

Teaching NeuroImages: Granulomatosis with polyangiitis causing Collet-Sicard syndrome and refractory headache

Nathaniel M. Schuster, MD
Stellios Karnezis, MD
Lucas Restrepo, MD, PhD

Correspondence to
Dr. Schuster:
nschuster@gmail.com

Figure 1 Clinical examination before immunosuppression



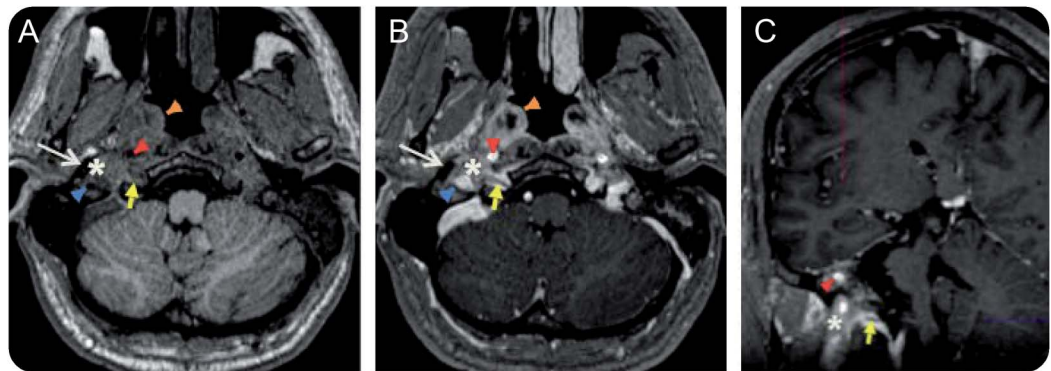
Examination revealed multiple cranial neuropathies as evidenced by palatal asymmetry (A), right trapezius atrophy (B) with scapular winging (C), and rightward tongue deviation (A).

A 52-year-old man presented for 4 months of progressive hypophonia and dysphagia with refractory headache and bilateral otitis media. Examination and MRI revealed abnormalities (figures 1 and 2). Serum proteinase 3 antibody level was 180 units (normal <21 units) and c-ANCA (antineutrophil cytoplasmic antibody) was 1:160, consistent with granulomatosis with polyangiitis (GPA;

previously known as Wegener granulomatosis). Lung nodule biopsy confirmed GPA.

Cranial neuropathies are a recognized complication of GPA.^{1,2} Collet-Sicard syndrome represents cranial nerve 9–12 palsies with sympathetic sparing. Collet-Sicard syndrome can be due to intracranial or extracranial pathology; differential diagnosis includes neoplasm (such as carotid

Figure 2 MRI brain retrostyloid parapharyngeal space heterogeneously enhancing material



(A) Axial T1 precontrast; (B) axial T1 postcontrast; and (C) sagittal oblique T1 postcontrast abnormally enhancing material (asterisk) between internal carotid/carotid canal (red arrowhead) and jugular vein/jugular foramen (blue arrowhead) extending to styloid process (white arrow) and nasopharyngeal mucosa/torus tubarius (orange arrowhead). Hypoglossal nerve (yellow arrow) is obscured past the hypoglossal canal.

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From the Montefiore Headache Center (N.M.S.), Department of Neurology, Montefiore Medical Center, Albert Einstein College of Medicine, Bronx, NY; and Departments of Radiology (S.K.) and Neurology (N.M.S., L.R.), Ronald Reagan–UCLA Medical Center, Los Angeles, CA. Go to Neurology.org for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

body paraganglioma), infection, and inflammatory disorders.

AUTHOR CONTRIBUTIONS

Nathaniel M. Schuster, MD, participated in design and conceptualization of the study, analysis and interpretation of the data, and drafting of the manuscript. Stelios Karnezis, MD, participated in analysis and interpretation of the data and drafting of the manuscript. Lucas Restrepo, MD, PhD, participated in design and conceptualization of the study, analysis and interpretation of the data, and revising the manuscript for intellectual content.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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