BJR|Case Reports, 2024, **10(1)**, uaae001
https://doi.org/10.1093/bjrcr/uaae001
Advance access publication: 19 January 2024
Case Review

Racemose neurocysticercosis: a case series

Srishti Sharma, MD[1], Anuradha Singh, MD[2], Vinita Elizabeth Mani, DM[3], Sarvesh Chandra Mishra, MD[2], Pragya Chaturvedi, MD[2], Shweta Jain, MD[4], Sunil Kumar, MD[5], Neeraj Jain DMRD, DNB[2,*]

[1]Department of Radiodiagnosis, All India Institute of Medical Sciences, Patna 801507, India
[2]Department of Radiodiagnosis, SGPGIMS, Lucknow 226014, India
[3]Department of Neurology, SGPGIMS, Lucknow 226014, India
[4]Department of Pathology, SGPGIMS, Lucknow 226014, India
[5]Department of Radiology, Medanta Super Speciality Hospital, Lucknow 226030, India
*Corresponding author: Neeraj Jain, DMRD, DNB, Department of Radiodiagnosis, SGPGIMS, Raebareli Road, Lucknow 226014, India (neerajdmrd@gmail.com)

Abstract

Neurocysticercosis (NCC) is a common parasitic condition of the central nervous system in certain parts of the world. The racemose variety of NCC is distinct from the commonly seen parenchymal form. It frequently infiltrates the basal cisterns and Sylvian fissures. Imaging plays a vital role in the diagnosis; however, as their signal intensity is similar to cerebrospinal fluid and due to the absence of enhancement in most cases, imaging diagnosis is often difficult on the conventional MRI sequences. Here, we present five cases of racemose NCC to emphasize the importance of a heavily T2-weighted sequence (Fast Imaging Employing Steady-state Acquisition) sequence in the diagnosing this entity.

Keywords: neurocysticercosis; racemose NCC; FIESTA; MRI.

Introduction

Neurocysticercosis (NCC) is a common parasitic infection affecting the central nervous system (CNS) and constitutes a public health challenge for most developing countries.[1] It is caused by the larval form of the pork tapeworm Taenia solium. Depending on the site of involvement, they are classified into intraparenchymal, extra parenchymal (subarachnoid-cisternal, meningeal, intraventricular) and mixed types.[2] Among these, parenchymal involvement is the most common. As with other haematogenously transmitted diseases, it typically involves the grey-white matter junction, as parasites often lodge here due to the smaller calibre blood vessels.[3] If larvae lodge in the meninges or choroid plexus, the infection may involve the ventricular system and subarachnoid space. The subarachnoid or racemose NCC are frequently large-sized lesions as the subarachnoid space provides ample space to grow compared to the parenchymal variety. Furthermore, unlike parenchymal NCC, racemose NCC clinical presentation is variable, making their diagnosis extremely challenging.

MRI plays a crucial role in the diagnosis; however, considering that the signal intensity of the lesion is similar to cerebrospinal fluid (CSF) signal intensity on all conventional sequences and the lack of enhancement in most cases makes their prospective diagnosis quite tricky. Herein, we present a case series depicting the MRI findings of racemose NCC. We emphasize the importance of a heavily T2-weighted sequence (FIESTA—Fast Imaging Employing Steady-state Acquisition) in their diagnosis (Table 1).

Case 1

A 52-year-old female patient presented with a history of fever, headache, and vomiting for 2 months. Based on her symptoms and background, an initial diagnosis of CNS tuberculosis (TB) was suspected. Subsequently, a gene expert test for TB was negative, and CSF analysis revealed normal protein, sugar, and lactate levels.

T2-weighted MRI imaging (T2WI) of the brain showed expansion of the left prepontine cistern with the subtle presence of fluid-signal intensity cystic lesions, which extended posterior to the midbrain and displaced it towards the contralateral side (Figure 1A and B). Moderate hydrocephalus was also noted. The lesions were better delineated on FIESTA images with thin septations as additional findings; these septations were not seen on T2 (Figure 1C and D). No scolex was identified, and there was no post-contrast enhancement. Based on imaging findings, a diagnosis of racemose NCC was made. The patient was put on conservative medications and is currently on follow-up.

