Maffucci syndrome is characterized by the sporadic occurrence of multiple enchondromas together with multiple hemangiomas. Patients with Maffucci syndrome are at increased risk of developing different kinds of malignant tumors. We report on a 39-year-old woman who was diagnosed with Maffucci syndrome together with intrahepatic cholangiocarcinoma (IHCC). Heterozygous somatic mutations in the isocitrate dehydrogenase 1 and 2 (IDH1/IDH2) genes are associated with a number of different tumor types (e.g., IHCC) and also with Maffucci syndrome. For IHCC, mutations in IDH1/IDH2 are associated with higher survival rates. IHCC tissue as well as normal liver tissue and peripheral blood were analyzed for IDH1/IDH2-mutations in our patient. In the tumor sample, we identified a recurrent somatic IDH1-mutation affecting Arg132, while in normal liver tissue and peripheral blood, no variants were detected, as expected. This case report presents the second patient in the literature exhibiting the features of Maffucci syndrome along with cholangiocarcinoma. This supports the hypothesis that IDH1/2-mutations, which can be present in different types of tumor tissue simultaneously, arise during embryonic development in a mosaic pattern; as a result, a more aggressive follow-up is proposed in patients with Maffucci syndrome to exclude neoplasms.