Improving People's Lives Through Innovations in Personalized Health Care

IgG4 related disease:
presentation of a paraspinal mass

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CC: arm weakness, L > R

Initial HPI

46yoF p/w LUE loss of function in her LUE progressing to RUE with chronic neck and shoulder pain. No lower extremity symptoms or sensory deficits.

Outside MRI obtained by prior surgeon demonstrated a mass

Non diagnostic outside biopsy

History

PMH: Arthritis, Migraines, Bipolar, Narcolepsy

PSH: Cervical diskectomy, c-section, RTC repair

Social Hx: every day smoker, 30+py

FMH: Mother w/ DM, Breast cancer

Father w/ DM, TB, Prostate cancer, Lung cancer

ALL: Abilify, Compazine, Phenergan, Prednisone, Tramadol

Current Rx: lots of Morphine, some Percocet, Gabapentin, Topomax, Dextroaphetamine, Propranolol

Initial PE

AFVSS

LUE:

0/5 deltoid, 0/5 biceps

2/5 triceps, 2/5 intrinsic hand muscles

Decreased to light touch and pinprick in multiple dermatomes

RUE:

4/5 shoulder abduction, otherwise 5/5 throughout

BLE: 5/5 throughout

Outside Imaging

Per notation:

Poor quality MRI of cervical spine

Postoperative changes at the diskectomy site

Left paraspinal mass from the right from the left neural foramina at C5-6-C6-7 and C7, T1 remodeling in the neural foramina.

The lesion right tracks along the course of the C5-C6-C7 and T1 nerve roots

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A&P

Paraspinal mass, concern for lymphoma

MRI with and without contrast of her cervical spine as well as thoracic, lumbar, brain MR

Referral to Hematology/Oncology

ESR, CRP ordered and elevated

F/u in 2 weeks

Heme/Onc eval

Outside PET with increased activity in paraspinal mass and lower activity in adrenal gland

+ pain x1y now poorly controlled, fatigue, poor appetite, 15# wt loss, low grade fever. No night sweats, chest pain.

Vaccines and cancer screening are UTD and normal

Additional PMH of skin lupus, FMS, Seizures

No LAD on exam

Directly admitted for diagnostic evaluation

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Admission Labs

UA normal

Chem normal

LFT normal

CBC w/ diff normal

Coags normal

Imaging

MRI C-Spine:

Significantly progressed contrast enhancing **left paraspinal and epidural mass from C4-T1**, invades the adjacent musculature and encases the left vertebral artery. There is invasion of the left greater than right C4-T1 neural foramina and circumferential involvement of the epidural space with compression of the spinal cord and encasement of the exiting nerve roots..

