GENERALIZED MYOSITIS FIBROSA: CASE REPORT

BY

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Generalized myositis fibrosa is rare, and its clinical appearance and behaviour are so well defined that it is considered justifiable to report a case recently observed.

Case Report

The patient, a single woman aged fifty-four years, had worked for four years as a radio-condenser assembler. She had had asthma from 1930 to 1941 for which she had been treated by an autogenous Streptococcus viridans vaccine prepared from a nasal swab, with improvement of symptoms. Skin tests against animal fluffs and food proteins had all been negative. All her teeth were removed for pyorrhoea alveolaris in about 1930. For six months previous to observation she had suffered intermittently from iritis in the left eye. She had never worked for many years, and had a nasal deformity of the left nasal spine and alae nasi; both hands, but no evidence of involvement of the joints concerned: wrist movements were full and free with the fingers partially flexed. Passive extension of the wrist caused flexion of the fingers, and vice versa. The extensor and flexor groups of muscles of both forearms felt firmer than is normal, having the texture of a muscle in spasm, though this was evidently not the cause of the induration. The small muscles of the hand appeared normal. There was marked equinovarus and cavus deformity of the feet, with hallux valgus and hammer toes, so that when the patient tried to walk the metatarsals were placed almost vertically, the anterior part of the foot being acutely plantarflexed on the talus (see illustration). This deformity had evidently arisen as a result of simultaneous contracture of the toe flexors and extensors. The tendo achillis was also markedly shortened, and there was minimal movement in the ankle joint and all the joints distally. The calf muscles appeared wasted (R > L) and all the leg muscles felt indurated. Hip and knee movements were normal, and there were no skin changes.

The respiratory, cardiovascular, alimentary, and genito-urinary systems were clinically normal; the nervous system showed only the pupillary changes noted, with absent ankle jerks and plantar responses which were accounted for by the deformity present.

Muscle biopsy from the left flexor digitorum profundus had been performed six weeks earlier. Sections from the tissue excised showed some fibres thinner than normal, with diminution of striation, collections of nuclei between muscle fibres, and some small round-celled infiltration. No evidence of fibrosis was found in the sections examined from this biopsy, and it was not considered justifiable to repeat the procedure in order to confirm the fibrosis.

Special Investigations.—The following further investigations were made. A radiograph of the cervical and lumbar spine showed slight osteo-arthritis changes. Radiographs of the arms and legs showed only the deformities described; there was no evidence of abnormal calcification in the soft tissues. A chest radiograph showed mild emphysema and a radiologically normal heart. The cerebrospinal fluid was normal.

The Wassermann reaction was negative in the blood and cerebrospinal fluid. The blood count was: Hb 100 per cent., red cells 5·4 million per c.mm. of blood, colour index 0·9, white cells 7,000 per c.mm., polymorphs 61 per cent., eosinophils 8 per cent., basophils 1 per cent., monocytes 12 per cent., lymphocytes 18 per cent.

The sedimentation rate was 15 mm. in the first hour (Westergren).

Plasma inorganic phosphorus was 4·6 mg. per 100 ml.
GENERALIZED MYOSITIS FIBROSA

Ellerman (1943) reported a case in which the skin was adherent over many of the widely scattered but relatively localized muscle lesions.

Benedek (1944), under the title pseudomyasthenic syndrome in polymyositis interstitialis chronica fibrosa, described a case which clinically resembled myasthenia, but showed the histological changes of myositis fibrosa.

Aetio logically no factor can be recognized which is in any way constant for these cases. Diagnosis is based on the clinical behaviour of the condition and the biopsy findings. Its onset is insidious, usually with stiffness affecting the legs, arms, or back, and its course chronic, progressing to contractures in the groups of muscles affected, with coincident wasting. Further progress may be prolonged through a series of remissions and exacerbations extending over many years. Pain is absent, the pain in the case here reported being due to the foot deformity rather than directly to the condition in the muscles.

The affected groups of muscles are characteristically indurated, and impart a sensation to the observer variously described as "doughy", "boggy", "firm", "like a sandbag". It may spread widely to affect practically the entire muscular system, though the face is spared. The skin usually remains normal throughout its course., thus distinguishing it from dermatomyositis, which in the earlier stages is the most likely differential diagnosis. The course is afibrile. Degenerative, inflammatory, and fibrotic changes are found histologically in muscle biopsies from affected areas. Hyaline and vacuolar degeneration may be seen in muscle fibres, which often appear granular and structureless with loss of striation. Round-celled infiltration is found round degenerating fibres, and fibrous tissue replacement is found in the more advanced lesions.

Bodansky and others (1929-30) investigated the creatine and creatinine metabolism fully in their patient, a negro boy of fourteen years who also suffered from pulmonary tuberculosis. The age and coincident disease in this case would render their conclusions questionable, but they demonstrated an inability to retain exogenous creatine; 40 per cent. of the total creatine and creatinine was excreted as creatine.