Case 2

A 10-year-old boy presented to our hospital with a history of frequent headaches and seizures for 3 months and visual disturbances for 4 days. MRI brain revealed a multiloculated cystic lesion in the pineal region extending into the third ventricle with resultant obstruction (Figure 2). It was hypointense on T1 (Figure 2A) with a thin rim of peripheral enhancement on the post-contrast study (Figure 2B). T2 and FIESTA images showed a characteristic multiloculated appearance (Figure 2C and D), classically called a "cluster of grapes" appearance. The lesion caused a mass effect on the tectum and thalami with the narrowing of the aqueduct of Sylvius.

Received: 2 June 2023; **Revised:** 20 September 2023; **Accepted:** 13 January 2024

Table 1. The demographics, clinical manifestations, and MRI findings of these cases.

Patient	Age/gender	Type and duration of symptoms	Lesion location	Internal septations (on FIESTA)	Scolex	Enhancement
1	52 years/F	Fever, headache, and vomiting—2 months	Prepontine cistern and the cisternal spaces along the midbrain	Present	Absent	None
2	10 years/M	Headache and seizures—2 months	Pineal region	Present	Absent	Yes
3	37 years/M	Headache and seizures—2 months	Under surface of the corpus callosum, roof of the third ventricle and basal cisterns	Present	Absent	Absent
4	56 years/M	Seizure disorder and slowness of gait—5 years	Ambient cistern and Sylvian fissures	Present	Absent	Absent
5	61 years/M	Headache and fever—2 days	Sylvian fissure	Present	Present	Present

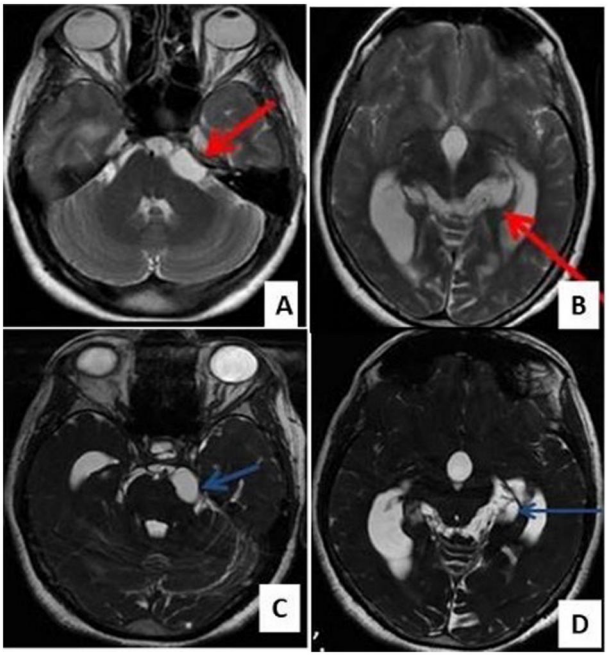

Figure 1. Axial T2-weighted images showing a CSF signal intensity cystic lesion in the prepontine cistern (A) and in the cisternal space along the midbrain (B). Axial FIESTA—Fast Imaging Employing Steady-state Acquisition images (C, D) aid better visualization of the lesions with thin internal septations.

Case 3

A 37-year-old male patient has a history of frequent headaches and recurrent seizures for 2 months. MRI revealed dilatation of the extra-axial CSF spaces with septations along the inferior aspect of the corpus callosum. The third ventricle roof was displaced posteriorly into the basal cisterns (Figure 3A and B). Moderate non-communicating hydrocephalus was also noted with the normal-sized fourth ventricle. The FIESTA sequence could depict the multiseptated nature of the lesion more clearly. Contrast enhancement and diffusion restriction were absent. The patient improved on conservative management and is currently on follow-up.

