The history of polymyalgia rheumatica/giant cell arteritis

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ABSTRACT

Only two years separate the initial descriptions of polymyalgia rheumatica (PMR) by Bruce in 1888 and giant cell arteritis (GCA) by Hutchinson in 1890. However, the existence of an association between these two conditions was definitely accepted only in 1964.

Even if PMR is generally accepted as a different disease from GCA, some authors deem PMR to be a manifestation of a generalised arteritis and use the term GCA to define PMR/GCA as a whole. The time has come to clarify this issue.

Introduction

In this paper I would like to try to describe separately the history of polymyalgia rheumatica (PMR), the history of giant cell arteritis (GCA) and the history of PMR/GCA as a whole.

Polymyalgia rheumatica

The first description of polymyalgia rheumatica was made by Bruce (1) (Table I) in 1888, who defined as "senile rheumatic gout" a painful condition involving the large joints of the girdles, with non-ankylotic stiffness. While compromising the general condition of the patient, the gout generally resolved within one or two years.

Thereafter no further observations were published on the subject until the end of World War II, in 1945, when a reference appeared which, in the light of recent findings, seems to be a forerunner: that is, the description of "peri" or even "peri-extra" articular involvement. Meulengracht outlined such a specific localization at the girdle that it could be confused with scapulo-humeralis periarthritis, as indeed was evoked in the denomination which he proposed, "peri-arthrosis humero-scapularis" (2). In a similar report, Holst and Johansen used the term "peri-extra-articular rheumatism" to describe 5 patients with a "special type of rheumatic disease", special in part because of the severe general involvement and their highly increased ESR (reaching 80 mm/ hr) (3).

In 1951 at the 2nd European Congress of Rheumatology in Barcelona, a complete description of the disease was made by Kersley in a presentation entitled "A myalgic syndrome of the aged with systemic reaction" (4). He reported 13 patients (median age 71 yrs) who presented with typical pain and stiffness of the girdle (and sometimes of the hands!), but also a severe general state of impairment and biological signs of strong inflammation. The disease spontaneously reversed with time. Kersley's presentation is historically important for another two reasons: it reports the first successful use of steroids and the typical negative muscle biopsy.

Table I. History of polymyalgia rheumatica (PMR).

Authors	Year	Name
Bruce	1888	Senile rheumatic gout
Holst & Johansen	1945	Peri-extraarticular rheumatism
Meulengracht	1945	Peri-arthrosis humeroscapularis with general symptoms
Kersley	1951	Myalgic syndrome of the aged with systemic reaction
Porsman	1951	In arthritis in old age
Bagratuni	1953	Anarthritic rheumatoid disease
Forestier & Certonciny	1953	Pseudo-polyarthrite rhizomelique
Barber	**1957**	**Polymyalgia rheumatica**
Levey	1963	PMR: A separate entity?
Ballabio	1971	Part of fibromysitic rheumatism

The first French publication, by Forestier and Certonciny, is also of particular interest in the context of the work that will be presented at this Conference (5). They once again stressed the localization at the fibrous structures, sheaths and tendons, and emphasized that the articular synovial structures are almost entirely spared.
In the same year Bagratuni described in great detail a group of patients with PMR as having "anarthritic rheumatoid disease" (6). In following the clinical course of these patients for more than 10 years, he did not observe any progression in erosive arthritis and, curiously enough, judged this disease to be a mild form of rheumatoid arthritis.
The term "polymyalgia rheumatica" was proposed by Barber in 1957 (7). This is the term which has received general acceptance and lasted in time. Interestingly, Barber describes in this context the possible flogistic involvement of the spine.
In the same period Gordon confirmed that the response to steroids was such a significant parameter that it qualified as a diagnostic criterion of the disease (8).
In 1963 the first case under this name was reported in the American literature by Levey, but still with a question mark: "PMR: A separate entity?" (9).
At different times various Italian researchers have suggested changing the name for the condition, such as for example Ballabio who coined the term "reumatismo fibromiositico [fibromyositic rheumatism]", considering the condition to be a variant of the inflammatory rheumatic diseases (10). Since then, at least in Italy, this rather misleading name has lasted.

Table II. History of giant cell arteritis (GCA).

Authors	Year	Name
? ?	1350 B.C.	Sine nomine
Ali' Ibn Isa	1000 A.C.	Sine nomine
Hutchinson	1890	Form of thrombotic arteritis
Horton	**1932**	**Arteritis of the temporal vessels**
Gilmore	**1941**	**Giant cell chronic arteritis**
Oldberg	1942	Arteritis senilis
Kilbourn & Wolff	1946	Cranial arteritis
Harrison	**1948**	**Temporal arteritis**

