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Intraventricular presentation of Rosai–Dorfman disease: a case report with review of literature

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Abstract

Background Rosai–Dorfman disease (RDD)/sinus lymphohistiocytosis is a rare benign lymphoproliferative disorder. Only 25% show extra-nodal manifestation, only 5% are seen involving the CNS, with intraventricular manifestation rarely reported. Our aim was to highlight important imaging features which would be useful in considering this entity as one of the differentials while encountering this rare entity.

Case description We present a case of a 34-year-old female with complaints of headache, altered behavior and visual disturbances. MRI brain showed T2 hypointense lesion arising from the left choroid plexus with dense homogenous enhancement, with multiple additional extra-axial dural-based lesions and a small lesion involving right choroid plexus. Left parietal craniotomy was done, and the lesion was excised. Histopathology showed large foamy macrophages in eosinophilic background, with lymphophagocytosis (emperipolesis), confirming the diagnosis of Rosai–Dorfman disease.

Conclusions Intraventricular Rosai–Dorfman disease is a rare entity. Imaging features of T2 hypointense homogenously enhancing lesion with blooming on GRE, without features of calcification or hemorrhage, may be helpful in prompting adequate histopathologic evaluation.

Keywords Rosai–Dorfman disease, Intraventricular lesions, Lymphohistiocytosis

Background

Rosai–Dorfman disease (RDD)/sinus lymphohistiocytosis is a rare benign lymphoproliferative disorder usually presenting in the first two decades of life. The aetiology remains unknown, and theories proposed include increased activation of suppressor macrophages, Epstein–Barr virus and HIV infection [1].

The disease presents usually as painless cervical lymphadenopathy in 95% of the cases with only 25% of cases showing extra-nodal manifestation, involving skin, respiratory tract, mucosa or soft tissues. Central nervous system involvement in Rosai–Dorfman disease (CNS-RDD) is noted in only 5% of the cases. It usually presents as dural masses mimicking meningioma with a broad base and dural tail. Rosai–Dorfman disease can be misdiagnosed with different histopathological types of meningioma and intracranial solitary fibrous tumors. Very few cases have been reported as non-dural/intraventricular/intramedullary lesions [2].

Confirmation of diagnosis is usually made with histopathological assessment where the cells show

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emperipolesis with large hyperchromatic histiocytes. Immunochemistry is definitive where CD68 and S100 positivity is observed with negative CD1a ruling out Langerhans cell histiocytosis, and other mimics including meningiomas and solitary fibrous tumors [3].

Majority of cases undergo spontaneous resolution. Complete surgical excision is the preferred option as it reduces the neurological symptoms and also serves for diagnostic purposes. If the lesion persists even after excision, chemotherapy is recommended. Some trials also suggest steroids for regression of lesion; however, there are no reliable data sources regarding treatment regimens owing to the rare nature of the lesion [4].

Our aim was to highlight important imaging features which would be useful in considering this entity as one of the differentials while encountering this rare entity.

Case presentation

We present the case of a 34-year-old female with complaints of headache since 7–8 months, memory impairment, behavioral changes and visual disturbances since 3 months. There was no history of associated vomiting,

