A real pain in the neck: Giant cell arteritis presenting with non-necrotising fasciitis and fever

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Word count: 499

Key words: Giant cell arteritis; non-necrotising fasciitis.

Giant cell arteritis (GCA) is the most common primary vasculitis in adults aged over 50 years affecting women more frequently¹. We present a case in which non-necrotizing fasciitis manifesting as neck pain and fever were the presenting features.

A 95 year old woman with a history of macular degeneration presented with fever, 3 weeks of acute neck pain, headache, night sweats and myalgias. Oral intake was reduced secondary to a “tired jaw”. Examination revealed prominent but non-tender pulsatile temporal arteries. Fundoscopy revealed a right swollen optic disc. Investigations were notable for a raised ESR 80mm/h, CRP 220mg/L, white cell count ($20 \times 10^9/L$), platelet count and ferritin and hypoalbuminemia.

MRI of the neck showed diffuse enhancement of the posterior fascial planes consistent with fasciitis. No gas locules were identified (Figure 1). There were no obvious abnormalities of the aortic arch or great vessels. Blood cultures were negative. Bilateral temporal artery biopsy confirmed the presence of GCA.

This is the author manuscript accepted for publication and has undergone full peer review but has not been through the copyediting, typesetting, pagination and proofreading process, which may lead to differences between this version and the Version of Record. Please cite this article as doi: [10.1111/imj.14317](https://doi.org/10.1111/imj.14317)

Within one month of treatment with oral prednisolone 40mg daily her symptoms had resolved and acute phase reactants had normalised. Due to neuropsychiatric side effects from prednisolone subcutaneous weekly tocilizumab 162mg/0.9mL was commenced. She remains well living independently and right eye fundoscopy is normal eight months after diagnosis.

GCA typically affects the extracranial branches of the external carotid artery and presents with a wide range of clinical manifestations of which headache is most common². Polymyalgia rheumatic (PMR) can be present in up to 50% of cases¹. Although neck pain is common in PMR¹, the striking feature in this patient is non-necrotizing fasciitis on MRI. Necrotizing fasciitis due to a bacterial aetiology is a medical emergency and the presence of gas along fascial planes is pathognomonic. Non-necrotizing fasciitis has previously been described associated with systemic inflammatory disorders such as systemic lupus erythematosus, Churg-Strauss vasculitis and dermatomyositis³. We believe this is the first published description of non-necrotizing fasciitis associated with GCA, likely a manifestation of concurrent PMR. While synovitis, bursitis and tenosynovitis have all been observed in PMR, a single report has documented the presence of fasciitis in the deltoid occurring with subdeltoid bursitis⁴.

Severe ischaemic manifestations of GCA occur not infrequently with jaw claudication in 41% of patients and permanent visual loss in 12.9%⁵. An abnormal temporal artery on examination², and the presence of traditional atherosclerotic risk factors⁵, increase the risk of ischemic complications.

Temporal artery biopsy has variable sensitivity (39% - 90%), according to the predominant artery involved, timing in relation to commencement of treatment⁶ and inconsistent gold standards. While advancements in imaging technologies have aided in the diagnosis of GCA (reviewed in⁶), we believed it prudent to perform temporal artery biopsy to confirm the diagnosis in this patient before committing her to high dose glucocorticoids.

This case highlights an unusual presentation of GCA in a patient with clinical features and biomarkers suggestive of GCA and PMR. This case adds to the list of systemic inflammatory conditions that need to be considered in the aetiology of non-necrotizing fasciitis.

Figure legend:

FIGURE 1. Axial (A) and sagittal (B) fat saturated, contrast enhanced T1 weighted images, demonstrate marked enhancement of the posterior neck, deep cervical fascia (arrows). This corresponds with fascial oedema on sagittal T2 fat saturated images (asterisks). Axial CT images demonstrate no gas within the fascia on soft tissue (D) or lung windows (E).

References:

1. Salvarani C, Cantini F, Hunder GG. Polymyalgia rheumatica and giant-cell arteritis. *Lancet*. 2008;372(9634): 234-245.
2. Gonzalez-Gay MA, Barros S, Lopez-Diaz MJ, Garcia-Porrúa C, Sanchez-Andrade A, Llorca J. Giant cell arteritis: disease patterns of clinical presentation in a series of 240 patients. *Medicine (Baltimore)*. 2005;84(5): 269-276.
3. Chaudhry AA, Baker KS, Gould ES, Gupta R. Necrotizing fasciitis and its mimics: what radiologists need to know. *AJR Am J Roentgenol*. 2015;204(1): 128-139.
4. Rozin AP. US imaging of shoulder fasciitis due to polymyalgia rheumatica. *Neth J Med*. 2008;66(2): 88.
5. Gonzalez-Gay MA, Pineiro A, Gomez-Gigirey A, et al. Influence of traditional risk factors of atherosclerosis in the development of severe ischemic complications in giant cell arteritis. *Medicine (Baltimore)*. 2004;83(6): 342-347.
6. Ninan JV, Lester S, Hill CL. Giant cell arteritis: beyond temporal artery biopsy and steroids. *Intern Med J*. 2017;47(11): 1228-1240.

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Title:

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Date:

2019-06-01

Citation:

Dwyer, K. M., Tai, A., Smith, P. & Buchbinder, R. (2019). Real pain in the neck: giant cell arteritis presenting with non-necrotising fasciitis and fever. INTERNAL MEDICINE JOURNAL, 49 (6), pp.802-804. <https://doi.org/10.1111/imj.14317>.

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