Case 4

A 56-year-old male patient came to our hospital with a history of seizures, gait impairment, incontinence, ataxia, and forgetfulness for 5 years. Based on the presence of hydrocephalus on the previous MRI, he was treated elsewhere on the line for CNS tuberculosis. However, despite 3 months of antitubercular treatment, he showed no signs of improvement.

A repeat MRI was done to look for an alternate diagnosis. It showed a dilated ambient cistern with indentation on the pons (Figure 4A). The lateral ventricles were markedly dilated along with dilatation of bilateral CSF spaces in the temporoparietal region with dominant involvement of Sylvian fissures (Figure 4B). On FIESTA MRI (Figure 4C and D), the lesion showed thin septations in the ambient cistern and Sylvian fissures, which strongly suggested the diagnosis of racemose NCC. Moderate hydrocephalus was also present. The patient was advised for VP shunt placement and is currently on follow-up.

Case 5

A 61-year-old male complaining of headaches for 3 days and fever for 2 days came to our outpatient department. He had a history of a right middle cerebral artery territory infarct in January 2020. Clinical examination was unremarkable except for a poor swallowing reflex. On T2W MRI, a hyperintense multilobulated cystic lesion was detected in the right Sylvian fissure (Figure 5A and B). It showed smooth peripheral enhancement (Figure 5C). Eccentric blooming focus representing scolex was seen on susceptibility-weighted imaging (SWI, Figure 5D). Scolex represent armed rostellum and body of cysticercus, as larvae progress through different stages scolex undergoes sequential changes and finally becomes calcified in calcified nodular stage. In addition to showing cystic lesions, the FIESTA image showed multiple thin septations in the rest of the Sylvian fissure, suggesting a more extensive involvement (Figure 5D and E). Based on imaging findings, a diagnosis of racemose NCC was suspected. He showed improvement on conservative treatment and presently is on follow-up.

Discussion

Cysticercosis is considered one of the common helminthic infections of the brain and a leading cause of acquired epilepsy worldwide.[4] Clinical manifestations of NCC are highly variable, with seizures being the most common presentation mode. There may be focal neurological deficits, intracranial hypertension, or cognitive decline. The onset of symptoms

