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Still's disease : a distinct pathological entity?

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STILL'S DISEASE

A Distinct Pathological and Etiological Entity?

By

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INTRODUCTION

Arthritis or rheumatism is an old disease, older in fact than man; but for all its antiquity much of its nature is unknown, or what is possibly a better term, obscure. This is especially true of chronic non-tubercular arthritis. A great amount of work has been done in attempting to unveil the mysteries of these conditions; however, since chronic arthritis remains pretty much a mystery, in many respects, there is much confusion. The terminologies and classifications in use are almost as varied as the number of authors writing on the subject. In the body of this paper the terminology used in the original article is employed and so to make clear the terms used, there is incorporated in this introduction a chart (taken from Pemberton and Osgood, which lists the various terms used by the many authors under the proper classification as accepted by the American Committee on the Problem of Rheumatism.)

So extensive and so voluminous has been the work on chronic arthritis that no attempt has been made to discuss the entire subject, but only those articles that directly relate themselves to the subject proper of this paper are considered.

The literature herein used is almost entirely American or English in origin and any reference to articles written in a foreign language have been indirectly taken from reviews of English or American writers.

CHIEF CLASSIFICATIONS OF CHRONIC ARTHRITIS

Name	Atrophic	Hypertrophic
Adams-1857	Polyarticular	Monarticular
Charcot-1881	Rapid Evolution	Gradual Evolution
Garrod-1890	Rheumatoid Arthritis	Osteo-arthritis
Bannatyne-1896	Rheumatoid Arthritis (Early)	Rheumatoid Arthritis (late)
Goldthwaite- 1904	Infectious Arthritis Atrophic Arthritis	Hypertrophic Ar- thritis.
Nathan-1907	Chronic pseudo-rheumatism Chronic secondary artic- ular rheumatism Rheumatoid Arthritis	Osteo-arthritis deformans
Hoffa and Wollenberg- 1908	Secondary chronic articular rheumatism. Primary progressive polyarthritits	Osteoarthritis deformans.
Llewellyn Jones-1908	Rheumatoid Arthritis	Osteoarthritis.
Nichols and Richardson 1909	Proliferating or Ankylosing arthritis	Degenerative or non- ankylosing arthritis.
Osler and McCrae	(a) Lesions principally in synovial membrane; (b) Atrophic changes in cartilage and bone.	Hypertrophy and over- growth of bone.
Ely-1914	Type I Primary proliferation of synovial membrane and marrow. Secondary atro- phy of cartilage and bone.	Type II Primary inflammation of synovial membrane and degeneration of synovial membrane and marrow. Secondary hypertrophy of bone and cartilage.

Name	Atrophic	Hypertrophic
Fisher-1923	Type II Primary synovial membrane invasion. Secondary cartilage and bone invasion.	Type I Primary cartilage and bone invasion. Secondary synovial membrane invasion.
	Type III (Mixed) Simultaneous invasion synovial membrane, cartilage and bone.	

STILL'S DISEASE

A Distinct Pathological and Etiological Entity?

In 1896, Still, in a paper read before the Medical and Chirurgical Society of London, described a form of chronic arthritis occurring in children which he claimed was sufficiently distinct to be differentiated from the usual form of rheumatoid arthritis that occurred in adults and on rarer occasions in children. He defined this disease as a chronic progressive enlargement of the joints of the body associated with a general enlargement of glands and enlargement of the spleen. He used for this study a series of twenty-two cases of arthritis occurring in children, twelve cases of which presented the above picture. He further stated that the disease usually had its onset before the second dentition and that the onset may be either insidious or acute. He found girls to be more often afflicted than boys, and established this ratio at 1:5 to one. He described the enlargement of the joints as being smooth and fusiform and of the periarticular tissue in contra-distinction to bony enlargement as it occurs in some forms of arthritis. He found no osteophytic growth or bony lipping even after several years had elapsed.

