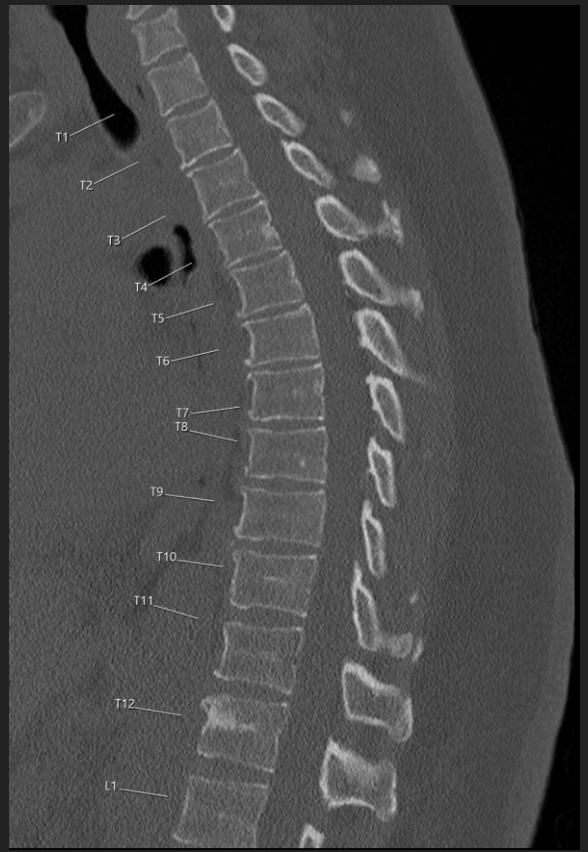
Immunoglobulin G4-related spinal pachymeningitis

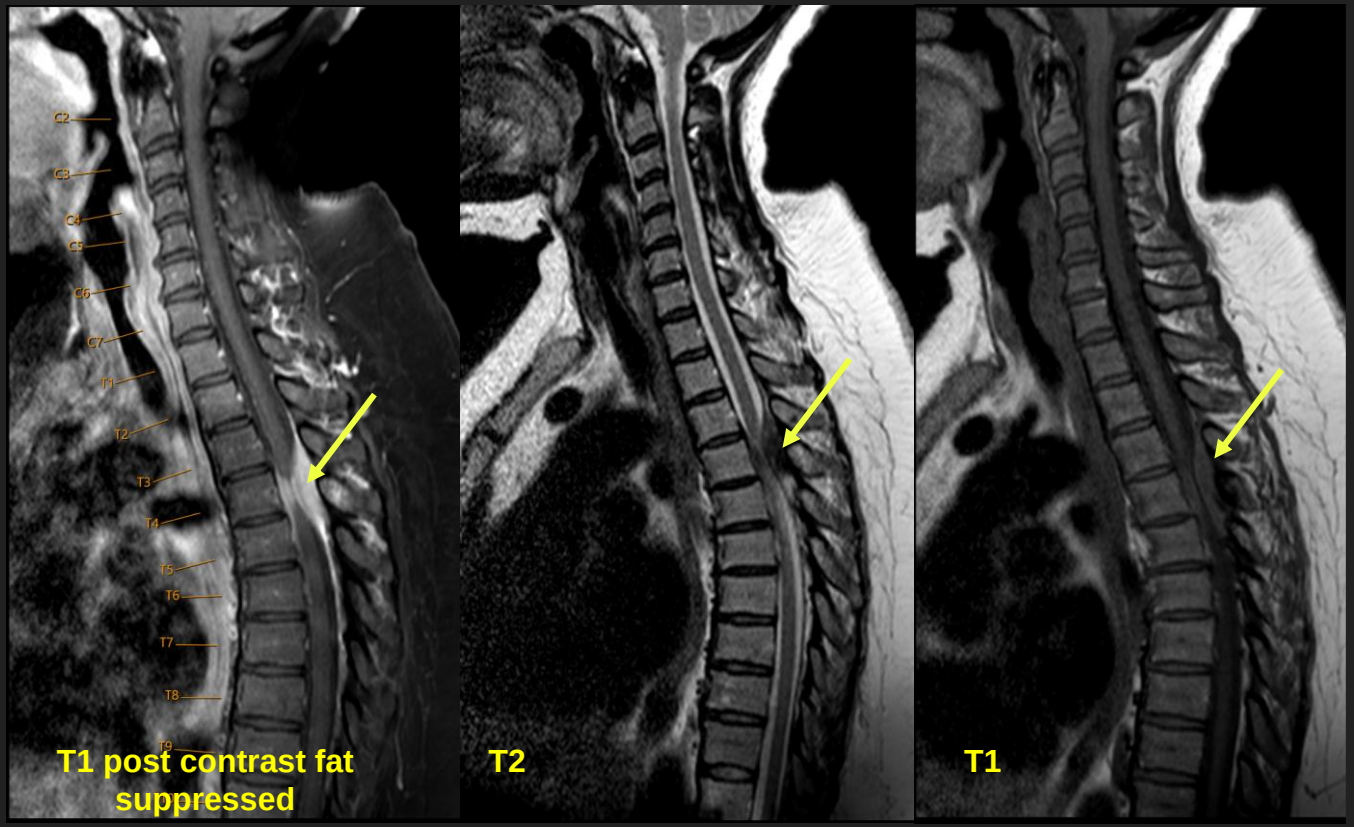
Abstract:

