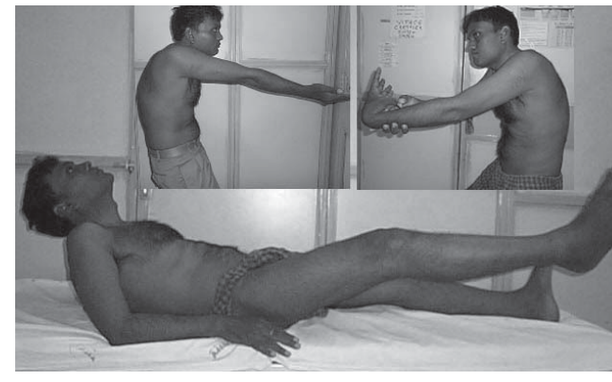

Fig 4: A typical case of ankylosing spondylitis with spinal deformity and fixed flexion deformity of right hip joint, who was not able to sleep in supine position because of these two deformities. Inside photo the same patient with hypermobility in upper limbs.

Results

The patients' demographic profile is given in Table–3. There were 23 males and 2 females with the mean age of 25.92 $\pm$ 7.54 years (age range 17-41 years). The mean duration of the disease was 6.25 $\pm$ 6.72 years. The mean Beighton's score for joint hypermobility syndrome was 6.36 $\pm$ 1.32. Schobers' test was positive in 13 patients and on tissue typing 23 patients were positive for human leukocyte antigen (HLA)-B27. ESR was elevated in 24 patients with the mean of 42.8 $\pm$ 29.34 mm/1st hour. All patients were positive for sacroilitis with 68 % were with

Table -3: Profile of the patients.

Age	Sex	DD yrs	ESR (mm)	HLA B27	Sacroilitis Grade	ST	BS	Associated Problems
17	M	10	43	+ve	II	-ve	8/9	Treated earlier for anterior cruciate ligament injury, medial meniscus injury, bilateral calcaneal spur
41	M	10	75	+ve	II	+ve	7/9	
21	M	0.5	50	+ve	II	+ve	5/9	Treated earlier as Rheumatoid Arthritis
17	M	1.5	22	+ve	III	+ve	8/9	Treated as Koch's hip, plantar fascitis, scoliosis, shortening of limb
20	M	3	40	+ve	II	+ve	5/9	Shortening of Limb
35	F	2	62	+ve	II	-ve	6/9	
16	M	1.5	09	+ve	III	-ve	6/9	Treated for rickets, earlier
20	M	1	145	+ve	II	-ve	6/9	Bilateral Plantar Fascitis. Family h/o AS
22	M	5	27	+ve	II	-ve	5/9	
28	M	3	57	+ve	II	+ve	6/9	
32	M	2	32	+ve	II	-ve	5/9	Known c/o Diabetes Mellitus, Chest expansion reduced
22	M	0.5	17	-ve	I	-ve	8/9	
18	M	3	17	+ve	II	-ve	5/9	Bilateral retrocalcaneal bursitis
28	M	7	50	+ve	II	-ve	8/9	Intervertebral Disc Prolapse
30	M	10	22	+ve	I	+ve	7/9	
32	M	12	55	+ve	IV	+ve	5/9	Brother had AS (died at 26 years of age), kyphosis, right hip fixed flextion deformity
23	M	2	18	+ve	I	-ve	9/9	
35	M	8	80	-ve	II	+ve	6/9	Chest Expansion reduced
20	M	14	30	+ve	I	-ve	7/9	ACL repair, Posterior subluxation of knee, ATT given for Koch's knee
19	M	10	19	+ve	III	+ve	8/9	Reduced Chest Expansion
30	M	0.3	30	+ve	II	-ve	6/9	Gouty Arthritis
19	M	3	60	+ve	II	+ve	6/9	Hallux Valgus
34	M	17	07	+ve	II	+ve	7/9	Left Eye Corneal Opacity, right healed uveitis
29	F	29	48	+ve	II	+ve	5/9	G3, A2, LSCS done due to growth retardation of child. Had ankle injury
40	M	1	55	+ve	II	+ve	4/9	Trivial AR, Type II Diastolic dysfunction, Deformity left shoulder, uveitis right eye

Abbreviations used: DD-duration of disease; ST-Schober's test; BS-Beighton's score

grade-II, 16% with grade I, 12% with grade III and remaining 4% with grade IV. Aortic regurgitation was observed in only one patient with type-II diastolic dysfunction. Two patients had healed uveitis in right eye and one of them had complete corneal opacity on left eye (Fig-3). Four patients had reduced chest expansion. One patient had a family history of ankylosing spondylitis. Two patients had plantar fascitis; bilateral retrocalcaneal bursitis, calcaneal spur and hallux valgus deformity was observed in each patient. One female patient had repeated abortions twice. Two patients had received treatment for tuberculosis of hip and knee joints. Many of the patients were having associated spinal deformity (Kyphosis, scoliosis) contractures in hip, knee and shoulder joints; but the same patients had Hypermobility in other joints (Fig-4). Two patients had anterior cruciate ligament injury

and one of them had associated medial meniscus injury of knee joint; one female patient had ligament injury in ankle joint. No patients had any varicose veins or hernias.

