Primitive neuroectodermal tumor (PNET) is a term used to describe a group of highly malignant neoplasms composed of undifferentiated round cell tumors of neuroectodermal origin with variable cell differentiation. PNET generally occurs in the central nervous system (CNS), but peripheral PNET occurring outside the CNS has also been reported. The most common location of PNET in head and neck region is the orbit, followed by the neck and the parotid gland. PNET of the maxilla is rare. The aims of this report are to present the case of a 41-year-old female diagnosed with PNET tumor of the maxilla after detailed clinical, radiological, and histopathological examinations and to review the previously reported cases of PNET of the maxilla in the literature.