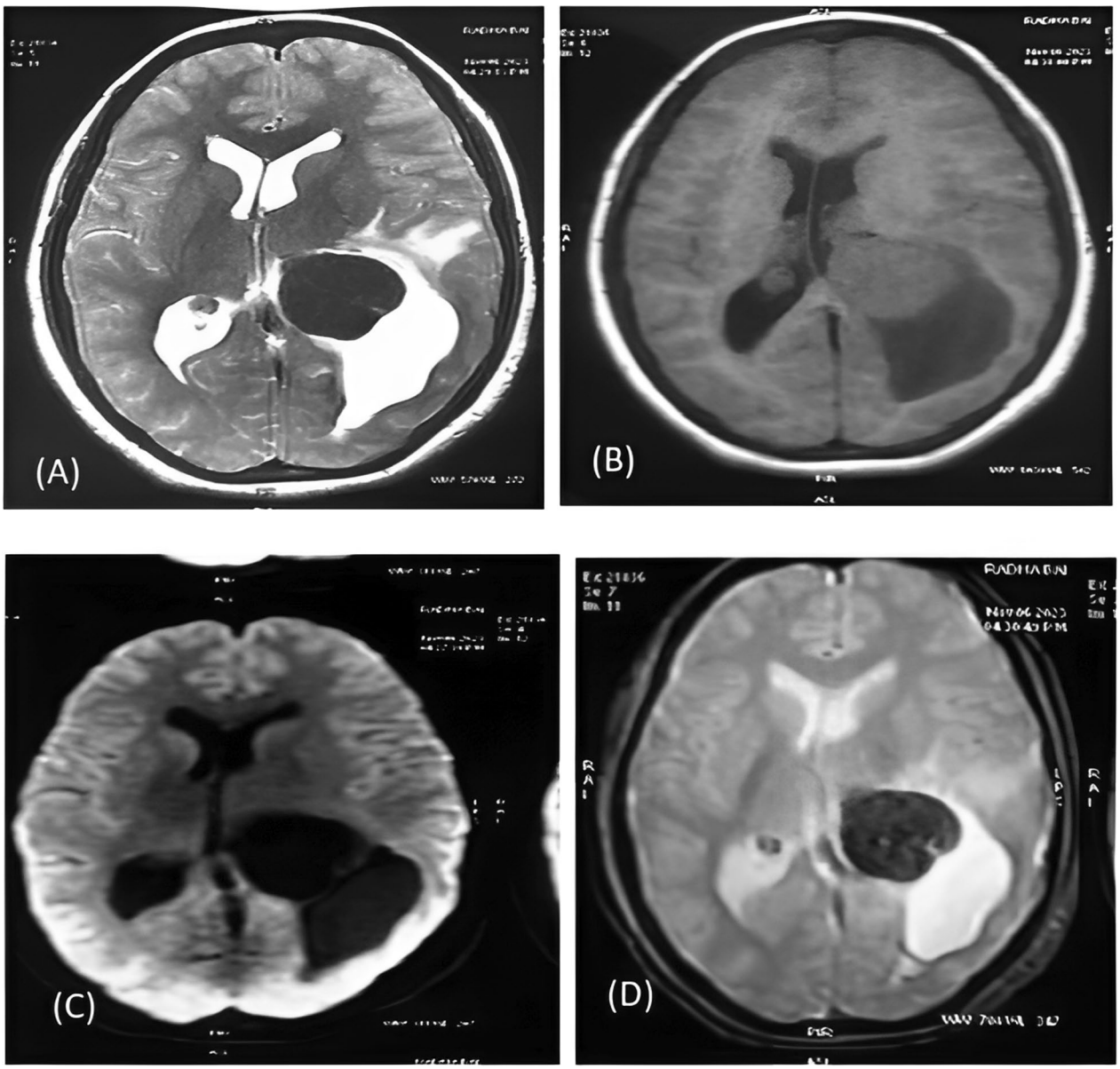

Fig. 1 T2 W image (**A**) shows markedly T2 hypointense lesion in left lateral ventricle atrium with dilatation of left lateral ventricle. The lesion is isointense on T1 (**B**) and does not show diffusion restriction (**C**). The lesion shows blooming on GRE image (**D**)