Immunoglobulin G4-related disease (IgG4-RD) is a rare inflammatory disorder that can affect multiple organs, including the pancreas, salivary and lacrimal glands, and retroperitoneum, mimicking malignancy, infectious diseases, and other inflammatory disorders. IgG4-related spinal pachymeningitis is particularly uncommon. Symptoms often include neck pain and signs of spinal cord compression. We report the case of a patient with worsening pain and numbness, found to have pachymeningeal thickening on cervical MRI and elevated serum IgG4 levels. Treatment included glucocorticoids, azathioprine, Imuran, and laminectomy, which resulted in some clinical improvement.

Clinical presentation:

A 54-year-old woman presented with progressive lower extremity weakness and numbness. She had mid-thoracic back pain following a fall and subsequent acute numbness from the sternum downwards. Initial CT scans were negative, and a steroid course provided no relief. Later the patient's symptoms worsened, requiring a walker. Her history included gastric bypass surgery, obesity, OSA, cholelithiasis, GERD, and incomplete RBBB. She had no family history of cancer and had an active lifestyle.





Homogeneous enhancing extramedullary extradural mass from T3 to T9 (yellow arrows). Increased T2 signal within the compressed cord compatible with edema/myelomalacia.

Management and follow-up:

Neurosurgery performed a T3-T5 laminectomy and debulking of the extradural spinal cord lesion. Pathological examination confirmed IgG4-RD with IgG4> 50, IgG4 in HPF > 200, and obliterative phlebitis with storiform fibrosis. The patient was initially treated with Imuran, later transitioned to azathioprine. At the six-month follow-up, the patient remains on azathioprine therapy, with stable disease and no progression of clinical symptoms.

Take home points:

- The rarity and nonspecific symptoms of IgG4-related spinal pachymeningitis can delay diagnosis. Progressive neurological symptoms are often nonspecific. After diagnosing spinal cord compression, a thorough systemic evaluation is essential due to the potential for the disease to involve multiple organs and the need to exclude other diagnoses. The differential diagnosis includes infectious, neoplastic, and other inflammatory causes of dural thickening, requiring comprehensive evaluation.
- MRI is the primary imaging modality, revealing diffuse or localized dural thickening. However, histopathological examination is still required to confirm the diagnosis, showing lymphoplasmacytic infiltration with IgG4-positive plasma cells.
- A comprehensive systemic examination is essential after diagnosing spinal cord compression due to the potential for multi-organ involvement in IgG4-RD. Early diagnosis and treatment are crucial to prevent irreversible neurological damage.

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