

Abstract

Chronic kidney disease (CKD) is a devastating disorder which carries high morbidity and mortality if left untreated. The congenital anomalies of the kidney and urinary tract was the predominate cause for CKD in younger patients. We present an adolescent who presented with chronic kidney disease due to bilateral renal agenesis. She had features of external and internal ear abnormalities with history of branchial cyst excision during early childhood. These features were tallying with the clinical diagnosis of Branchio-Oto-Renal syndrome. This case emphasis the importance of identifying relevant congenital associations of syndrome as there may be a reversible component of them which will improve the quality of life of the patient.