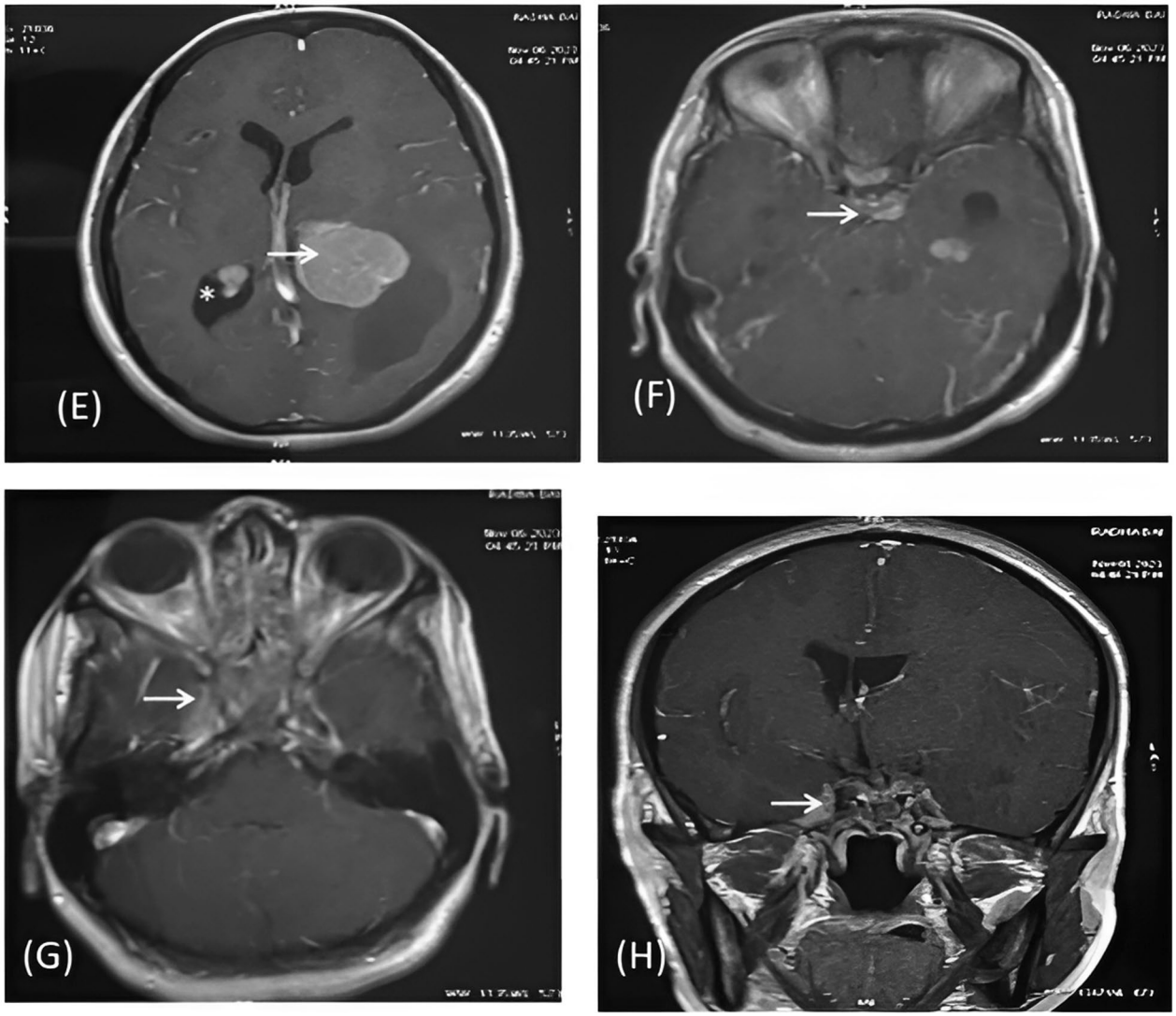

Fig. 2 Post-contrast T1 W axial image (**E**) shows enhancement of lesion in left lateral ventricle (arrow) with another enhancing lesion noted in right lateral ventricle glomus (*). Post-contrast T1 W axial image (**F**) shows extra-axial dural-based enhancing lesions in suprasellar cistern. Additionally dural thickening and pachymeningeal enhancement noted involving right cavernous sinuses (**G**, **H**)—arrows

seizures or loss of consciousness. She did not have any previous history of tuberculosis/granulomatous disorder, with no family history of malignancy/granulomatous disorder. On examination, there were no pallor, icterus, lymphadenopathy, pedal edema, cyanosis or clubbing. She was conscious and oriented, with Medical Research Council (MRC) scale power of 4/5 in right upper and lower limb. Her higher mental functions and cranial nerves examination were normal. The cerebellar examination was normal. Her blood workup including complete blood counts, erythrocyte sedimentation rate, liver and renal function tests was normal.

