Pleomorphic xanthoastrocytoma (PXA) is a rare primary brain tumour which occurs in children and young adults. PXA is designated as low-grade astrocytoma (WHO II), although an anaplastic variant and malignant potential have been described. Pathologically, cellular pleomorphism is the rule and includes spindle cells, mononucleated and multinucleated giant cells, and granular bodies in a reticulin-rich background with few mitoses; necrosis is usually absent. The tumour cells stain positive for glial fibrillary acidic protein (GFAP). Surgery is the mainstay of treatment with gross surgical resection being accomplished due to its well-circumscribed nature and peripheral location. The role of adjuvant treatment remains yet to be clearly defined. To date, the majority of PXAs have been reported as single or small case series; consequently data of this rare brain tumour are fragmentary. The present paper reviews the pathogenesis, neuroradiological features, prognostic factors and treatment options for PXA.