DISCUSSION

Epithelioid glioblastoma is a newly added distinct variant of glioblastoma in the 2016 revised World Health Organization classification of CNS tumors. It is classified under the IDH-wildtype glioblastoma along with Glioblastoma and Gliosarcoma[1].
Histologically they are characterized by uniform population of epitheloid to rhabdoid cells with eosinophilic cytoplasm and laterally positioned nucleus. Intervening neuropil is usually scant. Microvascular proliferation is present. Necrosis is usually zonal unlike the palisading necrosis of other types of GBMs.

The cells show immunoreactivity for Vimentin, S100 and epithelial markers like CK and EMA. Focal immunoreactivity for GFAP, NSE and synaptophysin is also seen.[2] IHC using the VE1 antibody for V600E mutant BRAF is positive in 50% cases.

50% cases show BRAF V600E mutation. A single case with H3F3A/K2M mutation has been reported but characteristically mutations for IDH1 and IDH2 are not seen. EGFR amplifications and loss of PTEN have also been reported in some cases.[3]

It is important to diagnose these cases as a distinct subset of GBMs as they show a particularly poor prognosis along with a dismal median survival. The identification of BRAF V600E mutation is important both to understand the biology of the tumor and as a potential therapeutic target.

CONCLUSION

Epithelioid Glioblastoma is a rare histological type of WHO grade IV astrocytic tumors with epithelioid to rhabdoid cells along with atypical mitosis, microvascular proliferation and zonal necrosis. It is a unique entity given its histological picture, IHC staining pattern and underlying mutation, which in 50% cases involves BRAF V600E unlike the typical IDH (secondary) and EGFR (Primary) mutations found in other glioblastomas. It is important to identify this entity given its rapid progression, particularly poor prognosis and low median survival rates. The BRAF V600E mutation not only helps understand the tumor biology, but can form an important therapeutic target in times to come.