There was an absence of redness and tenderness except in the acute cases but the limitation of motion was most marked. The disease tho polyarticular in nature effected the knees,

wrists, and cervical spine earliest and then later involved the ankles, elbows and fingers in the order mentioned. Noteworthy also was the symmetrical character of the disease. There developed no bony ankylosis; and suppuration of joints or glands did not occur. Glandular enlargement was noted in most of the peripheral lymph glands and particularly in the region of the joints affected. Significant also was the fact that the glands and the spleen fluctuated in size, enlarging during an attack and subsiding with the attack. Further he found no valvular heart disease, but did note adhesive pericarditis to be present in a good percentage of cases. Characteristically, each of the twelve patients were slightly anemic.

Still stated the disease was chronic and progressive with occasional intermissions. The disease was described as not being dangerous to life. He also noted spontaneous recovery in some instances following an acute disease. Recovery with no residual effects of the disease occurred in some cases, others continued and grew progressively worse. Others died of some inter-current infection. As a result of the death of three patients, three autopsies were performed and Still found the following: The joints showed marked hyperplasia and increased vascularization of periarticular tissue. There was no bone damage or bony overgrowth. The cartilage was intact except for occasional pitting of the cartilage about the periphery. The glands showed marked

Personal inference or opinion has been avoided as much as is possible since this thesis has as its purpose to report on the present status of the question - Is Still's disease a distinct pathological and etiological entity?

hyperplasia, but the organization was perfectly normal otherwise. No tuberculosis was found. The spleen exhibited hyperplasia but its markings were distinct.

In presenting his paper, Still argued that to his knowledge glandular enlargement and splenomegaly did not occur in the adult form of rheumatoid arthritis. Furthermore he felt the joint pathology was sufficiently distinct to warrant questioning the similarity of rheumatoid arthritis and the syndrome he described, pointing out that cartilaginous destruction, ankylosis and bony over-growth did not occur in his cases. Moreover, he argued that the difference in the pictures presented by the two forms of arthritis were not due to the ages of the patients, since the adult form of rheumatoid arthritis did occur in children, altho more rarely than did his form of arthritis. Significant also, according to Still, was the order and frequency in which the joints were affected. He pointed out that in the adult form the small joints were often first to be affected whereas in the disease he described the larger joints were involved first and the smaller joints secondarily. The absence of valvular heart disease in his cases and the findings of pericarditis in five of the twelve cases were felt to be particularly characteristic of the form of arthritis he described. Still did not attempt to prove an etiology for the form of arthritis his cases represented, however he did believe they were of an infectious nature.

Previous to Still's article little had been written of chronic arthritis in children. Moncorvo,⁽²⁵⁾ in 1878, reported fifty cases of chronic arthritis in children but nothing significant was recorded.

Barlow, in 1888, reported a case of an eleven and one-half year old girl with chronic arthritis showing fusiform swelling of joints and enlargement of lymph glands. The child was anemic and showed a definite leucopenia. In the same article he reported the case of a boy presenting the same picture clinically plus splenomegaly. A post mortem was performed on the boy and the pathology described agreed with the pathology described by Still. In fact, it is reasonable to assume that the latter case was included in Still's series of cases, since Still thanked Barlow among others for use of their cases.

Noteworthy was an article written by Chauffard and Ramond⁽²⁶⁾ earlier in the year 1896, in which they described a series of cases of chronic arthritis in adults with glandular enlargement and splenomegaly. Still acknowledged their priority but claimed the cases of Chauffard were sufficiently different clinically to consider them not the same disorder. At the same time Bannatyne and Wohlmann reported arthritis in adults with glandular enlargement.

Later, in 1897, Still in a lecture printed in the Clinical

Journal of London, reiterated his claims for the distinctness of his syndrome and offered a differential diagnosis for the various forms of arthritis, acute and chronic, occurring in children.

The work of Still and Chauffard and the relatively recent work of Pasteur stimulated interest in chronic arthritis and a great many writers appeared on the horizon, all attempting to prove a definite etiology for the disease. A great amount of work was done, much of it conflicting and inadequate, but all aiding in a better understanding of the subject.