Magnetic resonance imaging (MRI) brain was performed on 1.5 T MRI machine. Routine sequences of brain were obtained including (T1-weighted image, T2-weighted image, fluid-attenuated inversion recovery (FLAIR), diffusion-weighted imaging (DWI), gradient echo (GRE) with post-contrast ultrafast spoiled gradient echo 3D sequence. MRI revealed a lesion measuring 3.6 × 3.8 × 5.1 cm in left lateral ventricle, which was isointense on T1 and hypointense on T2 imaging with resultant dilatation of the trigone, occipital and temporal horns of the left lateral ventricle with associated intraventricular hemorrhage and periventricular edema. The lesion showed homogenous enhancement on contrast administration.

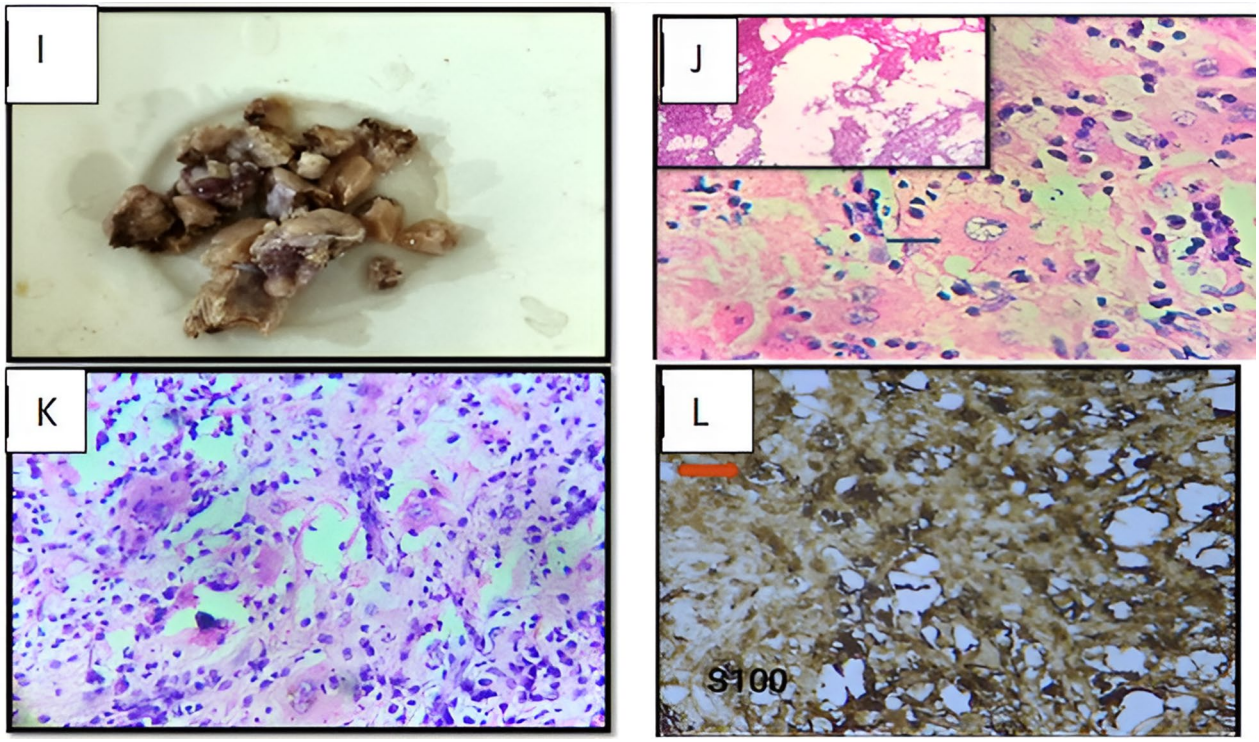

Fig. 3 Multiple grayish white firm tissue pieces (**I**), microscopy: (HE 40X) (**J**)Section shows diffuse proliferation of large pale histiocytes (Arrow) in dense fibrotic stroma admixed with inflammatory infiltrate comprising of numerous plasma cells, lymphocytes and neutrophils. Inset (PAP 40X) shows squash cytology smears of same case showing dense fibrous tissue along with few inflammatory cells. HE 40X) Section shows histiocytes (arrow) showing emperipolesis (**K**) of intact lymphocytes, plasma cells and neutrophils. IHC shows positivity for S100 (**L**)

Additionally similar characteristic enhancing dural-based lesions were also noted in the suprasellar cistern and right cavernous sinus. Another small similar lesion was also noted in right lateral ventricle trigone. The lesions also showed mild blooming on GRE, no diffusion restriction was noted. A preliminary diagnosis of a neoplastic lesion was made and it was planned for excision. Imaging features are shown in Figs. 1 and 2.

