MARIE-STRÜMPPELL SPONDYLITIS IN WOMEN

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A form of chronic arthritis of the spine characterized by certain distinguishing features was described by Marie (1898), Strümpell (1897), and von Bechterew (1899), at the turn of the 19th century. It is not an uncommon disease, but is very often unrecognized and from time to time articles appear in the medical literature calling attention to its importance in the differential diagnosis of low back and sciatic pain. In all of these articles, it is generally agreed that the disease is much more common in males than in females, the stated ratio varying from 4 : 1 (Simpson and Stevenson, 1949) to 10 : 1 (West, 1948).

It is usually assumed that the disease runs a similar course in both sexes, and this investigation was undertaken in order to study this feature. It occurred to us that there might be differences in the clinical manifestations and course of the disease which made the difference in sex distribution more apparent than real.

In the male we usually find an onset in early adult life with persistent, though at times relenting, progression. The severe systemic effects of the disease are chiefly characterized by loss of weight and strength, extensive muscle atrophy, and a marked tendency to forward-flexion deformity of the spine. The pallor of chronic disease is present in most cases, but is often not correlated with the blood picture. The pain of ankylosing spondylitis is its most prominent feature, and it is often paradoxical in that, though the pain is severe, it cannot be accurately located or described by the patient. Its most characteristic feature is that of being at its worst in the early hours of the morning or following any prolonged period of rest. The extensive ankylosing features are marked in males, and eventually lead to fixation of the spine and its adjacent articulations. The end-result is a pathetic picture of a thin, chronically ill, discouraged man standing stooped over with flexed hips and knees, and arms and head hanging forward.

Material of Present Study

In the past 20 years, sixty authenticated cases of Marie-Strümpell arthritis in women have been under observation in the combined arthritis clinics of the Presbyterian and New York Orthopaedic Hospitals. During the same period, over 450 cases have been observed in male patients. Cases admitted to these specialized clinics represent only the more obvious and severe examples of the disease, since all the patients will first have been treated by family physicians and have found their way into the general medical and orthopaedic clinics, whence, due to obvious findings or persistence of symptoms, they have been sent to the arthritis clinics.

The disease is generally similar in both sexes, but striking differences in the course and behaviour of the disease also appear. In the female, the marked systemic manifestations (such as weight loss, loss of appetite, pallor, exhaustion, and tendency to deformity) are usually lacking. This discrepancy in severity of symptoms may well account for some of the differences noted in the incidence of the disease in both sexes, since the milder manifestations in women may often be regarded as cases of chronic backache, sciatica, or referred pain from pelvic abnormalities, rather than as true cases of spondylitis ankylopoietica. In fact, the medical profession tends to be prejudiced in favour of the male having the disease, so that the evidence must be overwhelming in the female before the diagnosis is even entertained. Only seven of the sixty women in our series showed a degree of severity comparable to that usually seen in the male.

Age.—The age distribution of our sixty cases of Marie-Strümpell arthritis in women ranged from 22 to 60 years when first seen (average 32). The onset of the disease was often vague, the age of onset varying from 14 to 59 years (average 25).

Symptoms.—The site of the first symptomatic complaint referable to the osseous system is shown in Table I (opposite). The majority of patients first noticed symptoms in the low back region.

Previous or Concurrent Disease.—Sciatica also occurred at some time in more than half the cases (36 cases), eight of the sixty patients (13 per cent.) had a history of iritis, and some form of menstrual disorder was present in
22 per cent. Four patients had definite rheumatic fever in the past and four had had chorea.

Family History.—Marie-Strümpell arthritis was found to occur frequently in siblings and parents. In six patients (10 per cent.) there was a definite family history of Marie-Strümpell spondylitis. One woman had two brothers with spondylitis. A high incidence of rheumatoid arthritis was also noted; it was found in the immediate family of nine (15 per cent.).

Physical Examination.—The involvement of the spine and peripheral joints is shown in Table II. In contradistinction to the clinical picture usually found in males is the high incidence of involvement of the cervical spine. The involvement of shoulders and hips is about the same as in males. The involvement of the peripheral joints is the same as that in males. Three patients had psoriasis and six had enlarged hearts (four with definite evidence of mitral stenosis, one of whom was fibrillating). In seven patients the disease was severe with marked deformities, and the remainder had only mild or moderate deformities. Subcutaneous nodules were not found.

Radiological Examination.—The changes found in these patients by x ray are shown in Table III. To be noted is the high incidence of involvement of the sacro-iliac joints and the apophyseal joints of the lumbar spine. Involvement of the apophyseal and syndesmophyte joints occurred in more than 10 per cent.; it is seen only rarely in males (Golden and Tyson, 1952).
of the relative infrequency with which the disease is reported in women. This series of cases shows that the disease does appear in women and should be included in the differential diagnosis of all complaints referable to the spine.

REFERENCES

Spondylite Marie-Strümpell chez la femme

La spondylite Marie-Strümpell semble avoir une évolution plus sereine chez la femme que chez l'homme. A part l'atteinte symptomatique plus fréquente de la région cervicale et le preuve radiologique de l'atteinte de la symphyse pubienne, les images cliniques chez l'homme et chez la femme semblent peu différents. Le caractère un peu moins sévère chez la femme peut mener à un diagnostic erroné et même faux, mais il ne semble pas que ce soit la raison de la rareté relative des cas féminins publiés. Les cas relatés ici montrent la réalité de la spondylite Marie-Strümpell chez la femme, et la nécessité d'en tenir compte dans le diagnostic différentiel de la symptomatologie vertébrale.

Espondilitis Marie-Strümpell en la mujer

La espondilitis Marie-Strümpell parece tener un curso más benigno en la mujer que en el hombre. Aparte del compromiso sintomático más frecuente de la región cervical y del compromiso radiológicamente demostrable de la sinfisis pública, los cuadros clínicos en el hombre y en la mujer difieren poco. El carácter algo menos grave de esta enfermedad en la hembra puede llevar a un diagnóstico erróneo y hasta falso, pero esto no parece ser la causa de las observaciones relativamente raras de esta enfermedad en la mujer. Los casos relatados aquí muestran la realidad de la espondilitis Marie-Strümpell en la mujer así como la necesidad de tomarla en consideración en el diagnóstico diferencial de la sintomatología vertebral.
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