Giant cell arteritis

Temporal arteritis may have existed as long ago as 1350 BC (Table II). In Egyptian art, on the tomb of Pa-Aton-Em Eb dating from the 18th dynasty, a harpist is represented as follows: (i) his left eye is closed and he seems to be staring into space (he has been called "the blind harpist" by art historians); (ii) he is round shouldered, thus indicating his advanced age; (iii) interestingly, his temporal area is hollowed by a broken line strongly suggesting the temporal artery; (iv) the whole area surrounding the line is elevated and this detail is consistent with a painful inflammatory area; and finally (v) his face is emaciated. In contrast with these features, the flutist and the mandolin player appear to be young and healthy.
According to a study by Appelbloom & Van Heigem, 30% of the harpists in ancient Egypt were blind and the God of Music himself was often depicted as a blind musician (11). However, only the harpist depicted on the tomb of Pa-Aton exhibits the additional symptoms of GCA as well as an advanced age. Around 1000 AD a doctor in Mesopotamia, Alì Ibn Isa, first noted a relationship between the inflamed artery, headache and visual symptoms, and proposed a daring surgical remedy - cauterization of the artery. Dating from the Italian Renaissance, the portrait of Lorenzo Giambetti by Piero di Cosimo (1502) is even more suggestive of temporal arteritis. The tortuous and inflamed artery in this dignified elderly man is diagnostic.
The first complete clinical description of GCA (Table II) is the rightly famous one of Jonathan Hutchinson who in 1890 described " . . . a peculiar form of thrombotic arteritis of the aged" in an 80-year-old patient, the father of a porter at the London Hospital, whose superficial temporal arteries were so swollen and painful that he was unable to wear a hat (12).
In 1932 Horton *et al.* published the first biopsy description of granulomatous arteritis of the temporal vessels (13). Horton himself showed that the arteritic background was of the giant cellular type, but it took about 10 years before Gilmore, on the basis of careful anatomic studies, succeeded in indicating the presence of giant cells as the hallmark characterizing the disease (14). Since then, the name "temporal arteritis" has become synonymous with giant cell arteritis and suggests a localized affliction of the cranial arteries in the elderly. Soon the term "giant cell arteritis" took the place of the temporal arteritis.
Horton himself contributed significantly to define the clinical profile of the disease (13). He outlined the outstanding prevalence in the elderly and pointed to the pain and hyperaestesia in the temporal side and scalp, the headache, and the constitutional symptoms such as asthenia, weakness, nightly sweating, anorexia, loss of weight, and anemia. Jaw claudication and pain in the ear on opening the mouth or on chewing have been considered part of the disease ever since.
A few years later, after Gilmore, Kilbourn & Wolff drew attention to a form of vascular involvement outside the temporal artery (namely of the aortic and great extra-aortic vessels) and used the term "cranial arteritis" (15). Since then many confirmations have appeared of the aneurismatic complications at the thoracic and abdominal aorta and of their possible ominous rupture. However, Harrison in 1948 still called the disease "temporal arteritis" (16).
All of these findings have led to a different view of GCA. Temporal GCA is now thought of as a systemic disease with a number of potentially serious or even life-threatening cardiovascular complications. Temporal arteritis represents a single event in time of a phasic disorder that is generalized in terms of organ involvement (16).

Polymyalgia rheumatica and giant cell arteritis

In a work of the great Flemish painter Jan van Eyck in the municipal museum of Bruges, "The Holy Virgin with Canon

Table III. History of polymyalgia rheumatica/giant cell arteritis (PMR/GCA).

Authors	Year	Name (or observations)
Bain	1938	
Oldberg	1942	Frequency of muscle pain in GCA
Sjovall & Winblad	1944	
Porsman	1951	Rheumatic symptoms in GCA are similar to those in PMR
Paulley & Hughes	1960	Rheumatic symptoms in GCA are PMR
Hamrin	**1964**	**Arteritis in polymyalgia rheumatica**
Hamrin	1966	Polymyalgia rheumatica sive arteritica
Hamrin	1968	Polymyalgia arteritica

Van der Paele" (1436), the appearance of the Canon is striking. In the left temporal region, prominent arteries can be observed along with scar formation and the loss of hair in front of the left ear (a clear-cut case of temporal arteritis). In addition, the left hand has a significant appearance; the Canon is exerting great effort to hold his breviarium, indicating strong rheumatic pain, and the diffuse swelling of the hand indicates distal edema strongly resembling what we often observe in patients with polymyalgia rheumatica (the so-called RS3PE syndrome).

Porsman, in Barcelona in 1951 (Table III), when speaking in general terms about arthritis in old age, was the first to point out that in a series of patients studied by him, the symptoms in 29 out of 109 presumably presenting with PMR were quite similar to the "rheumatism" which he was used to observing in Horton's disease (18).

Many years before a specular feature was emphasized sporadically, i.e. that during GCA muscle pain can occur, in particular at the girdles (19, 20, 21).

In 1960 Paulley and Hughes discussed the finding that the rheumatic manifestations occurring in giant cell arteritis closely resemble those present in polymyalgia rheumatica and posed the question as to whether the latter might not represent a manifestation of the former (17). In 1964 the existence of this association was definitively accepted. The question posed by Hamrin regarding whether GCA is a rheumatic or an arteritic disease and consequently which term should be considered as more appropriate ("polymyalgia rheumatica sive arteritica") was resolved (22). At present Scandinavian clinicians deem polymyalgia rheumatica to be a manifestation of a generalized arteritis and usually adopt the term giant cell arteritis to define polymyalgia rheumatica/giant cell arteritis as a whole.

To conclude, I would like to make some simple considerations. First, we can observe (Tables I and II) the impressive fact that only two years separate the initial descriptions of the two diseases - PMR by Bruce in 1888 and GCA by Hutchinson in 1890. Secondly, if we really wish to understand the story of the history of PMR/GCA we must also note that there have been years of heated discussion about the most appropriate name for the disease, interrupted by long periods of onomastic dead calm. I believe that perhaps the time has arrived to change the name to polymyalgia.

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