Eosinophilic granulomatosis with polyangiitis guidelines

Sharon Chang, MD The first-ever ACR guide developed to manage granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic polyangyitis granulomatosis (EGPA) provides useful recommendations for managing these complex conditions in clinical practice. Sunday Session, ANCA-Associated Vasculitis: How to apply the new ACR guidelines, from 8:30 a.m. to 10:00 a.m. in the Thomas Murphy Ballroom 3 and 4, Building B at the Georgia World Congress Center, will discuss the draft of this new guide. Sharon Chang, MD, principal investigator and head of the core oversight team that developed the guidance, will review the new developed guidelines for the GPA and MPA and discuss its clinical application. Dr. Chang is an associate professor of medicine in the Department of Rheumatology and Director of the Vasculit Clinic at the University of California, San Francisco. Joining Dr. Chang will be a principal member of the Carol Langford Oversight Group, MD, MHS, who will review the guidelines for EGPA. Dr. Langford is director of the Vasculitis and Research Center at the Cleveland Clinic. Systemic vasculitides are a group of diseases that are rare, even for common rheumatologists, and can have organoth-threatening and life-threatening manifestations, said Dr. Chung. Given their many manifestations, potential mimiers, and now increasing treatment options, diagnosing, treating and treating these diseases can be challenging. Rheumatologists who don't treat the disease regularly, she said, often seek advice from colleagues and experts to develop treatment and management plans for their patients. The new guidelines apply to seven classic systemic vasculitides - giant cellular arteritis (GCA), arteritis Takayas (TAK), polyarteritis nodose (PAN), Kawasaki disease, and three ANCA-related vasculitides. For ANCA-related vasculitides, we will provide recommendations regarding the use of remission induction and supportive therapy, management of severe and non-severe illness, and laboratory studies to monitor diseases and guide treatments, said Dr. Chung. For large vasculitis vessels, the guidelines include recommendations regarding the use of imaging to diagnose and monitor patients, the use of glucocorticoid sparing therapy, and surgical interventions for management. The two sessions on Monday will cover new guidelines for GCA, TAK, PAN and Kawasaki disease. GCA, TAK and PAN: How to apply the new ACR guidelines (see Tuesday's story ACR Daily News) will take place from 8:30 - 10:00 a.m. in Hall B1, and the treatment of Kawasaki disease: Old and New in 2019 (see story on page A15) will take place from 11:00 a.m. - 12:00 p.m. in room B216-B217. During sessions in Dr. Chang said the faculty would present treatment and management recommendations scenarios where rheumatologists consult on Kawasaki disease, as well as recommendations for the treatment of polyarteritis nodosa, including the use of imaging and biopsy studies for diagnosis. This is ACR's first attempt to develop guidelines for the treatment of these complex diseases, Dr. Chung said. At the end of these sessions, we hope that participants will not only have a clear understanding of the basic principles regarding the treatment of these diseases, but also recognize that there are nuances relevant to each patient that will affect each patient's treatment plan. The new guidelines, developed in partnership with the Vasculita Foundation, are expected to be finalized, approved and published in spring 2020. Nguyen Y, Guillain L. Eosinophilic Granulomatosis with Polyangiitis (Hurg-Strauss). Semin Respir Crit Care Med. 2018 Aug 39 (4):471-481. (Medline). (Guide) Groh M, Pagnoux C, Baldini C, Bel E, Bottero P, Cottin V et al. Eosinophil granulomatosis with polyangiitis (Hurg-Strauss) (EGPA) Consensus Task Force recommendations for evaluation and management. Eur J Intern Med. 2015 Sep. 26 (7):545-53. (Medline). (Full text). Guillevin L, Cohen P, Gayraud M, et al. Hurg-Strauss Syndrome. Clinical study and long-term follow-up training of 96 patients. Medicine (Baltimore). 