MRI remaining T-L spine, Brain: normal

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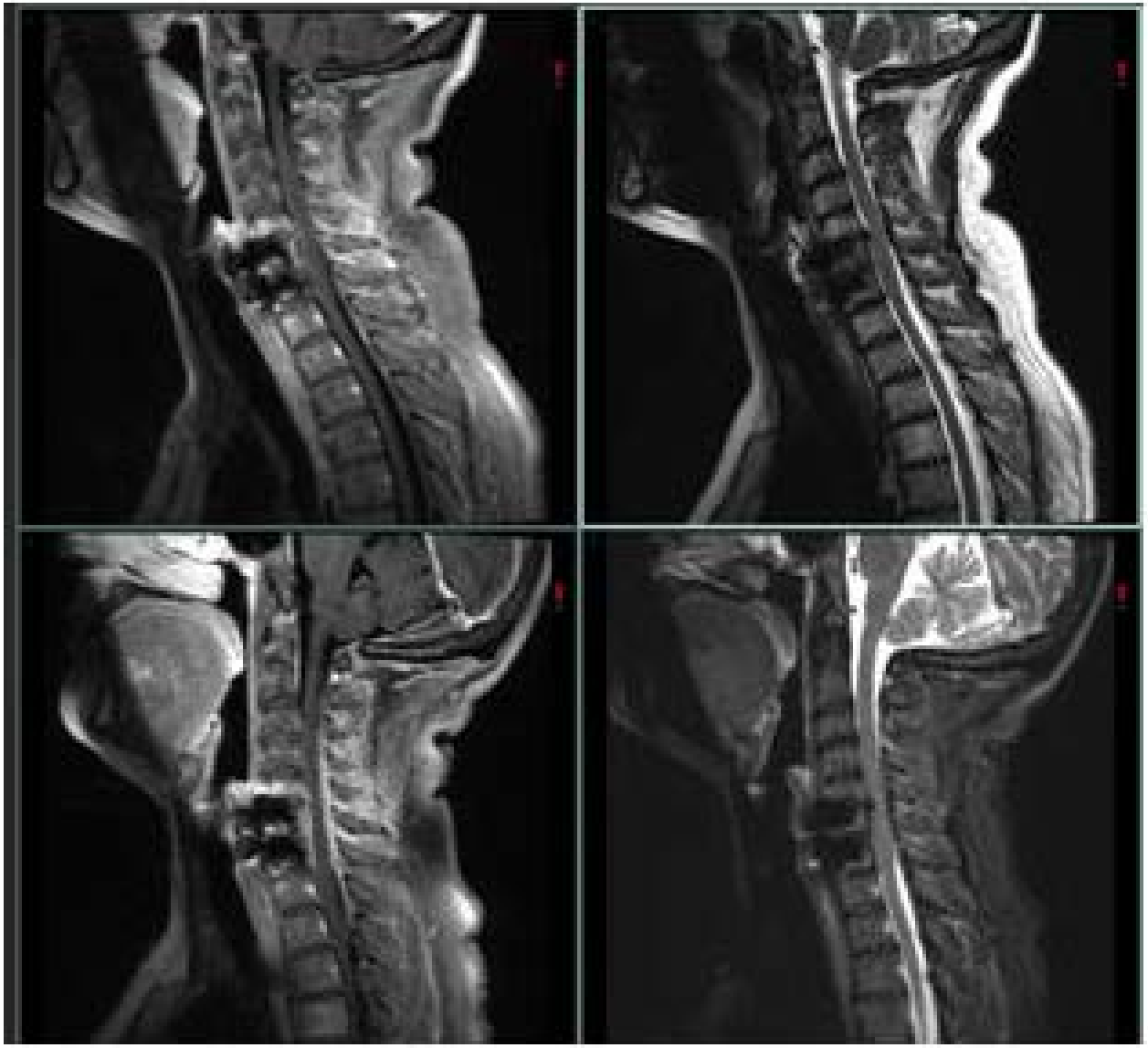
MRI



Imaging

CT Chest and Pelvis w/ IVC:

No lymphadenopathy or primary masses

CTA Spine:

Left vertebral artery is completely encased from C4 to C7. No focal intraluminal filling defect or occlusion

Surgical Pathology

Fibrotic tissue, vessel, and nerve with lymphoplasmacytic and acute inflammation.

No malignancy. Given the presence of dense fibrosis and many plasma cells, IgG 4 immunohistochemical stain was requested (sent out). Other differential consideration includes reactive process to prior surgery.

Focal histiocytes/ giant cells reaction is seen. Special stains for microorganisms were negative.

ADDENDUM: Scattered IgG4 positive plasma cells, focally reach 10/HPF.