The patient underwent a left parietal craniotomy for removal of the intraventricular lesion with placement of extra-ventricular drain. Intraoperative findings showed a grayish, firm to hard lesion arising from the choroid plexus with moderate vascularity with well-defined planes occupying the atrial region.

On histopathological assessment, there were multiple grayish white firm tissue pieces measuring 5 × 4 × 1 cm in size. H and E staining under 40x showed diffuse proliferation of large histiocytes admixed with fibrotic stroma and inflammatory infiltrate of plasma cells, lymphocytes and neutrophils (Fig. 3). Histiocytes show emperipolesis of intact lymphocytes, neutrophils and plasma cells without atypical mitotic figures or angiogenesis. Immunohistochemistry of the biopsy specimen revealed S100 positivity. A definitive diagnosis was made based on the histopathology and immunohistochemistry as histiocytic tumor—Rosai–Dorfman disease.

Discussion

Rosai–Dorfman disease is a rare non-neoplastic and non-Langerhans cell proliferation disorder of the histiocytes with central nervous system involvement (CNS-RDD) being rare (less than 5% cases), and less than 150 cases of CNS-RDD have been described[5]. In CNS-RDD most cases which have been described are dural-based lesions and intraventricular involvement is quite rare. We did a review of previous literature, and noted that there were a total of 8 cases showing intraventricular involvement and among them 5 cases had isolated intraventricular involvement (Table 1). From the table we can also observe that intracranial involvement by RDD can occur as an isolated entity and may not be associated with nodal/extra-cranial manifestations of the disease.

Radiological appearances of previously described cases of Rosai–Dorfman disease are similar to what we observed in our case. The previously described intracranial lesions of CNS- RDD are predominantly dural-based

Table 1 Table summarizing previously reported cases of intraventricular RDD, extra-ventricular features (if present), Immunohistochemical markers and histopathological features

Author and year	Age and sex	Intraventricular lesion location	Extra-ventricular involvement	Histopathology	IHC + for
Patwardhan et al., 2018 [5]	40y, female	Rt lateral ventricle—occipital horn, T1 isointense, T2 hypointense, Gd enhancement +	Nil	Emperipolesis of lymphocytes, psammoma bodies	CD 68, S100
Friconnet et al., 2021 [9]	30y, male	Lesion protruding into the fourth ventricle, isointense T1, T2 hypointense, Dota +, ADC ~ 1, perfusion normal, NAA, lipid peak	Nil	Emperipolesis, Neovascularization with a dense peri-vascular lymphocytic infiltrate extending inside the histiocytic proliferation	CD68 +, CD163 +
Morandi et al., 2000 [10]	22y,female	Marked homogeneous enhancing lesion in the floor of the fourth ventricle	Nil	Lymphophagocytosis +	CD68 +, CD163 +, CD1a negative
Jamali et al., 2022 [11]	8y male	Multiple large intraventricular masses	Nil	Histiocytes mixed with lymphocytes and plasma cells	CD68 +, CD163 +, CD1a negative
Ludemann et al., 2015 [12]	2y 10 month	Axial enhancing lesion with ventricular spreading mainly to the left occipital horn and bilateral frontal periventricular infiltration	Nil	Chronic inflammatory cells and histiocytes that showed emperipolesis	Nil
Luo et al., 2017 [13]	41y male	T1 isointense or slightly hyperintense signal, DWI images showed hypointense signal, and all the lesions showed homogeneous enhancement in contrast-enhanced scanning	Multiple ICSOL, also involving Lt parasellar and CP angle region	Lymphocyte and histiocyte predominance	CD68, S100 positivity
Catalucci et al., 2012 [14]	57y, male	Single lesion seen involving Rt lateral ventricle	Multiple dural-based enhancing lesions	Lymphocyte and histiocyte predominance	CD68 +, CD163 +, CD1a negative
Zhang et al., 2022 [2]	14y, female	Lesion involving left frontal lobe and lateral ventricle showing homogenous enhancement, peripheral edema	Left frontal lobe involvement	Polymorphous lymphohistiocyte infiltrate	CD 68, S100 positivity