1999 January 78 (1):26-37. (Medline). Perhg J, Strauss L. Allergic granulomatosis, allergic angiitis, and periarteritis nodoza. Am J Pathol. 1951 Mar-April. 27(2):277-301. (Medline). Hurg A. Pulmonary angiitis and granulomatosis again. Hum Pathol. 1983 Oct. 14(10):868-83. (Medline). Keogh KA, Sfox W. Hurg-Strauss syndrome: clinical presentation, cytoplasmic antibodies of antineutrophil and antagonists of leukotriene receptors. Am J Med. 2003 Sep. 115(4):284-90. (Medline). Masi AT, Hunder GG, Lie JT et al. American College of Rheumatology 1990 criteria for classification of Hurg-Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Reum. 1990 Aug. 33(8):1094-100. (Medline). Jennett JC, Falk RJ, Andrsassi K, et al. Nomenclature of systemic vasculitides. Proposal for an international consensus conference. Arthritis Reum. 1994 February 37(2):187-92. (Medline). Cottin V, et al.; Groupe d'Etudes et de Recherche sur les Maladies Orphelines Pulmonaires (GERMOP). Review of systemic vasculitis in eosinophil granulomatosis with polyangiitis (Hurg-Strauss): Study of 157 patients Groupe d'Etudes et de Recherche sur les Maladies Orphelines Pulmonaires and the Task Force of the European Respiratory Society for eosinophilic granulatosis with polyangite (Hurg-Strauss). Autoimmun Rev. 2017 January 16 (1):1-9. (Medline). Helmich B, Ehlers S, Chernok E, Gross VL. Update on the pathogenesis of Hurg-Strauss syndrome. Wedge Ext Revmatol. 2003 November-December. 21 (6 Suppl 32):S69-77. (Medline). Franco J, Artes MJ. Pulmonary eosinophilia associated with Chest. 1999 June 54 (6):558-60. (Medline). Wechsler ME, Finn D, Gunawardena D, et al. Hurg-Strauss Syndrome in patients receiving montelucast as asthma treatment. Breast. 2000 Mar. 117(3):708-13. (Medline). Hauser T, Mahr A, Metzler C, Coste J, Sommerstein R, Gross WL, et al. Antagonist of leukotriene receptors Montelukast and the risk of Hurg-Strauss syndrome: crossover study. Chest. 2008 Aug. 63(8):677-82. (Medline). Wechsler ME, Harpestad E, Flier SR, et al. Pulmonary infiltrators, eosinophilia, and cardiomyopathy after withdrawal of corticosteroids in patients with asthma obtain zafirlukast. Jama. 1998 Feb 11. 279(6):455-7. (Medline). Tuggay JM, Hosker HS. Hurg-Strauss syndrome associated with montelukastic therapy. Chest. 2000 55 (9): 805-6. (Medline). Vaglia A, Martorana D, Maggiore W, Grasselli S, Dzanetti A, Pesci A, et al. HLA-DRB4 as a genetic risk factor for Hurg-Strauss syndrome. Arthritis Reum. 2007 56(9): 3159-66. (Medline). Eustace JA, Nadasdy T, Choi M. Illness of the Month. Hurg Strauss Syndrome. J Am Soc Nephrol. 1999 September 10 (9):2048-55. (Medline). Hasley PB, Follansbee WP, Kulehan JL. Heart manifestations of Hurg-Strauss syndrome: case report and literature review. Am Heart J. 1990 Oct 120 (4):996-9. (Medline). Keogh KA, Spex W. Hurg-Strauss Syndrome: An update on clinical, laboratory and therapeutic aspects. Sarcoidosis Vasc diffuse lung Dis. 2006 Mar 23(1):3-12. (Medline). Boyer D, Vargas SO, Slattery D, Rivera-Sanchez YM, Colin AA. Hurg-Strauss syndrome in children: clinical and pathological review. Pediatrics. 2006 118 (3): e914-20. (Medline). Wechsler ME, Finn D, Gunawardena D, Westlake R, Barker A, Haranath SP, et al. Hurg-Strauss Syndrome in patients receiving montelukast as asthma treatment. Breast. 2000 Mar. 117(3):708-13. (Medline). Sinico RA, Sabadini E, Maresca AM. Hurg-Strauss syndrome, caused by mesalazin, in a patient with Crohn's disease and sclerosing cholangita. Wedge Ext Revmatol. 2006 Mar-Apr. 24 (2 Suppl 41):S104. (Medline). Orriols R, Munoz X, Ferrer J, Huget P, Morell F. Cocaine induced by Hurg-Strauss vasculitis. Eur Respir J. 1996 Jan. 9(1):175-7. (Medline). Sokolovsky BM, Sheklik VK, Vludarczyk A.A., Kuchiya PP, Yakiela BA, Gazior J. et al. ANCA-positive and ANKA-negative phenotypes of eosinophil granulomas with polyangitiit (EGPA): the result and long-term follow-up of 50 patients from one Polish center. Wedge Ext Revmatol. 