SOCIEDADE BRASILEIRA DE REUMATOLOGIA

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Review article

Scientific people named in the classification of vasculitis

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ARTICLE INFO

Article history:
Received 11 September 2015
Accepted 6 May 2016
Available online 25 August 2016

Keywords:
Vasculitis
Scientific people
Eponym

ABSTRACT

The first International Chapel Hill Consensus Conference was held in 1994. There have been suggestions about the nomenclature of systemic vasculitis. Important categories were added to the classification of vasculitis, and many changes were made for disease names in the second Chapel Hill Consensus Conference 2012, which were not included in the Chapel Hill Consensus Conference 1994. The new nomenclature was introduced instead of being referred to by many names such as Churg-Strauss and Wegener"s. New categories such as Behçet"s and Cogan etc. were also added. These people are honored by the classification. They contribute to science through their case studies, scientific articles, and observations. This article reviews only eponyms present in the current classification of vasculitis. The aim of this paper is to give information about scientists mentioned in the classification of vasculitis.

Nomes de cientistas usados na classificação das vasculites

Palavras-chave:
Vasculites
Cientistas
Epônimo

RESUMO

A primeira *International Chapel Hill Consensus Conference* (CHHC) ocorreu em 1994. Fizeram-se sugestões sobre a nomenclatura das vasculites sistêmicas. Na segunda CHHC, 2012, adicionaram-se importantes categorias à classificação da vasculite e fizeram-se várias mudanças em nomes de doenças que não estavam incluídas na CHCC 1994. Introduziu-se uma nova nomenclatura em vez de se usarem nomes como Churg-Strauss e Wegener. Também foram adicionadas novas categorias, como de Behçet e Cogan etc. Essas pessoas são homenageadas pela classificação. Elas contribuíram para a ciência com seus estudos de caso, artigos científicos e observações. Este artigo analisa apenas epônimos presentes na classificação atual das vasculites. O objetivo é prestar informações sobre os cientistas mencionados na classificação das vasculites.

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http://dx.doi.org/10.1016/j.rbre.2016.06.002

Introduction

Systemic vasculitis is an inflammatory condition. The primary inflammatory process involves the vessel wall of different organs and systems, affecting blood vessels of different types and sizes. The nomenclature and classification of systemic vasculitis has been a problem for researchers and clinicians for many years.[1] There are different ways of classifying vasculitides that include the size of predominantly affected vessels, type of inflammatory infiltrate (e.g. neutrophilic, lymphocytic), etiological agent (primary or secondary), disease extension (single-organ vasculitis or systemic vasculitis) and the pathophysiological mechanism involved (e.g. immune complex deposits, ANCA). The first International Chapel Hill Consensus Conference (CHHC) was held in 1994.[2] There was an attempt to replace eponyms by noneponymous terms which would represent the pathophysiologic process. Important categories were added to the classification of vasculitis, and many names of diseases were changed at the second CHHC 2012, which were not included in the CHCC 1994.[2,3] The new nomenclature was introduced instead of being referred to by many names such as Churg-Strauss and Wegener's. The new categories were also added to the classification system, such as variable vessel vasculitis. These people are honored by the classification. They contribute to science through their case studies, scientific articles, and observations. The article reviews only eponyms present in the current classification of vasculitis. The aim of this paper is to give information about scientists mentioned in the classification of vasculitis.

Large vessel vasculitis

Takayasu arteritis

Takayasu arteritis (TA) is a chronic, idiopathic, granulomatous arteritis of the aorta and its branches. It is a form of large vessel vasculitis, and usually affects younger patients (<50 years).[2,3] The disease is also known as pulseless disease. Here, the name comes from MikitoTakayasu. He was a Japanese ophthalmologist, born in 1860. Takayasu graduated from the Tokyo Imperial University in 1887. He reported a case at the 12th Annual Meeting of the Japan Ophthalmology Society.[4,5] The patient had no abnormality in her medical examination, except for peculiar changes of the retinal central vessels with aortitis. In the patient's history, there were visual disturbances, and complete loss of visual acuity with retinal abnormalities. After presentation of this case, it was published in the *Acta of the Ophthalmic Society of Japan* in 1908.[6] Similar cases were reported consequently. It was reported that the term '*Takayasu arteritis*' was first used by Yasuzo Shinmi, and officially named as '*Takayasu arteritis*' by the researchers committee of the Department of Health and Welfare of Japan in 1975.[4,7] MikitoTakayasu died in November 1938.[4,5] Although, there are other synonyms, the disease was called as Takayasu arteritis in the nomenclature of vasculitides at the 2012 international CHCC.[3]

