

Supplementary Online Content

Kim EJ, Park YE, Kim DS, Ahn BY, Kim HS, Chang YH, Kim SJ, Kim HJ, Lee HW, Seeley WW, Kim SY. Inclusion body myopathy with Paget disease of bone and frontotemporal dementia linked to *VCP* p.Arg155Cys in a Korean family. *Arch Neurol*. 2011;68(2):doi:10.1001/archneurol.2010.376

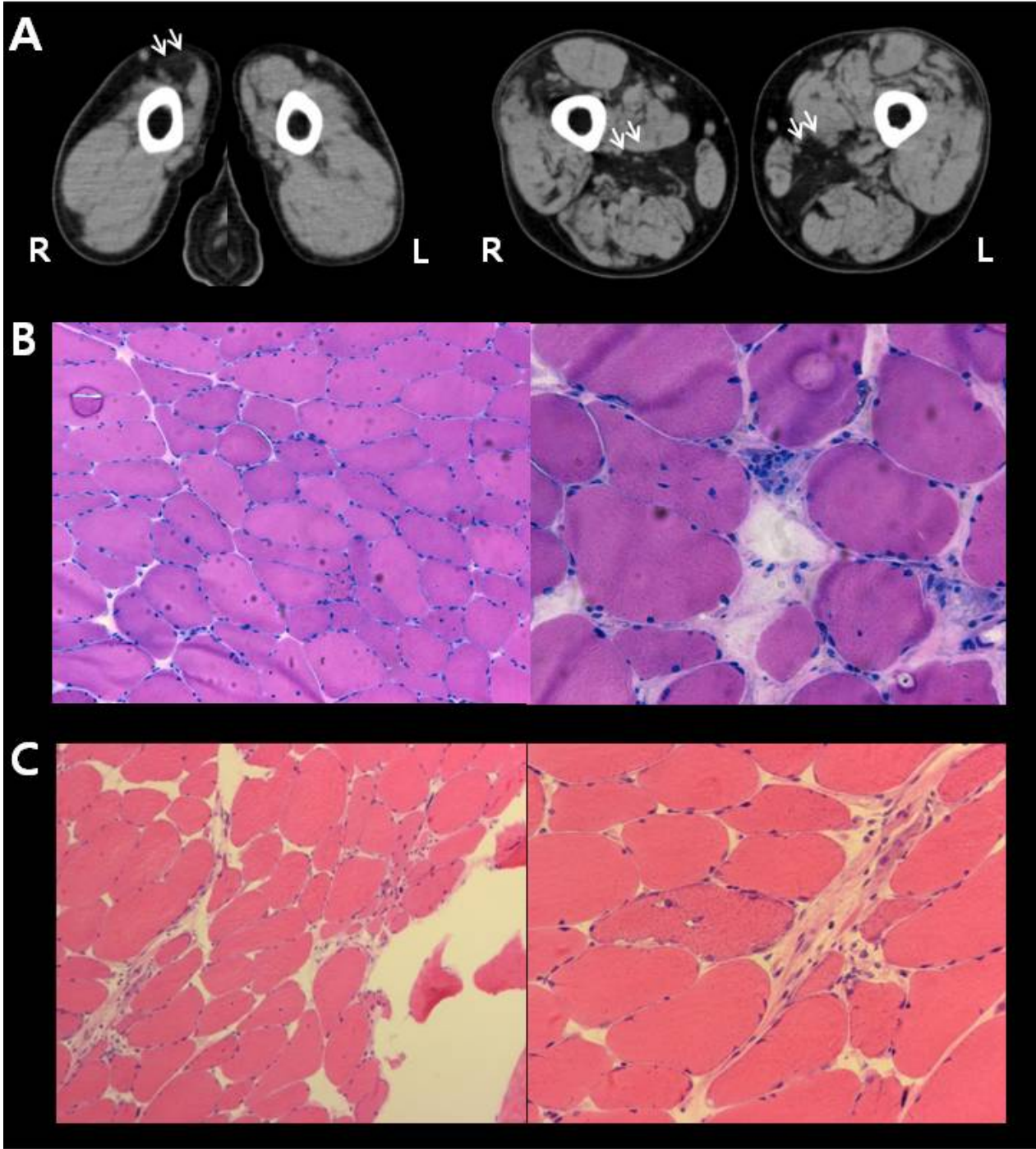
eFigure 1.

eFigure 2.

eFigure 3.

This supplementary material has been provided by the authors to give readers additional information about their work.

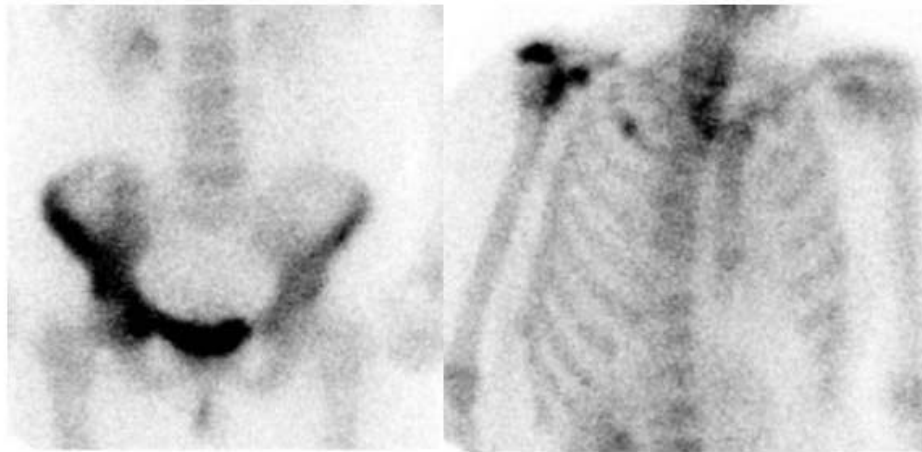
eFigure 1.



eFigure 1.