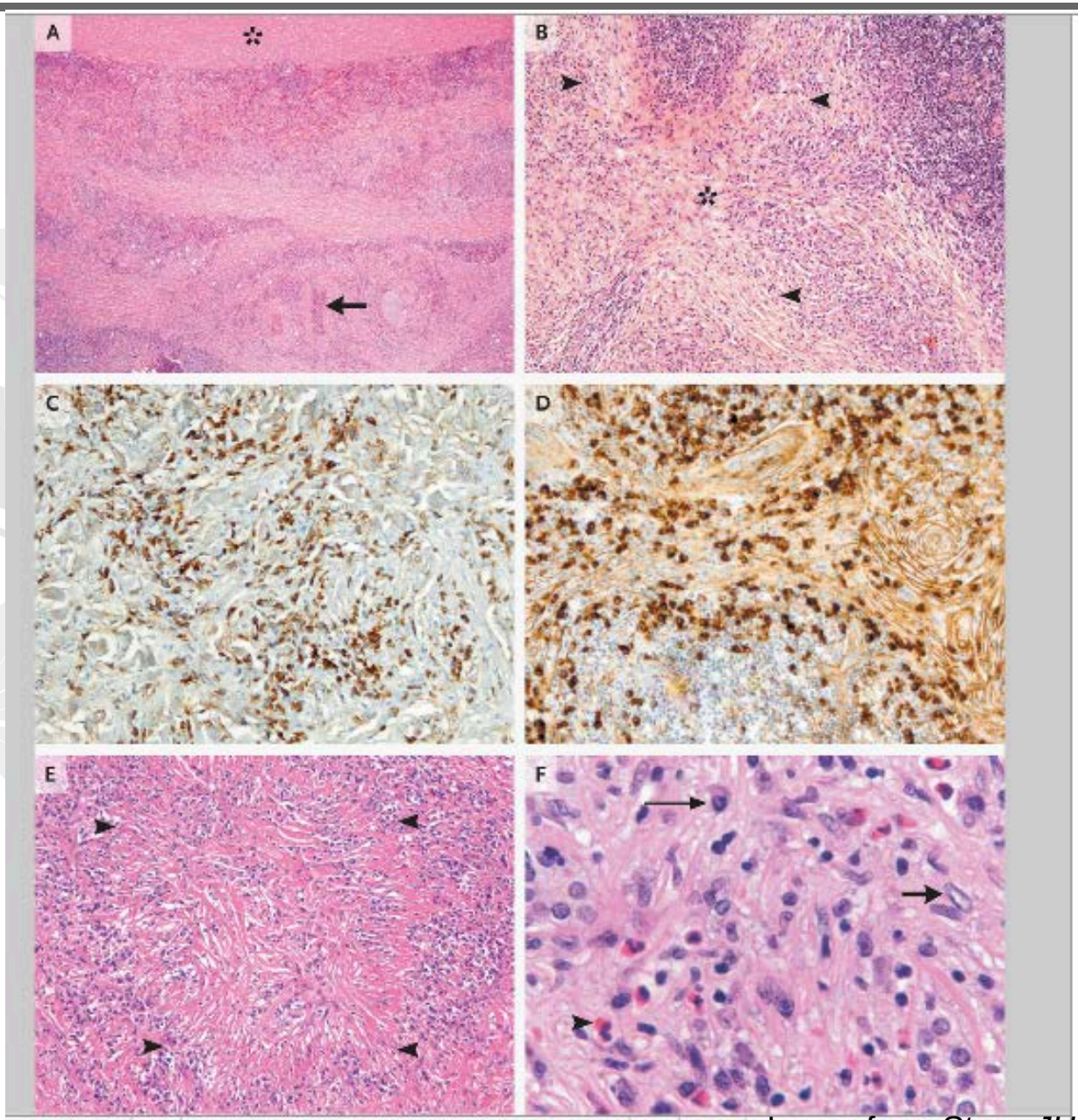


Figure 2. Histopathological Features of IgG4-Related Disease.

A tissue specimen from a patient with IgG4-related aortitis shows virtually the entire wall of the aorta (Panel A, hematoxylin and eosin). Although the media (inner layer, asterisk) is relatively unaffected, a dense lymphoplasmacytic infiltrate is present on the adventitial aspect (outer layer) of the aorta, and a vein obliterated by inflammation is indicative of obliterative phlebitis (arrow). Storiform fibrosis (Panel B, hematoxylin and eosin) is characteristic of IgG4-related disease, such as IgG4-related dacryoadenitis. The pattern is often likened to a cartwheel, with the bands of fibrosis (arrowheads) emanating from the center (asterisk) representing the spokes of the wheel. On immunoperoxidase staining, nearly all the plasma cells in specimens from a patient with IgG4-related aortitis (Panel C) and a patient with IgG4-related dacryoadenitis (Panel D) are strongly positive for IgG4, whereas the small lymphocytes are negative. A specimen of a venous channel (Panel E, hematoxylin and eosin) is characterized by total obliteration (i.e., obliterative phlebitis). Arrowheads mark the periphery of the vein. A high-power image of the specimen shown in Panel E (Panel F) shows lymphocytes, plasma cells (long arrow), eosinophils (arrowhead), and fibroblasts (short arrow).

Image from *Stone JH, Zen Y, Deshpande V. NEJM.2012 Feb*.

This particular case pathology is pending approval for image use.

Further testing

CSF cytology normal

SPEP normal

Multiple cultures negative

TB quantiferon negative

Hepatitis panel negative

ANA negative

IgG4 38 (normal 4-86 mg/dL)

Interval Hx

Diagnosed with IgG4 related disease

Started high dose steroids

Rheum consulted for pathology result

Discharged next day on Prednisone 50 mg QD PO

1m F/U: mild improvement in symptoms, remained on Prednisone 50mg QD PO

Labs stable, TPMT present

Started Azathioprine up-titrated to 100 mg QD PO, prolonged steroid taper initiated with prophylactic measures

Interval Hx

Continue clinical improvement

Gross motor intact with residual for 4/5 left bicep, 3/5 left shoulder abduction

Continued paresthesia in left hand with diminished sensation to light touch in left upper extremity

Remains on AZA and prolonged Prednisone taper for IgG4 disease

IgG4 discussion

Most common presentation = Type 1 Autoimmune Pancreatitis (AIP)

- sclerosing cholangitis
- salivary gland disease
- orbital disease
- retroperitoneal fibrosis
- tubulointerstitial nephritis
- lung opacities, mediastinal fibrosis
- lymphadenopathy
- aortitis, periaortitis
- thyroid disease