Hunt, in 1898, described the case of a boy six with arthritis, glandular enlargement and splenomegaly which followed an acute tonsillitis. He attempted to prove an infectious etiology, but on aspiration of the joints involved, he found them to be free of organisms. The case, though typical of Still's Disease, did not prove or disprove anything.

In 1899, there was recorded in the St. Thomas Hospital report of London, the case of a child, age five and one-half, with arthritis showing enlarged glands and splenomegaly, with anemia and involvement of the spine. The patient was given cod liver oil and improved temporarily, returning with the same complaint one year later. Also reported was the case of a twenty-one year old woman with rheumatoid arthritis accompanied

by glandular enlargements and splenomegaly. She improved under care which consisted for the most part of physical therapy.

Then in 1902, Lemke reported the case of a boy age eleven with rheumatoid arthritis and glandular swelling and splenomegaly showing a leucopenia and anemia and involvement of the spine. He ruled out tuberculosis as a possible etiological agent by biopsy of a gland. He found the joints to be free of bacteria. Further he was of the opinion that the pathology found present in his case was very similar to that present in the adult form of the disease.

Shortly after this McCrae, in 1904, in a comprehensive review of arthritis dismissed Still's disease by saying he believed it to be a part of the general picture of atrophic arthritis, and stated that he believed it to have an infectious etiology.

In a lecture on rheumatoid arthritis, Herringham, in 1909, offered the opinion that the glandular swelling and splenomegaly occurring in Still's Disease was due to a difference in the age of the patients, rather than a different etiological agent, and that Still's Disease and adult rheumatoid arthritis were one and the same condition. He gave no proof for his statement.

Nichols and Richardson did much to clarify the problem of

chronic arthritis when they published their comprehensive work in 1909. Their paper was based upon a pathological and clinical study of sixty-five cases of chronic, non-tubercular, deforming arthritis. Their work included cases of Still's Disease. They concluded that these joint lesions can be divided with great definiteness into two pathological groups:

1. Those which arise from primary proliferative changes in the joints, chiefly in the synovial membrane and in the perichondrium.
2. Those which arise primarily as a degeneration of the cartilage.

The first type they called proliferative arthritis and the second type degenerative arthritis. They stated further that these two pathological groups are characterized by distinct gross and histological differences, but added that these two pathological types, however, do not correspond to two definite etiological factors; that is, two definite and distinct diseases. Thus it became evident that the reaction of joint tissue to many and variable etiological agents were limited; so that although the etiological agent may be quite variable, the pathology is the same - being either the proliferative or degenerative type.

Since the work of Nichols and Richardson has been confirmed by many succeeding writers using a better, more modern technic

and having available a larger amount of material with which to study, and since their work deals with material visible and fixed, one cannot but accept it as being so. As a result of their work it became evident that Still's Disease could not claim a distinct pathology; but this, as they pointed out, did not indicate an identical etiology or even a similar etiology. Thus the claim of a distinct etiology for Still's Disease remained to be proved or disproved.

Barker in his book "Monographic Medicine" stated that he believed Still's Disease to be one of the infectious arthritides and not a distinct entity. He based this statement on the clinical findings and the rather indefinite information that was accumulating in the literature on focal infection as a cause or the cause of rheumatoid arthritis.

Then in 1912, Luff differentiated Still's Disease from rheumatoid arthritis on the basis of clinical findings, using the following as differential points:

1. Enlarged glands.
2. Enlarged spleen.
3. Peculiar feel of the joints and the absence of bony grating and the absence of osteophytic out-growths.
4. And that the disease began in the knees and

wrists and affected the fingers later.

Here again the differences in the clinical picture were brought out and although it seemed to indicate a different etiology, it does not prove it.

Lichfield and Mason, in 1922, reported a typical case of Still's Disease and concluded that although they were of the belief that Still's Disease was not a distinct disease as differentiated from rheumatoid arthritis in adults, it was sufficiently distinct to warrant special mention.