- (A) Computed tomography (CT) scans of the biceps muscles (left) and mid-thigh muscles (right) in P2 showing mild to moderate focal fatty replacement (arrows). R: right, L: left.
- (B) Right biceps muscle biopsy in P2 at the age of 54 showing only minimal muscle size variation (right, x 100) with few necrotic fibers (left, x 200) in hematoxylin-eosin staining.
- (C) Left biceps muscle biopsy in P2 at the age of 57 revealing grouped atrophic fibers with angulated fibers (right, x 100) and cytoplasmic rimmed vacuoles (left, x 200) in hematoxylin-eosin staining.

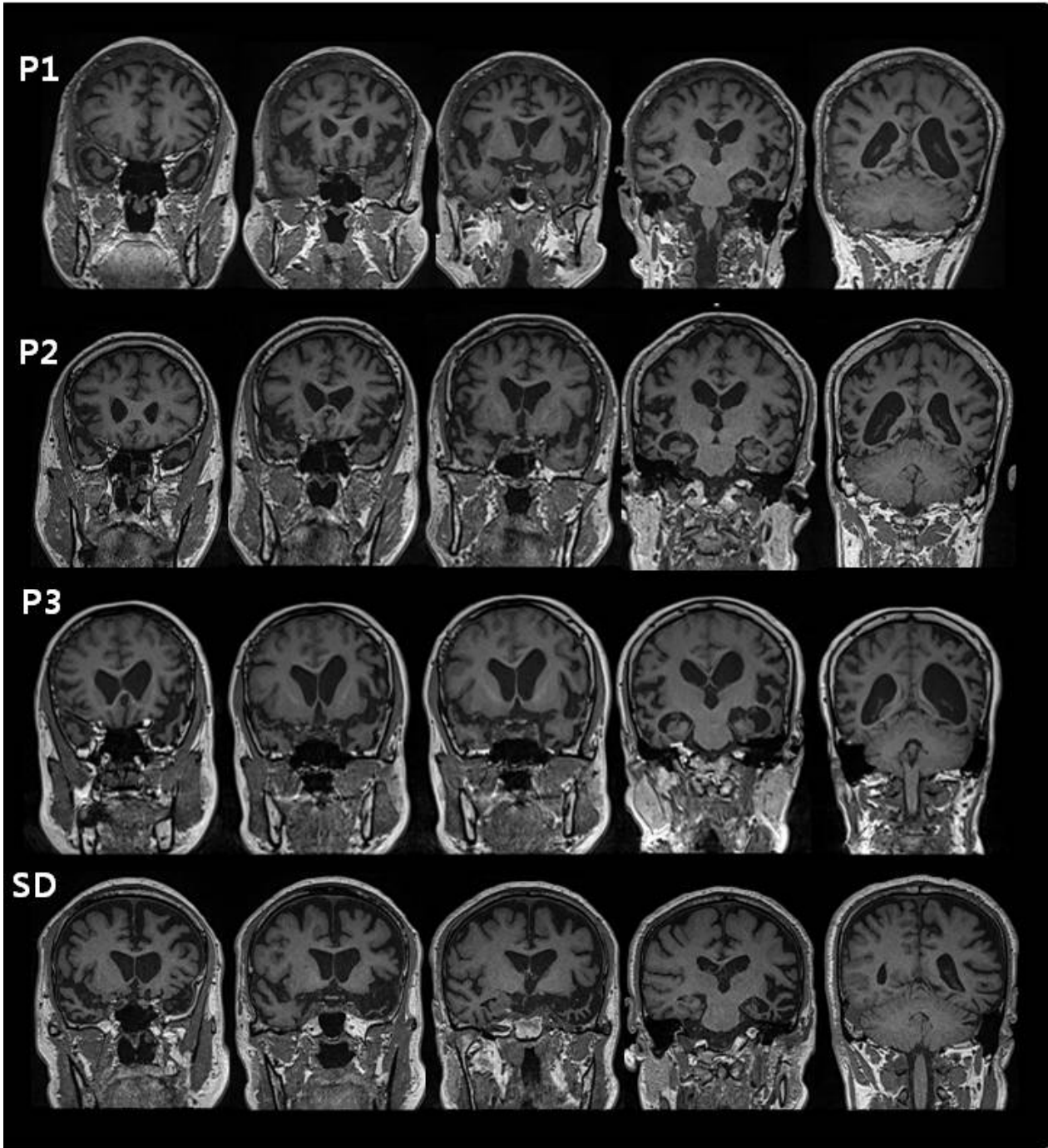
eFigure 2.



eFigure 2.

A ^{99m}Tc bone scan of P3 demonstrating active bone lesions on the right pelvis and right shoulder, compatible with Paget's disease of bone.

eFigure 3.



eFigure 3.

T1-weighted coronal brain MRIs of three affected IBMPPFD patients (P1, P2, P3) and a 62-year-old male with typical semantic dementia (SD). Brain MRIs of the IBMPPFD patients demonstrated asymmetric anterior and lateral temporal and inferior parietal atrophy (left predominant atrophy in P1 and P3, right predominant atrophy in P2) compared to the MRIs of the SD patient, which showed severe left anterior temporal and frontal atrophy.