Medium vessel vasculitis

Kawasaki disease

Kawasaki disease is a medium-sized vessel vasculitis (visceral arteries, its branches, and especially coronary arteries), and usually occurs in young children. The disease is characterized by fever, erythematous rash, conjunctivitis, strawberry tongue, lymphadenopathy, and specific desquamations.[2,3] The disease name comes from Tomisaku Kawasaki. He was a Japanese pediatrician, born in Tokyo in 1925. Kawasaki graduated from the School of Medicine, Chiba University in 1948.[8] He described a boy aged 4 years and 3 months with high fever, mucocutaneous features, and cervical adenopathy in 1961, and presented seven cases entitle '*Non-scarlet fever desquamation syndrome*' in 1962 at the Chiba Prefecture Pediatric Meeting, and 20 cases entitle '*Twenty cases of ocular-mucocutaneous syndrome*' in 1964 at the meeting of the 15th Eastern and Central Japan Pediatric Meeting in Matsumoto.[9–11] He published a clinical observation of 50 patients in 1967 under the title "*Acute febrile mucocutaneous syndrome*".[12] In this article, patients had lymphoid involvement with specific desquamation of the fingers and toes. Later, Kawasaki et al. reported 50 cases in September 1974 at *Pediatrics* entitled '*A new infantile acute febrile mucocutaneous lymph node syndrome prevailing in Japan*'.[13] He retired from the pediatric department of Japan Red Cross Hospital.[8]

Small vessel vasculitis

Granulomatosis with polyangiitis (Wegener's)

Granulomatosis with polyangiitis (Wegener's) is an ANCA-associated multifocal necrotizing granulomatous vasculitis that affects small to medium-size vessels of the kidney, lower and upper respiratory tract.[2,3] The disease is named after Friedrich Wegener, a German pathologist, born in 1907in Varel, Germany.[14–16] He completed his medical education in 1932, studied at the pathology department of Kiel University.[16] He was reported to be a member of the Nazi party, as was half of German doctors during World War II.[17–19] In Kiel, he described a case with generalized angiitis, and necrotizing granuloma of the upper and lower respiratory system, kidney and spleen.[14] Although he worked as a pathologist in Lodz (a localized Jewish ghetto), there were conflicting reports about where he worked in the health office.[16,19] Despite the suspicion, Wegener was released due to lack of evidence as a war criminal.[16] It is reported that he was silent about the events until his death.[20] Wegener reported a peculiar rhinogenic granulomatosis with marked involvement of the arterial system and kidney, and published articles in 1936 and 1939.[21,22] In these articles, the disease's clinical and pathological features were defined. The term of '*Wegener's granulomatosis*' was first used by Godman and Churg.[23] Falk et al.recommended an alternative name for Wegener's granulomatosis: Granulomatosis with polyangiitis (Wegener's).[18] Thereafter, 'Granulomatosis with polyangiitis (Wegener's)' instead of 'Wegener's granulomatosis' was used by the CHHC

2012.[3] It was identified as Wegener's in brackets to avoid confusion in literature. He received a master clinician prize from the American College of Chest Physicians in 1989.[24] Rosen reported at the CHEST in 2007 that the board voted almost unanimously to rescind the award.[25] He died in 1990, in Lübeck.[14–16]

Eosinophilic granulomatosis with polyangiitis (Churg-Strauss)

Eosinophilic granulomatosis with polyangiitis (Churg-Strauss) is ANCA-associated necrotizing granulomatous vasculitis that affects predominantly small to medium-size vessels. It is generally associated with asthma, paranasal sinusitis, pulmonary infiltrates, neuropathy, and eosinophilia of peripheral blood or tissue.[2,3] The disease is named after Jacob Churg and Lotte Strauss. Jacob Churg was a pathologist, born in 1910, Dolhinow.[26,27] He was interested in medicine since the age of eight, worked in the department of internal medicine, but was reported as saying 'I was not very successful with patients'.[27] He then worked as an assistant in the pathology department of Vilna University. He described a patient with asthma, lymphadenopathy, and eosinophilia, who died of cranial hemorrhage.[27] The biopsy specimens of lymph node showed eosinophilic infiltration, early granulomas, vasculitis in cranial arteries, and granulomas in various tissues. Dr. Strauss also had similar cases.[26] Lotte Strauss was a pathologist, born in 1913in Nuremberg, Germany.[26] She specialized in pediatric and perinatal pathology. They, both reviewed cases with asthma, fever, and hypereosinophilia. Most of them had characteristic, specific anatomical lesions with histopathologic entity termed '*allergic granuloma*'. Their article was published in 1951 entitle '*Allergic Granulomatosis*, *Allergic Angiitis*, and *PeriarteritisNodosa*'.[28] Churg and Strauss described the entity and called it the Churg-Strauss syndrome. When searching the term 'Churg-Strauss syndrome' on PubMed, we found an early article reported by Abul-Haj Sk and Flanagan P, in 1961 entitled '*Asthma associated with disseminated necrotizing granulomatous vasculitis, the Churg-Strauss syndrome. Report of a case*'.[29] Jacob Churg died in 2005, and Lotte Strauss in 1985.[26]

IgA vasculitis (Henoch-Schönlein)