Discussion

When a patient with Hypermobility syndrome develops features of spondyloarthopathy it is difficult to label him in a particular group as either BJHS or AS because features like back pain, fatigue, joint swelling, range of motion of joints and spine may overlap and cause confusion. In this series some patient were neither diagnosed as ankylosing spondylitis nor BJHS but they were previously treated elsewhere as joint tuberculosis, rickets, gout and juvenile RA. The elevated alkaline phosphatase was stated to be the reason to treat one patient for rickets, but Sheehan NJ et al had reported that alkaline phosphatase level is also elevated in patients with ankylosing spondylitis.[2] Ligament and menisci injuries are rare in ankylosing spondylitis but in this series we observed that because of associated hypermobility few patients had sustained soft tissue injuries.

Hypermobility an inherent laxity of ligaments may be present in up to 10% of individuals in western populations and about 4-7% in general population are having lax joints.[3, 4, 5] The range of joint movement varies considerably between individuals and with age and race. People from Indian and African continents do have a greater range of joint mobility than Caucasians of the same age and sex.[6] Hypermobility is advantageous in few professionals like dancers (Bharathanatyam [classical Indian dance] dancers, ballet dancers) and musicians. If a patient with Hypermobility syndrome develops spondylitis it is difficult to define the mobility criterion for either disease especially if the Hypermobility syndrome has not been strictly defined prior to the onset of symptoms of spondylitis.[7] Thus the mobility criteria for both diseases will be at least partly eliminated. In this series nearly half of the patients had reduced spinal movement and four patients had reduced chest expansion.

The problems associated with hypermobility are often thought to diminish with time as the degree of hypermobility wanes with age, whereas problems with spondyloarthropathy progress as the age advances. Existence of these two conditions in the same individual can cause stressful life from early childhood to till old age. It is not known exactly as how does hypermobility help in spondyloarthopathy, especially in preventing contractures and spinal deformity. Both the conditions are associated with cardiovascular complications. In BJHS the development of mitral valve prolapse (MVP) is 7%, whereas in AS aortic root diseases and valve disease are also very common[8], thus leading to high morbidity in such patients. In the current series associated cardiac problems were less, but there are chances of developing in later stages. Many patients with BJHS also report a range of extra-articular symptoms, some of which are related to psychological stresses such as anxiety and depression but others possibly associated with autonomic dysfunction[9]. In addition to pain and stiffness in AS patients fatigue, physical look, sleep disturbances, mood, and social relationship problems are also common.[10] Hypermobility patients do complain of swelling with stiffness in the joints[11], morning foot pain and low back pain without any structural abnormality and which interferes in their vocation. The same symptoms were also observed in AS with backache, joint swellings, early morning stiffness and foot pain due to associated plantar fascitis. A person with coexistence of these conditions had to suffer more pain compared to either AS or BJHS alone.

Bird et al [12] hypothesized that there are three main factors that contribute to the joint hyperlaxity; the inherited collagen structure (though this may sometimes be modified by applied physical factors, particularly in sports), the shape of the bony articular surfaces and the neuromuscular tone that controls joint movement. Sutro C.J[13] expressed his doubt that this laxity might be due to disproportion in the relative rate of growth of the bone and their attached ligaments. He also mentioned the formation of isolated joint laxity as a compensatory mechanism just adjacent to a fixed or stiff joint. Charpel A and Marks R [14] from their study found that there is an increased incidence of joint injury and joint degeneration in BJHS. Tarsal tunnel syndrome, carpal tunnel syndromes, musculoligamentous lesions and early onset osteoarthritis are also common in patients with BJHS.[4, 15, 16] These compartmental syndromes are also observed in ankylosing spondylitis. Mobile flat foot, hindfoot valgus and hypermobile ankles are also observed in BJHS.

In this series we observed that majority of our patients were positive for HLA B-27 and sacroilitis. It is not known if any relation existed between this and the Hypermobility and till now no studies have focused this aspect. When it comes to management aspect the problems are whether drugs like methotrexate and salfasalazine along with NSAIDs should be given or not. Patients do require these drugs in later stages of AS. And last but not the least the exercises, which may lead directly or indirectly to stretching of tendon or joint ligament should be avoided and in this context the isometric strengthening exercises would be the exercise of choice. But in the joints where contractures have already developed, stretching exercises are required. Hypermobility patients are good in sports activities like swimming and dancing which are more helpful for AS patients to maintain range of motion of joints and spine and these activities should be encouraged. Prevention of soft tissue injuries is very

important in these patients because if injury occurs it may further worsen the disability from which they are already suffering. The next major important aspect is the sexual functions. Hypermobility patients have problems in sexual functions because of depression and anxiety whereas ankylosing spondylitis patients find difficulty due to ankylosis of hip joint, pain, stiffness and kyphotic deformity of the spine. Therefore adequate counseling is required.

In conclusion coexistence of these two opposite disorders causes confusion in the diagnosis and management and leads to double disability. Hypermobility can be utilized for a stiff joint. By taking proper care of the joints and spine with proper exercises, soft tissue injuries and further deformities can be prevented.

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