Rosai-Dorfman disease

Sinus histiocytosis or Rosai-Dorfman disease (RDD) is a rare but well-recognized disorder characterized by an unusual proliferation of histiocytic cells. Intracranial localization is a rare manifestation of RDD.

Etiology

Rosai-Dorfman disease (RDD) is a rare disease that can be triggered by either viral or bacterial infection. Several parts of the body can be involved, from the CNS to the pelvic regions had been reported.

Diagnosis

Conventional MRI, combined with diffusion-weighted imaging and ADC mapping, is an important diagnostic tool in evaluating RDD patients. An accurate diagnosis of RDD should consider the clinical features, imaging characteristics, and the pathological findings [1].

FDG-PET/CT image of a cystic central nervous system Rosai-Dorfman disease [2].

Differential diagnosis

Meningioma.

Rosai-Dorfman disease: especially if extracranial lesions are also identified. Usually in young adults. Isolated intracranial involvement is rare. MRI: duralbased enhancing mass with signal characteristics similar to meningioma, may have dural tail. Most common intracranial locations: cerebral convexities, parasagittal, suprasellar, cavernous sinus. Pathology: dense fibrocollagenous connective tissue with spindle cells and lymphocytic infiltration, stains for CD68 & S-100. Histiocytic proliferation without malignancy. Foamy histiocytes are characteristic. Surgery and immunosuppressive therapy not effective. Low-dose XRT may be the best option.

Some case reports highlights the necessity to consider Rosai-Dorfman disease as a potential posterior fossa tumor differential diagnosis and/or intraventricular tumor.

Treatment

At present, there is a serious lack of guidelines as to how to treat cases of RDD involving the spine.

Current trends show that surgery remains the first method of choice to cure this disease, but in refractory or recurrent RDD, repeat surgery cannot guarantee total resection. Under such circumstances, adjuvant therapy can be very useful.

Case series

A pooled cohort of 124 cases (81 male and 43 female), with a mean age of 39.7 years, included 11 cases from our cohort and 113 cases from 80 prior studies. Twenty-nine patients (23.4%) had multiple lesions. Seventy-four patients (59.7%) experienced gross total resection (GTR), 50 patients (40.3%) had non-GTR, 15 patients (12.1%) received postoperative adjuvant radiation, and 23 patients (18.5%) received postoperative steroids. A multivariate Cox regression revealed that GTR (HR = 4.52; 95% CI 1.21-16.86; p = 0.025) significantly improved PFS, and multiple lesions (p = 0.060) tended to increase the hazard of recurrence. Neither radiation (p = 0.258) nor steroids (p = 0.386) were associated with PFS. The overall PFS at 3, 5, and 10 years in the pooled cohort was 88.4%, 79.4%, and 70.6%, respectively. The PFS at 5 and 10 years in patients with GTR was 85.4% and 85.4%, respectively, which was 71.5% and 35.8%, respectively, in patients without GTR. Gross total resection significantly improved PFS and was recommended for PIRDD. Radiation and steroids were sometimes empirically administered for residual, multiple, or recurrent PIRDD, but the effectiveness remained arguable and required further investigation.Systematic review registration number: CRD42020151294 [3)]

Case reports

Rosai-Dorfman disease case reports.

1)
Cheng X, Cheng JL, Gao AK. A Study on Clinical Characteristics and Magnetic Resonance Imaging Manifestations on Systemic Rosai-Dorfman Disease. Chin Med J (Engl). 2018 Feb 20;131(4):440-447. doi: 10.4103/0366-6999.225053. PubMed PMID: 29451149; PubMed Central PMCID: PMC5830829.
2)
Kong Z, Wang Y, Ma W, Cheng X. FDG-PET/CT image of a cystic central nervous system Rosai-Dorfman disease. Eur J Nucl Med Mol Imaging. 2020 Jan 3. doi: 10.1007/s00259-019-04671-3. [Epub ahead of print] PubMed PMID: 31901102.
3)
Zhang GJ, Ma XJ, Zhang YP, Hao LF, Wang L, Zhang JT, Wu Z, Li D. Surgical management and outcome of primary intracranial Rosai-Dorfman disease: a single-institute experience and pooled analysis of individual patient data. Neurosurg Rev. 2023 Mar 27;46(1):76. doi: 10.1007/s10143-023-01983-9. PMID: 36967440.