Bilateral symmetric changes in the cerebral hemispheric white matter are found with increasing frequency using CT and MRI techniques. These unspecific changes of the white matter signal are often called leukoaraiosis. They differ from the normal white matter signal. These changes are found with increasing frequency in persons older than 60 years and also patients with dementia and cerebrovascular diseases. The pathogenesis, clinical significance and morphological substrate are unclear. The aim of this review is to summarise the actual knowledge about the etiology and clinical signs and symptoms found in patients with leukoaraiosis. This term should not be used when white matter changes are found in patients younger than 35 years, with an unilateral onset, asymmetric distribution, and extensive changes all over the infra- and supratentorial white matter area. Neuroradiological and clinical criteria are given to differentiate between leukoaraiosis and diseases of the white matter, especially enlarged Virchow-Robin spaces, lacunar infarction, subcortical arteriosclerotic angiopathy (Binswanger's disease), leukencephalopathy of different origin, and demyelinating diseases.