It is considered most likely that the iritis in the case here reported was due to the presence of a foreign body and was not connected aetio logically with the general disease process.

Treatment has usually been conservative and confined to measures designed to maintain and improve function in the affected groups of muscles. Drug therapy has produced no significant benefit, and

Radiograph of one foot of the patient here described. The appearance in the other foot was almost identical.

of blood, calcium 10 mg. per 100 ml., potassium 24 mg., and uric acid 2·9 mg. A twenty-four-hour specimen of urine contained 110 mg. of creatine and 274 mg. of creatinine.

The electrocardiogram and electrical reactions were normal.

Discussion

From a review of the literature it would appear that the condition is rare; it is difficult to determine the precise number of cases recorded, as some are inadequately described and in some the diagnostic criteria are not completely satisfied, but it would appear to be about thirteen.

Burton and others (1923-4) reviewed the cases previously described, accepted five as being generalized as opposed to the localized form of myositis fibrosa, and added one of their own. They reported the progress of their case after eight years, after a prolonged remission (Burton and others, 1931). Ornstein (1935) reported the condition in a forty-two-year-old male in whom the spinal muscles were principally affected. The case superficially suggested a form of myotonia, but biopsy showed the usual histological changes of myositis fibrosa.

Blau (1938), in a full review, accepted eight previous cases and added two of his own.

Somers (1939) reported a patient who had suffered from asthma which had been relieved by removal of nasal polypi.
Somers (1939) manipulated his patient, who died shortly afterwards from what was thought to be coronary occlusion.

Summary

A case of generalized myositis fibrosa is reported. The subject is briefly reviewed.

I wish to thank Dr. G. L. S. Konstam, under whose care the case was observed, for permission to report it; Dr. B. Buckley Sharp for the biopsy sections, and Mr. H. G. Blunt for the reproduction of the x-ray photographs.

REFERENCES


Myosite Fibreuse Generalisée : Observation

RÉSUMÉ

Environ treize cas de myosite fibreuse généralisée ont été rapportés dans la littérature. L'auteur de cet article en décrit un quatorzième : la maladie est une femme célibataire âgée de 54 ans, qui a travaillé pendant quatre ans à l'assemblage de condensateurs de radio. Elle a été atteinte d'asthme et d'irrité. La maladie a affecté les doigts des deux mains, les muscles des avant-bras, les pieds, et les muscles des mollets. Le sujet est passé en revue brièvement.

ABSTRACTS

[This section of the ANNALS is published in collaboration with the two abstracting Journals, Abstracts of World Medicine, and Abstracts of World Surgery, Obstetrics and Gynaecology, published by the British Medical Association. The abstracts are divided into the following sections: acute rheumatism; chronic arthritic rheumatism (rheumatoid arthritis, osteo-arthritis, spondylitis, miscellaneous); sciatica; gout; non-articular rheumatism; general pathological articles; other general articles. At the end is a list of articles that have been noted but not abstracted. Not all sections may be represented in any one issue.]

Acute Rheumatism


This is a somewhat procrastinate attempt to fit the changes in the coronary arteries, as found in some cases of acute rheumatic disease, into the three stadia described by Klinge. The changes described here are those of the rheumatic process, not the precocious atheroma, which is rare despite the cardiologists who quote it so often. The author also refuses to follow the fashion of saying that this is just a variant of polyanteritis nodosa (Kussmaul). He emphasizes [rightly] that varying degrees of myocardial infarction are not infrequently revealed on microscopy of the rheumatic heart, a fact that has not been widely appreciated. He discusses the appearances in the coronary in the light of recent work on experimental hyperergic lesions. A. C. Lendrum.


This paper is based on the correlation of the radiographic and clinical findings in the course of a follow-up in 1944-45 of 494 patients who, as children, had one or more attacks of rheumatic fever. In nearly 22% of the cases the radiographic findings were important for deciding the diagnosis and prognosis.


In order to assess the importance in rheumatic children of a loud blowing systolic murmur in the absence of demonstrable cardiac enlargement a comparison was made of the after-history of 144 children having such a murmur with that of 171 similar patients with potential and possible heart disease but without more than a slight systolic murmur.

The observations suggest that the diagnosis of mitral insufficiency, based on a loud blowing apical systolic murmur, is justified in children and carries a grave prognostic significance. H. E. Holling.


The author analyses a follow-up study of 547 children observed for 10 years after the onset of rheumatic fever. At the end of the 10-year period over 90% of the group had been located. At the end of the initial attack 41% gave no evidence of heart disease; of these, over 76% still showed no evidence of heart disease at the end of 10 years, while only 5% had died of rheumatic infection or bacterial endocarditis. Over 40% of children with rheumatic heart disease in the first attack had died by the end of 10 years of rheumatic infection or bacterial endocarditis, while in just under 10% the signs of cardiac involvement had disappeared. The results agree with...