Osteopetrosis (OP) is a rare hereditary bone disorder characterized by a marked increase in bone density due to a defect in remodeling caused by the failure of normal osteoclast function. There are two major clinical forms; the autosomal dominant adult form and autosomal recessive infantile form.

Adult form OP is the most common form of OP. Although the first findings in adult form OP are generally nontraumatic long bone fractures and osteomyelitis of the mandible, the infantile form, if untreated, is fatal during infancy or early childhood.

Typical radiographic features of OP were characterized by generalized uniform increase bone density. The aim of this report was to present a case of adult OP, complicated by osteomyelitis of the mandible, with cone beam computed tomography findings.