DISCUSSION

Pineal gland is located in the supratentorial midline above the superior colliculi and below the vein of Galen. It is made up of cells called pineocytes. Pineal tumors account for 1% of all brain tumors.[1] Pineal gland often considered to be vestigial, is the site of occurrence of a small percentage of intracranial space occupying lesions. Radiologic techniques, and stereotactic surgery have made the tumors of this region more accessible and amenable to treatment.

Tumors of the pineal region have a varied histology that are generally germ cell tumor and non-germ cell derivatives. Hence, most tumors are result of displaced embryonic tissue, malignant transformation of pineal parenchyma cells or transformation of surrounding astrocytes.[2] PTPR is a relatively uncommon tumor of the pineal gland and was initially described by Jouvet et al, in 2003.[3] This entity was included in WHO 2007 CNS classification of tumors. PTPR is a neuroepithelial tumor that is localized in the pineal region and characterized by a combination of papillary and solid areas with epithelial –like cells. The biologic behavior of this tumor is variable and may correspond to grade II or III. These tumors arise exclusively in the pineal region and occur most commonly in adults with slight female preponderance.

On neuroimaging, PTPR present as well circumscribed heterogenous masses composed of cystic and solid portions and centered by the posterior commissure or the pineal region. Aqueductal obstruction with hydrocephalus is a frequent associated finding. Radiologically these tumors are misdiagnosed as tectal gliomas.[4]

Morphologically, this is an epithelial looking tumor with papillary features and more densely cellular areas with ependymal like differentiation. Cells with clear and vacuolated cytoplasm are seen and occasionaly may have eosinophilic periodic acid Schiff positive cytoplasmic mass. The mitotic count ranges from 0 to 13 mitoses per 10 high power fields. Ki67 proliferation index ranges from 1 to 29.7%. The most distinctive feature that distinguishes this tumor from other native tumors of the pineal gland is reactivity for keratins (AE1/ AE3, CAM5.2, and CK18). GFAP is less commonly seen. These tumors also stain for vimentin, S100, NSE, NCAM1 and transthyretin. Synaptophysin and chromogranin are sometimes weakly and focally expressed.[5]