Typical of the findings in the literature from about 1900 to the present, are the following two articles: Rosenfeld, in 1917, reported a case of Still's Disease which he attributed to a focus of infection in the teeth. On removal of the focus, the patient improved. And further, injection of a culture from the focus into rabbits resulted in arthritic signs and symptoms in the rabbits. The organism was a type of streptococcus. Poynton, in 1925, reported a case of Still's Disease with definite foci of infection, which were removed with no improvement in patient. The case was typical and the foci as evident as in Rosenfeld's case, but the ultimate results of removal of the foci were directly opposite. These two articles are particularly significant, since Poynton was a firm believer in the theory of focal

cannot, on the basis of a white count alone, differentiate two diseases or syndromes so characteristically similar otherwise, except for the ages of the patients.

The work of Felty, which served as a new impetus to writers on this subject, was followed by several articles reporting similar cases. In 1936 Williams reported a case of Felty's Syndrome with autopsy report. The case was typical of those described by Felty. The autopsy report was not particularly valuable, except that culture of the spleen was negative while that of the lung yielded a streptococcus viridans. Williams was of the same opinion as Felty as to its classification, likening it to a Still's type of arthritis.

Fitz, in 1935, reported a typical case of Felty's Syndrome. He used liver and iron as treatment. On this one point he raised the question of similarity of Felty's Syndrome to atrophic arthritis and Still's Disease. He states that liver and iron therapy had marked effect in gout and Still's Disease, but produced no effect in his case of Felty's Syndrome. Of course, differentiation of Still's Disease from Felty's Syndrome on the basis of one patient and a single therapeutic measure is wholly inadequate; but further study along this line is indicated.

In 1936 Singer and Levy reported two cases of Felty's Syndrome, with autopsy. The cases were quite typical and autopsy

results showed streptococcus viridans in both spleens. Singer and Levy concluded, on the basis of a review of the literature by continental European writers and their personal observations, that Felty's Syndrome and Still's Disease were not distinct entities but a part of the general picture of atrophic arthritis. They reviewed the foreign literature, listing many authors who reported cases of arthritis plus adenitis and splenomegaly; and so concluded that adenitis and splenomegaly is not so rare a finding as some persons are prone to believe.

In an article which called attention to the arrested growth in Still's Disease, Kuhns and Swain, in 1932, eliminated syphilis and tuberculosis as possible causes of the disease. This article is not particularly significant except that a certain few continental writers were still of the belief that Still's Disease had a tubercular origin.

Dawson, in "Nelson's Loose Leaf Medicine", states that although striking in some of its characteristics, it is identical with adult infectious arthritis in that the pathology, blood findings, and prognosis are the same.

In confirmation of Dawson's statement, Blair and Hallman, in 1935, in experimental studies on blood of atrophic arthritis, showed a high agglutination for streptococcus hemolyticus and a high streptolysins titer in practically all cases of atrophic

arthritis, including Still's Disease.

Keefer, in 1935, in an article discussing the etiology of atrophic arthritis, makes the statement that glandular swelling is a side reaction, not characteristic of atrophic arthritis, but occurring in some cases. He is of the opinion that Still's Disease is a part of the general picture of atrophic arthritis and divides the pathological lesions of rheumatoid arthritis in to three parts, as follows:

1. Primary-Synovitis, periarticular changes with or without subcutaneous fibroid nodules.
2. Secondary- Destruction of cartilage, atrophy of bone, new bone formation, sublimation, ankylosis (fibrous and bony), muscular atrophy.
3. Incidental lesions- Lymphoid hyperplasia, calcification of blood vessels, amyloidosis, growth disturbances, and pigmentation of skin.

Six rather interesting cases from a series of two thousand cases of chronic arthritis were reported by Kauffman in 1937. These six cases were all women showing atrophic arthritis plus glandular swelling, which produced acute abdominal symptoms during an acute attack of the arthritis. Kauffman also states he believes glandular swelling to be a rare occurrence of atrophic arthritis and not significant otherwise.

Cohen, in 1937, in attempting to prove an allergic basis for chronic arthritis, stated that he is of the opinion that Still's Disease is not a distinct entity but a part of the general picture.