lesions, mimicking meningioma, showing a broad base with dural tail, most commonly involving the suprasellar region, convexity, parasagittal region, cavernous sinus, petroclival region and cerebellum. Rarely, the lesions presented with dural encroachment and pachymeningitis like features [5]. In our cases, also there were similar dural-based lesions in suprasellar cistern with dural thickening in parasellar regions.

The signal characteristics of these previously described lesions were isointense on T1 and iso–hypointense on T2 showing homogenous/heterogenous dense enhancement on contrast administration, similar to what we observed in our case.

The imaging differentials for intracranial RDD include meningioma, metastasis, lymphoma/leukemia, granulomatous diseases. Meningioma remains the most important differential. However, presence of T2 hypointense signal can point toward RDD as a possibility as most meningiomas are T2 hyperintense/isointense. Additionally, presence of blooming on gradient echo imaging (GRE) without evidence of calcification/hemorrhage on Computed Tomography (CT) can be an important clue for possible preoperative imaging diagnosis of RDD as compared to the rest of differentials (2). Lobulated margins and irregularly thickened meningeal wall extending into brain parenchyma—pseudopodium sign have been described as an important imaging signs favoring intracranial RDD, in addition to T2 hypointensity and blooming on GRE/susceptibility-weighted imaging (SWI) [2, 6].

Advanced Imaging including MR spectroscopy may show a non-specific choline peak. Perfusion imaging may show no significant alteration in cerebral blood flow/volume or may show decreased perfusion [6]. However one recent case report has described increased perfusion indicating increased vascularization with positive markers for CD34 and CD31 antibodies [7]. Few cases reports have described high fractional anisotropy (FA) and low apparent diffusion coefficient (ADC) values [6].

Microscopic features of RDD include large macrophages with foamy eosinophilic cytoplasm, showing lymphophagocytosis with preserved architecture of the engulfed cell, also known as emperipolesis. This feature is highly specific and suggestive of Rosai–Dorfman disease. This diagnosis is confirmed by the immunohistochemical profile of the histiocytes: positive for S-100 protein, CD68, CD163, and negative for CD1a [8].

Conclusions

Intraventricular Rosai–Dorfman disease is rare and requires multidisciplinary approach for diagnosis and management. Imaging features such as T2 hypointensity, blooming on GRE/SWI (without hemorrhage/calcification on CT), lobulated margins sign and pseudopodium sign help as important imaging features favoring intracranial RDD.

Abbreviations

CNS-RDD	Central nervous system involvement in Rosai–Dorfman disease
CT	Computed tomography
DWI	Diffusion-weighted imaging
FLAIR	Fluid-attenuated inversion recovery
GRE	Gradient echo imaging
MRC	Medical Research Council
MRI	Magnetic resonance imaging
RDD	Rosai–Dorfman disease
SWI	Susceptibility-weighted imaging

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None.

Author contributions

Dr. Saranya Ravi and Dr. Nishtha Yadav contributed to the literature search, figures, data collection, data analysis, data interpretation, and writing. Dr. Diya Bajaj was involved in the literature search, figures, revision and editing of manuscript. Dr. Shailendra Ratre participated in the literature search, revision and editing of manuscript. Dr. Sonjjay Pande revised and edited the manuscript.

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Availability of data and materials

All data generated or analyzed during this study are included in this published article [and its supplementary information files].

Declarations

Ethics approval and consent to participate

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Consent for publication

It was taken from patient.

Competing interests

The authors declare that they have no competing interests.

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