2014 May-June. 32 (3 Suppl 82):S41-7. (Medline). Sabre-Furtasso R, Cohen., Mahr A, Panu S, Roton L, Jane D. et al. Antineurthril cytoplasmic antibodies and Hurg-Strauss syndrome. Anne Intern Med. 2005 November 1. 143(9):632-8. (Medline). Choi YH, Im JG, Han BK, Kim JH, Lee KY, Myoung NH. Thoracic manifestations of Hurg-Strauss syndrome: radiological and clinical results. Breast. 2000 January 117 (1):117-24. (Medline). Kim Y, KS, Choi DC, et al. Spectrum of Eosinophil lung disease: radiological findings. J Comput Help Tomogr. 1997 November-December. 21(6):920-30. (Medline). Bushman DL, Waldron JA Jr., King TE junior Hurg-Strauss pulmonary vasculitis. High-resolution CT scan and pathological results. Am Rev Respir Dis. 1990 Aug. 142(2):458-61. (Medline). Worthy SA, Muller NL, Hansell DM, Flower CD. Hurg-Strauss syndrome: a range of pulmonary CT results in 17 patients. AJR Am J X Roentsol. 1998 February 170 (2):297-300. (Medline). Mavrogeni S, Carabela G, Gialafos E, Stavropoulos E, Spiliotis G, Katsifis G, et al. Cardiac Participation in ANCA Syndrome (C) and ANCA (-) Hurl-Strauss is rated cardiovascular magnetic resonance. The allergy of the drug targets is ignited. 2013 Oct 12(5):322-7. (Medline). JT's lies. Histopathological specificity of systemic vasculitis. Reum Dees Wedge North Am. 1995 November 21(4):883-909. (Medline). Katzenstein AL. Diagnostic features and differential diagnosis of Hurg-Strauss syndrome in the lungs. Review. Am J Wedge Pathol. 2000 November 114 (5):767-72. (Medline). Chemmalakuzhy AJ, Chou XJ, Hedayati SS. ANCA-negative glomerulonephritis associated with neastmatic Hurg-Strauss syndrome. Nat Klin Pr. Nephrol. 2008 Oct 4 (10):568-74. (Medline). Lhot F,Coen, Gilpain., Guilleun L. (Hurg-Strauss syndrome). Reverend Prat. 2008 June 15. 58(11):1165-74. (Medline). Hervier B, Durant C, Masso A, Ponge T, Hamidou M, Mussini JM. Use of muscle biopsy to diagnose systemic vasculitides. J Revmatol. 2011 March 38(3):470-4. (Medline). Grau W. Hurg-Strauss Syndrome: Update 2005-2008. Curr Rheumatol Republic 2008 Dec 10 (6):453-8. (Medline). Muhtyar C, Flossmann O, Hellmich B, Bacon P, Sid M, Cohen-Tervaert JW, et al. Results of studies of antineutropyl cytoplasm antibodies associated vasculitis: a systematic review of the European League against Rheumatism systemic vasculitis task force. Ann Reum Dis. 2008 July 67 (7):1004-10. (Medline). Mohammad AJ, Hot A, Arndt F, Moosig F, Gerry MJ, Amudala N, et al. Rituximab for the treatment of eosinophilic granulomatosis polyangiitis (Hurg-Strauss). Ann Reum Dis. 2016 February 75 (2):396-401. (Medline). Jachiet M, Samson M, Cottin V, Kang JE, Le Guenno G, et al. Anti-IgE monoclonal antibodies (omalizumab) in fireproof and recurrence of eosinophil granulomatosis with polyangiitis (Hurg-Strauss): Data from 17 patients. Arthritis Revmatol. 2016 March 4. (Medline). Tiliakos 4th, Shaya S, Hostoffer R, Kent L. Use of infliximab in a patient with steroid-dependent persimmon syndrome persimmons. J Wedge Revmatol. 2004 Apr 10(2):96-7. (Medline). Casian A, Jane D. Plasma exchange in the treatment of wegener's granulomatosis, microscopic polyangiitis, Hurg-Strauss syndrome and renal limited vasculitis. Carr Opin Revmatol. 2011 January 23 (1):12-7. (Medline). Guillauvin L, Cevallos R, Durand-Gasselin B. and Treatment Treatments globeroneprit in microscopic polyangyitis and Hurg-Strauss syndrome. Plasma exchanges, meta-analysis of 2 randomized trials for 140 patients, 32 with glomerulonephritis. Anne Med Intern (Paris). 1997. 148(3):198-204. (Medline). Bonaci-Nikolic B, Nikolic MM, Andrejevic S, S, Bukilich M. Antineutrophil cytoplasmic antibodies (ANCA)-associated autoimmune diseases caused by antithyroid drugs: comparison with idiopathic ANCA vasculitides. Arthritis Res Ther. 2005. 7 (5):R1072-81. (Medline). Wechsler ME, Akuthota P, Jane D, Khoury P, Clyon A, Langford CA, et al. Mepolizumab or Placebo for eosinophil granulomatosis with polyangiitis. N Engl J Med. 2017 May 18. 