In the same year Colver, in reporting on the prognosis of Still's Disease, stated that one in four recover, and those patients which survive the first three years of the infection are in no danger of life. This is interesting, in that all cases of Felty's Syndrome that have been reported as such have all ended fatally.

Then from the article "The Present Status of the Problem of Rheumatism", for 1936, comes the statement by Meltke that Still's disease is not a distinct entity, since polyadinitis is present in thirty-seven percent of cases of adult atrophic arthritis; and only the pericarditis present variably in Still's Disease occurs rarely in adults, all other findings being found in adults. (10)

The same review for the following year gives the percentage of cases of atrophic arthritis presenting glandular enlargement as varying between forty percent and fifty-three percent, quoting as their authority Douthwaite's article of 1933 and Coates and Delicati's article of 1931. Further, they state splenomegaly occurs in this type of arthritis in ten percent to fifteen percent

of cases according to Coates and Delicati. (11)

In the same article, Monorieff is recorded as saying he believed Still's Disease to be a distinct entity, a statement with which the authors of the article disagree. Also, in the review, Castellani is reported to have written an article in which he states Felty's Syndrome and Still's Disease to be distinct entities. (11)

Then in the same review for the next year, Collins is reported as claiming Still's Disease and Felty's Disease are rare varieties of atrophic arthritis.(12)

Of far greater significance than any of the above statements is the opinion of Pemberton and Osgood, as made in their very complete review and discussion of chronic arthritis in the book "The Medical and Orthopaedic Management of Chronic Arthritis". They conclude that Still's Disease is not a distinct entity, and that all the features and characteristics of Still's Disease are found sufficiently often in atrophic arthritis to make them of little value in attempting to differentiate two so similar conditions.

COMMENT

After reviewing the literature, one can arrive at certain rather definite conclusions as to exactly what is known and what

is not known. Thus the joint pathology of Still's Disease is known to be the same as that found in atrophic arthritis and not different as at first thought by Still, so that now claims for a distinct pathology for Still's Disease are unfounded. Also known is the fact that evidently glandular enlargement and splenomegaly are not so rare an occurrence in arthritis and may be found at any age. Most atrophic arthritis patients are known to be anemic and the white count is not particularly characteristic except in the case of Felty's Syndrome. In all probability, however, Felty's Syndrome merely represents either a more virulent form of the disease, or what is even more likely, the same condition and the same degree of virulence as found in other forms of atrophic arthritis but occurring in a patient of markedly decreased resistance.

Furthermore, it is known that blood findings in the cases of Still's Disease, Felty's Syndrome, and other forms of atrophic arthritis, are similar. It is also apparent that the prognosis of these afflictions, the less favorable at both ends of the age ladder is pretty much the same.

The unknown feature of the whole problem of atrophic arthritis is the etiology. The exact cause is as yet unknown; but from all indications, the condition seems to have an infectious basis. Furthermore, it appears quite likely that some strain,

or possibly strains, of the streptococcus organism is to blame; however, this is not proved. An allergic basis for the disease is claimed by some authors, and still others are in favor of a metabolic etiology for the affliction. These latter two theories have less foundation in fact than does the infectious theory. There are unknown quantities in all three theories, and the ultimate result may be that all three are wrong. It is interesting, however, that our concept of allergy is constantly changing, and recent work as yet unpublished seems to indicate a more probable allergic theory along the lines of our new understanding of the subject.

CONCLUSIONS

1. The joint pathology of Still's Disease and atrophic arthritis are one and the same, but this does not indicate an identical etiology.
2. The etiology of Still's Disease and atrophic arthritis are as yet unknown; however, the evidence accumulating in the literature seems to point to a similarity of the two.
3. Still's Disease, though striking in many of its characteristics, cannot claim to be a separate disease entity on the basis of the proof offered in the literature to date.

4. However, the evidence points to a similarity of the two conditions; one cannot definitely prove that Still's Disease is not a distinct etiological entity.

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