Brain metastases (BM) represent the main cause of intracranial neoplasms in adults, while being relatively less common in children. Today, better treatment options of the primary malignancy lead to higher remission rates as well as prolonged stable clinical conditions. This may in part explain the increased incidence of BM. Morbidity and mortality rates in patients with malignancies deteriorate significantly in cases of metastatic involvement of the central nervous system. Nowadays, especially modern management using surgical, medical, and radiotherapeutic options for treatment of BM tends to improve survival rates and enhance quality of life. Nonetheless, almost all treatment options are considered as palliative. In this review, we outline current knowledge of the incidence, diagnostic facilities, and therapeutic management of rare BM, with consideration of the basic aspects of the primary malignancy.