Giant Cell Arteritis

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Overview

- Giant cell arteritis (GCA), also referred to as temporal arteritis, is a medium to large-size vessel vasculitis that usually affects the temporal or occipital branches of the external carotid artery, but can also involve the ophthalmic, vertebral, distal subclavian, and axillary arteries, as well as the thoracic aorta
- Ischemic optic neuropathy is a medical emergency and early steroid treatment is essential to avoid permanent visual loss; a biopsy should be performed but treatment must be prioritized
  - Other complications include scalp necrosis, cerebrovascular events, and aortic dissection and aneurysm
- Can occur concurrently with polymyalgia rheumatica (PMR) in up to 50% of patients
- Most commonly seen in white females over the age of 50, with peak incidence in the seventh and eighth decades

Clinical Presentation

- Classically presents as a unilateral headache unresponsive to analgesics that is associated with exquisite tenderness overlying the temporal or occipital arteries; affected vessel can exhibit decreased or absent pulsation
- Fever, malaise, and jaw claudication are common symptoms
- When a patient has concurrent PMR, symmetric proximal muscle stiffness and arthralgias are also seen
- Cutaneous manifestations are not always present, but the affected vessel can be tender, hard, or tortuous under inflamed, cyanotic, or necrotic skin (Fig. 52.1)
  - Vesicles or bullae may occur and later may evolve into scalp necrosis and gangrene
  - Scalp necrosis is a rare complication that may be associated with optic neuropathy and other severe complications
  - Tender nodules overlying the affected artery or scalp, prurigo nodularis, lingual artery involvement with tongue necrosis, ecchymoses, retiform purpura, urticaria, alopecia, and actinic granuloma have been described

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- Visual disturbances include partial or total vision loss and diplopia
  - Vision loss is reported in 10–20% of patients, typically due to anterior ischemic optic neuropathy or less frequently from retinal artery occlusion, which can be irreversible even with treatment
- Other rare and severe complications include extremity claudication, cerebrovascular events, aortic aneurysms and dissections, vascular stenosis, and myocardial infarctions

Histopathology

- A primarily lymphohistiocytic inflammatory infiltrate of the transmural arterial wall is present with interruption of the internal elastic lamina (Fig. 52.2)
  - Multinucleated giant cells may be seen within the inner media, but are not required for diagnosis
  - Elastica van Gieson stain can help highlight elastic lamina fragmentation
- Serial sectioning is helpful as the lesions can be focal, and some patients require bilateral temporal artery biopsies to demonstrate the inflammatory infiltrate

Differential Diagnosis

- ANCA-associated vasculitis, notably granulomatosis with polyangiitis: can often be distinguished by history and physical exam, supplemented with laboratory testing
- Rarely, systemic amyloidosis can present with temporal artery involvement: may have other signs of amyloidosis (pinch purpura, macroglossia), and different histology
- Takayasu's arteritis: can be indistinguishable on histology and imaging, but age of onset and distribution of lesions should preclude this diagnosis

Work-Up

- Temporal artery biopsy is the diagnostic gold standard; however, biopsy can be negative given the focal nature of the arteritis or in those with predominantly subclavian arterial involvement
  - Ideal biopsies consist of a segment at least measuring at least 2 cm
  - Treatment should not be delayed for biopsy if the diagnosis is strongly suspected
- Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are elevated in almost all patients
  - An ESR >50 mm/hr has a sensitivity of 84–86%, albeit low specificity, and is a part of the American College of Rheumatology's diagnostic criteria for GCA
  - Additional criteria include: age $\geq$ 50, new headache, temporal artery symptoms including tenderness and decreased pulsation, and consistent biopsy findings
- As a large vessel vasculitis can occur in up to 25% of patients with GCA, magnetic resonance angiography (MRA) or computed tomographic angiography (CTA) of the aortic arch should be considered
  - These imaging modalities may also be useful in patients in whom GCA is suspected, but not confirmed on biopsy
- Fundoscopy should be performed in patients with visual disturbances in whom GCA is a concern

Treatment

- Systemic corticosteroid monotherapy is effective in the majority of patients
  - Treatment is initiated with prednisone
  - A taper of corticosteroids is usually begun once clinical symptoms and inflammatory markers have normalized with long-term treatment often required
- As ischemic optic neuropathy is a medical emergency, intravenous methylprednisolone should be initiated promptly in patients in whom there is concern for ischemic complications, even if it delays diagnostic biopsy
- The use of steroid-sparing agents has not shown to produce the therapeutic efficacy of steroid monotherapy

Suggested Readings

1. Weyand CM, Goronzy JJ. Clinical Practice giant cell arteritis and polymyalgia rheumatica. N Engl J Med. 2014;371:50–7.
2. Salvarani C, Pipitone N, Versari A, Hunder GG. Clinical features of polymyalgia rheumatica and giant cell arteritis. Nat Rev Rheumatol. 2012;8:509–21.

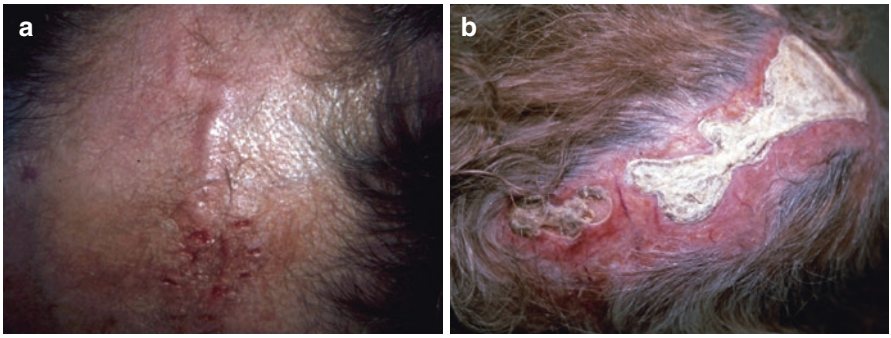

Fig. 52.1 Giant Cell Arteritis: (**a**) Hard, tender, pulsatile temporal artery characteristic of giant cell arteritis. (**b**) Necrosis can result from scalp infarction in giant cell arteritis (Courtesy of Natalie Wright MD & Sam Moschella MD).

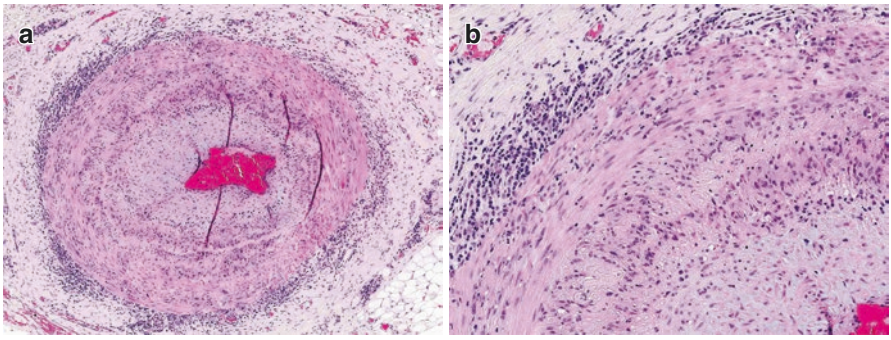

Fig. 52.2 Giant cell arteritis *(10x, 20x; H&E)*: (**a**) Temporal artery demonstrates marked intimal thickening and transmural inflammation with a dense inflammatory infiltrate composed of lymphocytes and histiocytes. (**b**) Multinucleated giant cells also may be seen transversing the arterial wall (Courtesy of Natalie Wright MD & Sam Moschella MD).