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Neurological picture:

Paravertebral Waldenström's macroglobulinaemia presenting as pseudoclaudication

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A 60-year-old man presented with a 1-year history of progressive, radiating back and leg pain worsened by standing consistent with pseudoclaudication. Neurological examination and electro-diagnostic studies were normal. MRI showed enhancement of the paravertebral soft tissues adjacent to the L5-S1 vertebrae with neuroforaminal involvement (figure 1A). An IgM monoclonal protein was detected in serum and bone marrow biopsy demonstrated plasmacytoid lymphocytes. Image guided biopsy of the abnormal L5 paravertebral tissue confirmed lymphoplasmacytic lymphoma/Waldenström's macroglobulinaemia (WM) (figure 1E–G). Rituximab therapy and lumbosacral radiation were instituted resulting in reduction of the size of the monoclonal protein and soft tissue mass, and improvement of pain. One year later the patient developed recurrent pain. Repeat MRI showed abnormal enhancement in paravertebral soft tissues adjacent to the T10-L3 vertebrae with bilateral foraminal involvement (figure 1B). Positron emission tomography/CT (PET/CT) demonstrated abnormal FDG uptake in paraspinal tissues adjacent to the T10-L3 vertebrae distinct from vertebral bone marrow uptake and marked hypometabolism in previously irradiated lumbosacral vertebrae (figure 1C,D). Image-guided aspiration of the abnormal tissue adjacent to the L3 vertebra was diagnostic of recurrent lymphoplasmacytic lymphoma (WM). Rituximab therapy was reinstituted with clinical and radiological improvement.

WM is a sporadic low-grade lymphoplasmacytic lymphoma characterised by proliferation of lymphoplasmacytic cells in the bone marrow with IgM monoclonal proteins in the serum. The pathological effects of WM result from immune mediated injury, direct tissue infiltration or changes in blood viscosity with central nervous system ischaemia.¹ The most common neurological complication is peripheral neuropathy occurring in 47% of WM patients in the largest cohort study from indirect immune effects.² Direct tissue infiltration by malignant plasmacytoid lymphocytes typically involves bone marrow, lymph nodes and solid organs. Direct infiltration of the nervous system is less frequent, but involvement of the brain, spinal cord and nerve roots has also been reported as rare complications of WM.³

Occasionally tumour infiltration may indirectly cause neurological dysfunction by invasion of neighbouring structures resulting in secondary compression syndromes. Al-Halabi *et al* reported myelopathy from an epidural mass with extension to the spinal cord.⁴ Tojo *et al* reported a paravertebral mass in a patient with WM. This case occurred in the context of

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advanced disease and a coexistent B-cell lymphoma.⁵ Our case-presented with lumbosacral pseudoclaudication due to invasion of WM lymphoplasmacytic cells into the paravertebral soft tissues impinging on the neuroforamina. In this case, the localisation was predicted by the clinical presentation of pseudoclaudication and confirmed with MRI and PET studies. PET imaging studies assisted in the identification of regions containing active disease as previously irradiated tissue had little PET activity consistent with the therapeutic effect of prior radiation, whereas recurrent WM infiltrated areas were hyper-metabolic allowing for targeted biopsy and treatment.

The present case of WM had recurrent infiltration of paravertebral soft tissues with nerve root compromise that responded to radiation and rituximab. This case illustrates a common neurological presentation (pseudoclaudication) resulting from an uncommon illness, that is, Waldenström's macroglobulinaemia.

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Figure 1.

T1 gadolinium-enhanced fat suppressed MRI of (A) L5-S1 and (B) L3 paravertebral mass (arrowheads) with neuroforaminal involvement (arrow). FDG-PET/CT showing increased F18-FDG uptake adjacent to the T10-L3 vertebrae (arrows), and reduced uptake in irradiated vertebral bodies (*) seen in (C) axial and (D) coronal planes. (E) H&E, (F) CD3 and (G) CD20 immunohistochemistry of biopsy of L5 paravertebral mass.