Hirayama disease, also known as Juvenile Muscular Atrophy of Distal Upper Extremity, is a benign, uncommon disease predominantly seen in young men of Southeast Asian descent. It often presents with insidious onset of muscular atrophy of the hands and forearms, and generally spares the brachioradialis. This is known as oblique amyotrophy. Patients may also feel trembling and “cold paresthesia”, or weakness of fingers when exposed to cold.

The primary predisposing factor is an imbalance in growth of the cervical spine that occurs during neck flexion. This causes the posterior wall to shift and compress the cord against the vertebral bodies. The chronic flexion of the neck likely causes micro-vascular ischemia to the anterior horn resulting in gliosis and atrophy of the cord.

Hirayama disease is best characterized with MR of the cervical spine in neutral and flexed positions. In the neutral position, MR reveals localized lower cervical cord atrophy, asymmetric cord flattening, and loss of attachment. In flexed position, MR reveals anterior displacement of posterior dural sac, loss of attachment, and increased posterior epidural tissue containing serpiginous flow voids. Focal gliosis and atrophic narrowing of the involved lower cervical cord can occur over time from chronic spinal cord impingement and persists on imaging in both flexion and neutral positions. The serpiginous flow voids demonstrated on MRI corresponds with engorgement of the epidural venous plexus. While the epidural venous plexus engorgement is thought to be a passive and transient process in association with neck flexion, it may contribute to the anterior horn damage of the spinal cord. Application of a cervical collar for 3 to 4 years has been shown to stop progression of the disease.

Another treatment alternatives include anterior cervical decompression, duraplasty, and reconstruction with tendon transfer.

References