IgA vasculitis (Henoch-Schönlein) generally affects small vessels with immunoglobulin A deposits and granulocytes in the vessels. The clinical features include palpable purpura, arthritis, abdominal pain, gastrointestinal bleeding, and glomerulonephritis.[2,3] The disease is named after Eduard Heinrich Henoch and Johann Lukas Schönlein. Eduard Heinrich Henoch was a German pediatrician, born in Berlin on July 16, 1820, and died in Dresden on August 28, 1910.[30,31] He completed his medical education in 1843, and studied with Moritz Romberg and Johann Lukas Schönlein. He worked as head of the pediatric clinic of the Royal Charité Hospital. He published his famous textbook called '*Lectures on Children's Diseases*'.[32] On the other hand, he reported a fifteen-year-old-male patient with abdominal pain, arthralgia, purpura; a seven-year-old male with purpura, nephritis, and a patient with hematuria and purpura. This article was published in 1868 in the '*KlinWschr*'.[33] These findings were also described by Johann Lukas Schönlein as an entity. Schönlein was a German Professor of Internal Medicine, born in Bamberg, Germany on 30 November 1793.[34,35] He worked in Würzburg, Zürich, and Berlin. Schönlein wrote few papers, and described rheumatica purpura rubra with cutaneous, arthritic symptoms and renal lesions.[36,37] He also described typhoid crystals in patients' stools, and the causative agent of favus. Johann Lukas Schönlein retired in 1859 and died in Bamberg in January 1864.[34–36]

Variable vessel vasculitis

Cogan's syndrome

Cogan's syndrome is a rare chronic inflammatory disease characterized by large to small-sized vessel vasculitis, nonsyphilitic interstitial keratitis and vestibulo-auditory dysfunction such as tinnitus, hearing loss, and vertigo.[3] The disease is named after David Glendenning Cogan. He was an American ophthalmologist, born in Massachusetts in 1908.[38] He was graduated from the Dartmouth College, Harvard University, and continued his career at Harvard Medical School, Chicago University Clinics, and Massachusetts Eye and Ear Infirmary. Cogan was certified by the American Board of Ophthalmology in 1937, and became a member of the editorial board at Archives of Ophthalmology, Investigative Ophthalmology, Albrecht von Graefes Archiv fur Klinische und Experiementelle Ophthalmologie, and Journal of Neurological Science.[38] In 1945, he reported five patients with nonsyphilitic interstitial keratitis and vestibulo-auditory symptoms as a syndrome at the *Archives of Ophthalmology*.[39] Hence, this clinical entity has come to be called as Cogan's syndrome. He was interested in neuro-ophthalmology, and ophthalmic manifestations of systemic vascular disease, and he published many books on these topics.[40,41] He died in Wayne after a heart attack on September 9, 1993.[42]

Behçet's disease

Behçet's disease is a chronic, inflammatory, multisystem vasculitis and is characterized by recurrent oral ulcers, genital ulcers, and uveitis.[3] The disease generally affect ethnic groups such Mediterranean and East Asian along the Silk Road. Hulusi Behçet was a Turkish dermatologist, the first professor in the Turkish academic life, born in Istanbul, Turkey on February 20, 1889.[43,44] He completed his medical education, and graduated from Military Medical School in 1910, then worked as an assistant in the Gulhane dermatology clinic until 1914. In July 1914, he was appointed as Chief Assistant at the Kırklareli Military Hospital, and worked as a dermatologist at the Edirne Military Hospital. He also worked at Charité Hospital, Hasköy Dermatology and Venereal Diseases Hospital, and Guraba Hospital.[45] He was interested in syphilis, leishmaniasis, and mycosis. Hulusi Behçet described two cases with recurrent oral and genital aphthous ulcers and uveitis with hypopyon. He published the findings and his opinions in 1937 in *Dermatologische Wochenschrift* and with more details in the same journal in 1938.[46] The disease was named as '*Morbus Behçet*' by professor

Mischner in 1947 at the International Medical Congress of Geneva.[45] The disease is encoded as Behçet's disease (M35.2), at ICD-10 Version:2015.[47] He was honored with the TÜBİTAK Science Award in 1975, and Eczacıbaşı Science Award in 1982.[45] Coins were also produced for his tribute during the National Dermatology Congress in 1996. Hulusi Behçet died in March 8, 1948.[43–45]

Conclusion

Although the named vasculitis in this article had been previously described by other valuable scientists, these eponyms are commonly used in the nomenclature of classification of vasculitis after scientists have honored them. The American College of Rheumatology, the European League Against Rheumatism, and the American Society of Nephrology recommend diagnostic terms instead of eponyms such as Churg-Strauss, Wegener's etc. Many new nomenclatures, and diagnostic terms explain clinico-pathological information about the disease. As a result, these people who have contributed to science emphasize about the significance of clinical observation, approach and research.

Conflicts of interest

The authors declare no conflicts of interest.

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