376 (20):1921-1932. (Medline). Cohen., Guillevin L. , Baril L. et al. Preservation of cytoplasmic antibodies of antineutrophil (ANCA) in patients with systemic polyarteritis nodose or Hurg-Strauss syndrome: the subsequent development of 53 patients. Wedge Ext Revmatol. 1995 March-April. 13(2):193-8. (Medline). Espinosa LR. Combination therapy for the treatment of persimmon-strauss syndrome: corticosteroids with short- or long-term impulses of cyclophospamid. Curr Rheumatol Republic 2008 Dec 10 (6):427-9. (Medline). Yamasaki A, Tomita K, Fujii J, Hasegawa Y, Watanabe M, Sano H. et al. Repressed aneurysms of the silt artery in Hurg-Strauss syndrome after combined treatment with glucocorticoid and cyclophosphamide. Revmatol Int. 2008 Aug 8. (Medline). Ribi C, Cohen P, Pagnoux C, Mahr A, Arne JP, Lauque D, et al. Treatment of Hurg-Strauss syndrome without factors of poor prognosis: multicenter, promising, randomized, open trial of 72 patients. Arthritis Reum. 2008 February 58 (2):586-94. (Medline). Tatsis E, Schnabel A, Gross WL. Interferon-alpha treated four patients with Hurg-Strauss syndrome. Anne Intern Med. 1998 Sep 1. 129(5):370-4. (Medline). Metzler C, Schnabel A, Gross WL, Helmich B. Phase II study of interferon-alpha for the treatment of fireproof Hurg-Strauss syndrome. Wedge Ext Revmatol. 2008 May-June. 26(3 Suppl 49):S35-40. (Medline). Beasley R, Bibby S, Weatherall M. Leukotriene receptor antagonist therapy and Hurga-Strauss syndrome: culprit or innocent passerby?. Chest. 2008 Oct 63 (10):847-9. (Medline). Giavina-Bianchi. Three months of anti-IgE administration to a patient with Hurg-Strauss syndrome. J Allergy Wedge Immuno. 2007 May. 119(5):1279; 1279-80. (Medline). Green RL, Vayonis AG. Hurg-Strauss syndrome after zafirlukast in two patients who do not receive systemic steroid treatment. Lancet. 1999 Feb 27. 353(9154):725-6. (Medline). Guillain L, Lhot F, Gayro M, etc. Predictive Factors in Polyarteritis Nodoze syndrome and Hurl-Strauss. Promising study in 342 patients. Medicine (Baltimore). 1996 January 75 (1):17-28. (Medline). Kaushik VV, Reddy HV, Bucknell RC. Successful rituximab in the patient's patient hurg-Strauss syndrome. Ann Reum Dis. 2006 Aug 65 (8):1116-7. (Medline). Khopers,, Erdbrugger W, Grossheim M, Meyer GP, Hissa M, Gwinner W, et al. Endothelial microparticles as diagnostic aid for cardiomyopathy caused by Hurg-Strauss vasculitis. Wedge Ext Revmatol. 2008 May-June. 26(3 Suppl 49):S86-9. (Medline). Lie JT. Illustrated histopathological classification criteria for selected vasculitis syndromes. Subcommittee of the American College of Rheumatology on vasculitis classification. Arthritis Reum. 1990 Aug. 33(8):1074-87. (Medline). Pabst S, Tiyerili V, Grohe C. Apparently the answer to anti-IgE therapy in two patients with fireproof forme fruste Hurg-Strauss syndrome. Chest. 2008 Aug. 63(8):747-8. (Medline). Roccatello D, Baldovino S, Alpa M, Rossi D, Napoli F, Naretto C, et al. Effects of anti-CD20 monoclonal antibodies as a life-saving treatment of ANCA-related idiopathic systemic vasculitis with or without excessive kidney involvement. Wedge Ext Revmatol. 2008 May-June. 26(3 Suppl 49):S67-71. (Medline). Todd DC, Cockcroft DW. Long-term survival in Hurg-Strauss syndrome. Anne Allergy Asthma Immuno. 2004 January 92 (1):92-3. (Medline). Greco A, Rizzo MI, De Virgilzio A, Gallo A, Fusconi M, Ruoppolo G, et al. Hurg-Strauss Syndrome. Autoimmun Rev. 2015 Apr 14 (4):341-8. (Medline). Wechsler ME, Akuthota P, Jane D, Khoury P, Clyon A, Langford CA, et al. Mepolizumab or Placebo for eosinophil granulomatosis with polyangiitis. N Engl J Med. 2017 May 18. 376 (20):1921-1932. (Medline). Wechsler ME, Akuthota P, Jane D, Khoury P, Clyon A, Langford CA, et al. Mepolizumab or Placebo for eosinophil granulomatosis with polyangiitis. N Engl J Med. 2017 May 18. 376 (20):1921-1932. (Medline). (Full text). Text. eosinophilic granulomatosis with polyangiitis treatment guidelines