The purpose of the present study was to evaluate periodontal health status in children with sickle cell disease (SCD). Forty-nine children with SCD and 39 systemically healthy sex- and age-matched children were enrolled in the study. Plaque index, gingival index, bleeding on probing, pocket depth, salivary volume, and hyperplastic index were recorded. In addition, the histopathological evaluation of gingiva was made in a child with SCD. There were significant differences between the groups with regard to hyperplastic index (P < 0.05), whereas there were no differences in other parameters. Gingival enlargement was detected in 27 children (55.1%) in the SCD group and 6 children (15.4%) in the control group (P < 0.001). However, there were no differences in periodontal health status of children in the SCD and control groups, the most important finding of this study that the gingival enlargement was more prevalent in children with SCD. Sickling and chronic inflammation seen in SCD may affect gingival tissues. Therefore, physicians and dentists must be aware of the effects of SCD on gingival tissues.