Multiple myeloma related amyloidosis: a rare neurological manifestation
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Abstract
Amyloidosis secondary to multiple myeloma can present as a multisystem disease. Neurological manifestations of amyloidosis are manifold — most commonly lower motor neurone in nature, but only rarely upper motor neurone disorders can be a form of presentation of this disorder. We present here a case of amyloidosis which presented as multifocal peripheral neuropathy and extradural spinal cord compression. The diagnosis of neurological amyloidosis was aided by a histological diagnosis of amyloidosis made on a mucocutaneous lip swelling biopsy. Upon further investigation, it was revealed that the patient also has concomitant myeloma. This case highlights that multiple myeloma and amyloidosis can present as rare neurological presentations, and in similar situations treating physicians must always consider these when faced with diagnostic dilemmas.

Keywords
Access adequacy, nursing staff, haemodialysis, renal clinic, unplanned activities.

Case
A 65-year-old male presented with painful wrists and hands. Clinically there was evidence of median nerve compression bilaterally and this was confirmed on nerve conduction velocity. Bilateral carpal tunnel syndrome was diagnosed and decompression on either side was performed. His symptoms transiently improved following this. However, two years later his symptoms reappeared and gradually progressed to such an extent that he was unable to drive. His general condition also deteriorated. He had a shuffling gait, stiff shoulders, aches and pains in his legs and struggled to lift his legs to climb stairs. He became wheelchair-bound. A diagnosis of Parkinson’s disease with bilateral carpal tunnel syndrome was made.

Three months later he was noted to have a 5 cm in diameter firm nodular swelling on his lower lip. On biopsy this revealed amyloidosis. He was normotensive with clinically no peripheral oedema or hepatosplenomegaly. Further investigations at this point showed Hb 14.5 g/l, white cell count 5.5 x 10⁹/l, neutrophils count 3.3 x 10⁹/l, platelets 245 x 10⁹/l, prothrombin time 12.3 seconds, serum creatinine 69 micromol/l, serum proteins 71 g/l, serum albumin 46 g/l and serum globulin 25 g/l. His serum immunoglobulins were normal and no serum paraprotein was detected. Urinary electrophoresis showed 0.3 g/l proteinuria with Bence Jones proteinuria of 0.15 g/l. Serum free light chains were detected. The kappa free light chains were quantified as 5572 mg/l against a normal of 3.3–19.4 mg/l; lambda free light chains 3.5 mg/l (normal 5.7–26.3 mg/l) and the kappa/lambda ratio was 1592 (normal 0.26–1.65). His liver function tests and serum beta-2 microglobulin were in the normal range.

The patient was referred to the haematologists. In view of the amyloid deposit on the lip and evidence of peripheral neuropathy with raised serum free light chains, investigations were directed to search for systemic amyloidosis.

A bone marrow aspiration and trephine biopsy were performed. The aspirate showed 9% plasma cells with normal appearance. The trephine biopsy demonstrated 50% plasma cells with large nodules of Congo red staining deposit in the connective tissue including the walls of the blood vessels. A SAP scan was performed and this did not show any evidence of visceral amyloid. An echocardiogram performed on him did not reveal any findings suggestive of cardiac amyloid.

His skeletal survey showed lytic lesions in the skull.

To evaluate his neurological condition further, an MRI of his cervical spine was performed (Figure 1). It revealed an abnormal low signal soft tissue mass in the epidural space anterior to the posterior rings of the first two cervical vertebrae causing compression of his spinal cord. This extradural mass on biopsy revealed an amyloid deposit.

Hence, the diagnosis of systemic amyloidosis with asymptomatic multiple myeloma was confirmed. The patient underwent a successful decompression procedure and is currently receiving anti-myeloma therapy.

Discussion
Amyloidosis is a multisystemic disorder characterised by extracellular deposition of amyloid (Kim et al., 2006). It may present as a localised or a systemic disorder (Hwang et al., 2000).

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The most common neurological manifestation is a small-fibre axonal neuropathy (Gutierrez, Turkewitz, Correa & England, 2006). Also there have been reported deposits of amyloid causing epidural cord compression (Haridas, Basu, King & Pollock, 2005; Belber & Graham, 2004). However, amyloidosis continues to be a difficult disorder to diagnose because the symptoms at presentation are vague (Gertz, Lacy, Dispenzieri & Hayman, 2005), even more so when it presents as an atypical polyneuropathy (Gutierrez et al., 2006). Our case is interesting as the patient had neurological signs and symptoms which led to the diagnosis of an atypical presentation of Parkinson’s disease and it was only after a few years that amyloidosis was diagnosed.

The patient had widespread amyloidosis; however, the 113 radiolabelled SAP scan did not reveal any evidence of amyloidosis. The diagnostic sensitivity of SAP scanning in AA- and AL-amyloidosis is 90% (Hazenberg et al., 2006). Localised deposits such as the lip amyloid lump are often not detected on SAP scanning as the uptake of the tracer is proportional to the quantity of amyloid in the deposit (Hachulla et al., 1996).

Serum free light chains gave us a clue to the diagnosis in this case. They are a useful investigation in the diagnosis and management of plasma cell dyscrasias such as myeloma, amyloidosis and light chain deposition disease (Mayo & Johns, 2007).

Multisystemic amyloidosis is associated with multiple myeloma. Up to 43% of cases of amyloidosis in a series were stated to be in patients with myeloma (Saba, Tohme, Abadijan, Haddad & Ghayad, 2005). As the pathogenesis of AL-amyloidosis is similar to that of multiple myeloma, similar therapeutic regimes, directed towards reduction of plasma cell load are used in both these conditions (Hall, Nord & Heimdahl, 1996).

**Implications in clinical care**

Extradural spinal cord compression can be due to various causes. The essential is to have a histopathologic diagnosis for the compressing mass, whenever possible.

Amyloidosis is a rare disorder, and, hence, not one of the differentials usually considered initially. At the same time, amyloidosis is a treatable condition and so the outcome of managing it with high-dose chemotherapy is encouraging (as in this case). The neurological features related to the compression by the amyloid lump completely regress with treatment.

If the biopsy of the compressing mass suggests amyloidosis, immunohistochemistry should be performed. Once AL-amyloidosis is confirmed, the patient should be considered for chemotherapy as in most plasma cell dyscrasias.

**References**


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**Figure 1:** MRI cervical spine reveals an extradural lesion compressing spinal cord at level C2.