(emperipolesis) (Figure 1A and B). No cytologicatypia, mitosis, or necrosis was identified. Additionally, the lesion has a well-circumscribed interface with the duramater.

A diagnosis of Rosai Dorfman Disease was made.

On IHC, the histiocytes were immunoreactive for CD68 and S100(Figure 2A, 2B) while being negative for CD1a and langerin. The background lymphoid cells were labeled with either CD3 (T-cells) and CD20 (B-cells) (Figure 2C and D). The scattered plasma cells were positive for CD138, EMA and expressed both the light chains. The Ki-67 proliferation index was low (1%). Pancytokeratin, PR, HMB-45 and CD34 were negative.

In view of the intracranial location, paucity of plasma cells, and absence of storiform or other forms of fibrosis and obliterative phlebitis, an IgG4 and IgG IHC were not conducted in this case. Epithelial and melanocytic markers were negative. The morphology and immunophenotype confirmed the rendered diagnosis.

PATHOLOGIC DIAGNOSIS: MENINGEAL ROSAI-DORFMAN DISEASE

DISCUSSION:

Rosai Dorfman Disease (RDD) was first described by Destombes in 1965 and was recognized as a distinct clinicopathologic entity by Rosai and Dorfman in 1969. It is considered to be a reactive process, however, a subset of RDD has a clonal nature as kinase mutations have been described in nodal and extranodal RDDs. The mutations include ARAF, MAP2K1, NRAS, and KRAS. BRAF V600E mutation (exon 15) has been described in RDDs. Infectious agent-related pathogenesis has also been suggested which is either because of immunodeficiency or autoimmunity.

The patients with meningeal RDD of the brain are within an age group of 6 years to 79 years with male preponderance. The clinical manifestations range from headache, seizures to non-specific symptoms of meningeal irritation, raised intracranial pressure and focal neurologic deficits.