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Popeye Deformity: A Red Flag for Wild-Type Transthyretin Amyloidosis

To the Editor:

CASE REPORT

A 72-year-old man with past medical history of hypertension was referred to a cardiology consultation due to recent heart failure hospitalization and newly diagnosed atrial fibrillation. Physical examination revealed a large bulge on his anterior mid-upper left arm, more pronounced during elbow flexion (Figure). This deformity had developed several months before, after lifting a heavy object. He remembers hearing a "pop" sound, followed by a sudden sharp pain and weakness in his left arm. He did not seek medical care and started self-treatment with cryotherapy and nonsteroidal anti-inflammatory drugs. The pain totally relieved within a few days and no longer affected his daily life.

Resting electrocardiogram revealed atrial fibrillation with a normal ventricular rate. Transthoracic echocardiogram showed asymmetric septal hypertrophy with granular sparkling appearance, normal left ventricular ejection fraction yet reduced global longitudinal strain (−9.7%) with "apical sparing" pattern, impaired diastolic function, and severe biatrial dilatation.

Transthyretin amyloidosis was diagnosed after diphosphonate scintigraphy displaying strong myocardial uptake of 99mTc-DPD (Perugini scoring system: grade 3) and laboratory testing that excluded immunoglobulin light-chain amyloidosis. Genetic testing was negative for hereditary transthyretin amyloidosis, indicating wild-type transthyretin amyloidosis. Treatment with tafamidis was initiated.

Figure Patient's left arm in flexion position. A deformity (Popeye sign) is perceived in the anterior mid-upper arm.

DISCUSSION

This typical finding on physical examination is known as Popeye sign or Popeye deformity, named after the ball-shaped biceps of the fictional cartoon character of the 1930s. It is caused by bulging of the biceps muscle belly after rupture of the long head of the biceps tendon.[1] Rupture of this tendon is not uncommon;[2] however, a patient presenting with the Popeye sign is rare.

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The risk factors include older age, smoking habits, corticosteroid use, fluoroquinolone antibiotics, and biceps muscle overuse. As forearm flexion and supination are maintained by the short head of the biceps, conservative management is sufficient for most cases. Nonetheless, patients whose occupations demand full arm strength may require surgical repair.[1]

Cardiac involvement in systemic amyloidosis, due to either immunoglobulin light-chain or transthyretin amyloidosis, is a major determinant of unfavorable outcome.[3] It is still mis/underdiagnosed, although effective treatments in improving patient survival are now available, particularly for transthyretin amyloidosis. Recognition of early clinical findings, some of them preceding cardiac manifestations, are key to achieving a timely diagnosis. Clues to extracardiac amyloid deposition in wild-type transthyretin amyloidosis include bilateral carpal tunnel syndrome, lumbar spinal stenosis, or biceps tendon rupture, which may produce the Popeye sign.[3]

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CONSENT

The authors confirm that written consent for submission of the case report including images and associated text has been obtained from the patient in line with COPE guidance.

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