IgG4 Dx

Presentation and differential depends on organ involvement

Tends to occur in middle age men although this is biased toward AIP presentation

Autoimmune and allergic components

Responds very well to steroids

Elevations in IgG4 are non-specific and can be seen in other rheumatic disorders

Diagnosis is based on mainly on characteristic pathology findings

IgG4 Dx

Pathologic diagnosis of IgG4 related disease requires the presence of 2/3 histological features:

1) dense lymphocytic infiltrate
2) fibrosis, arranged at least focally in a storiform pattern
3) obliterative phlebitis

Correlation with clinical and radiological findings, serum IgG4 levels, as well as steroid treatment response is generally recommended

IgG4 Dx

Treatment algorithms are not well established

Glucocorticoids are the only

Case studies using Ritximab

Azathioprine and Mycophenolate mofetil have been studied in AIP

Prognosis not established

Morbidity and mortality relate to organ involvement

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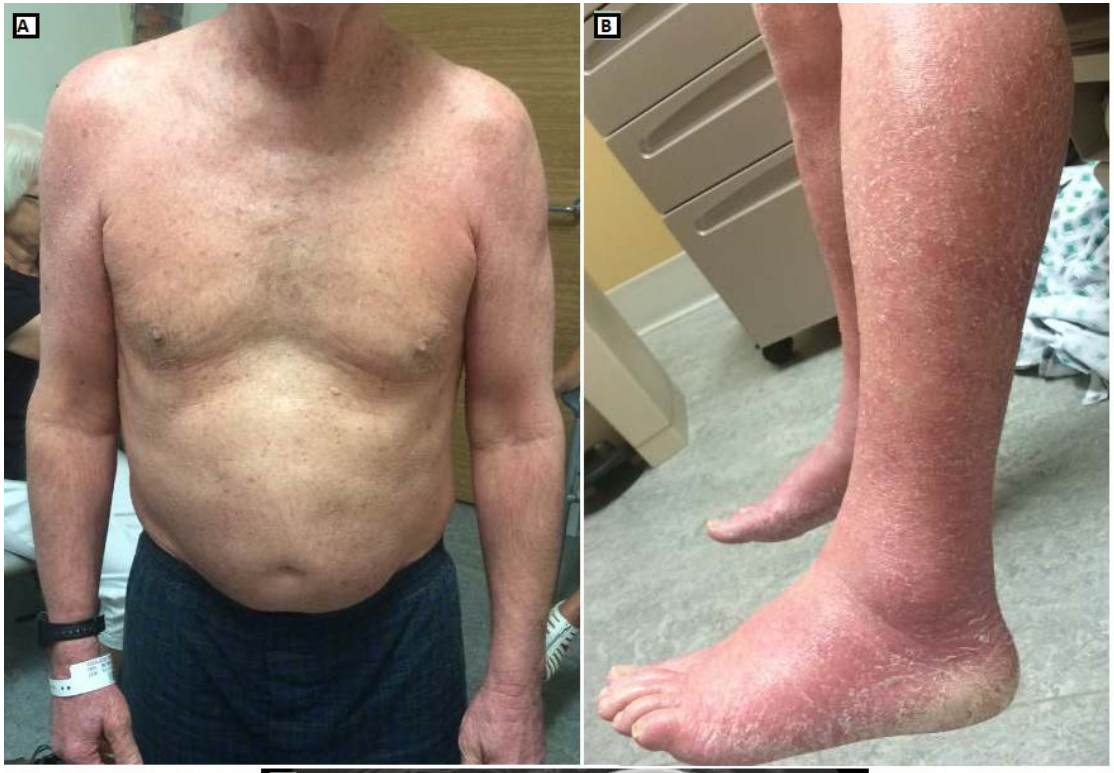
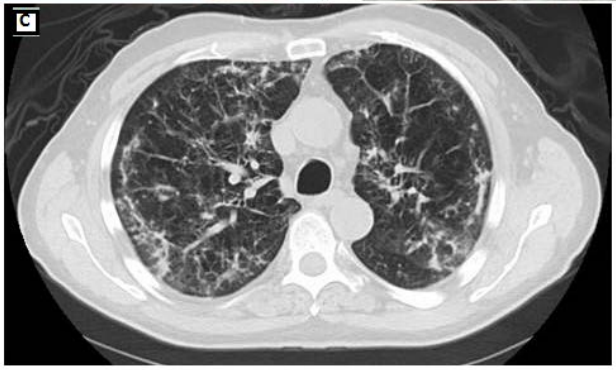
Other OSU IgG4 Case





Presented like cutaneous T cell lymphoma

Images from Stephanie Fabbro, M.D., dermatology resident

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