Lipoid proteinosis is a rare hereditary disease which often results in bilateral calcifications in the medial temporal region. Thirty-four adults living with lipoid proteinosis (>10% of the world population) were extensively assessed with standardized neuropsychiatric and neuropsychological measures. Of these, 27 patients representing a homogenous group living in the Northern Cape were matched with 47 controls. Subjects with lipoid proteinosis had a high incidence of neuropsychiatric disorders and performed poorly on facial recognition of positive and negative emotions and on many neuropsych...