Pseudo-Popeye syndrome

Extramedullary plasmacytoma manifesting in skeletal muscle

Figure 1 Clinical presentation

Front and side views of the patient demonstrate the Popeye effect. The patient complained of painful swelling of both forearms (A), most evident on the left side (B). A massive tumorous bulge caused compartment syndrome, which affected the median and ulnar nerves on the left side.

Figure 2 Histopathologic findings

Muscle biopsy revealed some atrophic muscle fibers displaced by infiltrating immunocytes, visualized using hematoxylin & eosin, 100 $\times$ (A). Immunohistochemical analysis identified plasma cells using CD138 antibody, 50 $\times$ (B) with isolated secretion of kappa light chain, 400 $\times$ (C). Plasma cell character was confirmed ultrastructurally, PC plasma cell, M macrophage (D).

A 75-year-old woman with medullary plasmacytoma reported progressive swelling of the forearms, left more than right (figure 1), with median nerve contribution pain. Examination revealed distal sensory loss and left hand weakness with atrophy of the median and ulnar nerve-innervated muscles. Arterial pulses were palpable. Electrodiagnostic studies showed ulnar nerve conduction block and left thenar muscle denervation. MRI showed subcutaneous and muscle edema in the forearm flexor muscles, mimicking Popeye syndrome.¹ Fasciotomy for compartment syndrome and muscle biopsy were performed. Muscle showed lymphoplasmacytic infiltrates (figure 2), a rare extramedullary manifestation of plasmacytoma.^{2,3} Sensory loss and muscle strength improved after fasciotomy. Therapy for plasmacytoma was bortezomib/dexamethasone plus radiotherapy of both forearms.

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