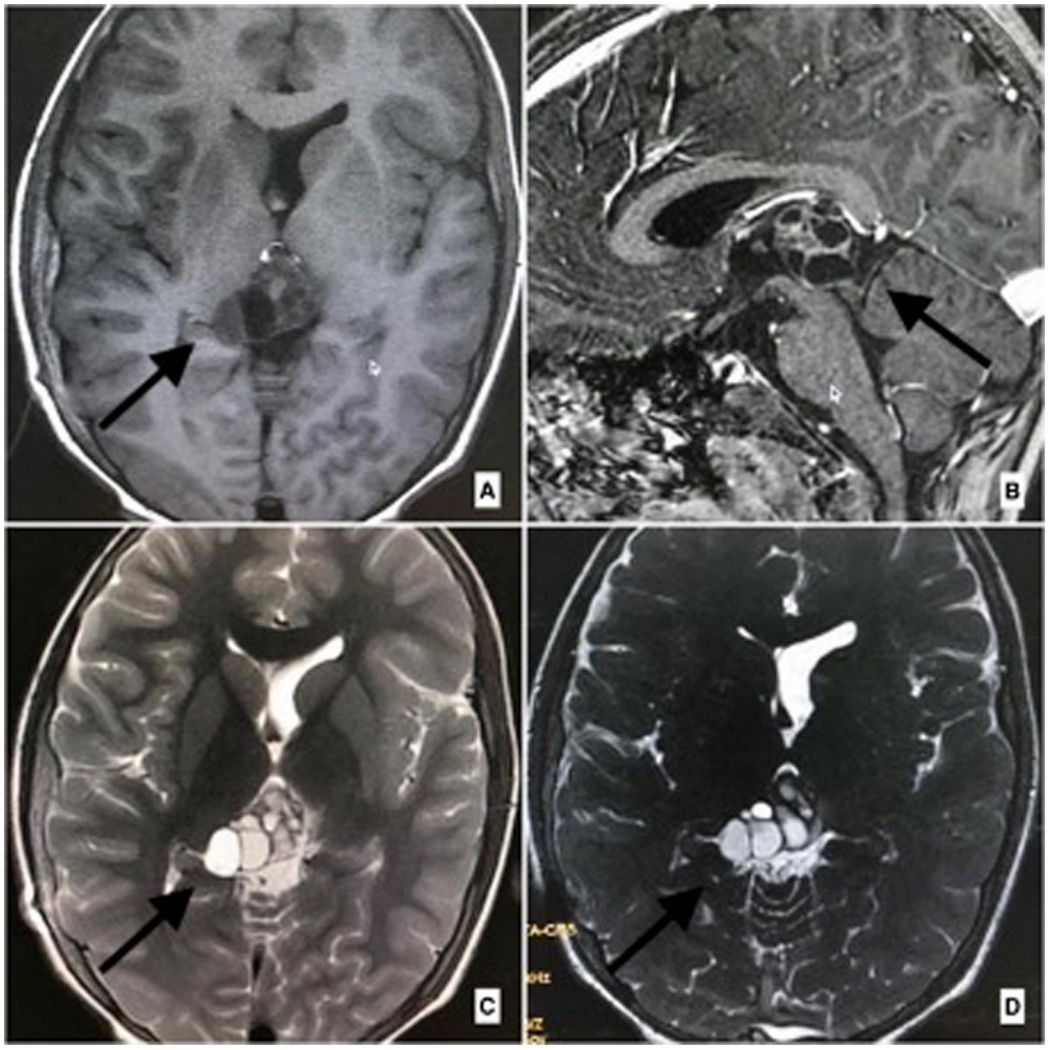

Figure 2. Axial T1-weighted image (A) showing a multiloculated cystic lesion in the pineal region with peripheral enhancement on the post-contrast sagittal image. (B) Axial T2 and FIESTA—Fast Imaging Employing Steady-state Acquisition images show a multiloculated cystic lesion in the pineal region (arrow).

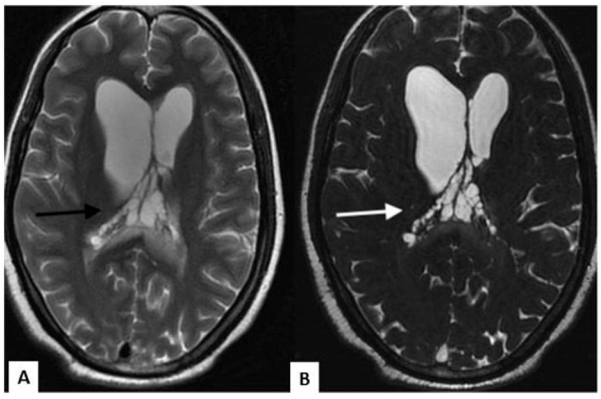

Figure 3. Axial T2-weighted image (A) shows a multiloculated cystic lesion along the roof of the third ventricle with further extension along the undersurface of the corpus callosum. The axial FIESTA—Fast Imaging Employing Steady-state Acquisition (B) image shows a better definition with thin internal septations.

after the initial infection is variable, and it depends on the cysts' number, size, location, and the host's immune response to the parasite.[5]

While the brain parenchyma is often considered the most common location for NCC, a few studies suggest that the subarachnoid-cisternal location may be more prevalent.[6-9] These studies argue that parasites' "parenchymal" location, as described in cross-sectional imaging, may represent subarachnoid cysticercosis located in deep sulci or in perforating branches of perivascular spaces.[10] Nonetheless, the distinction between parenchymal and extraparenchymal location has significant clinical and prognostic implications. The parenchymal forms behave differently clinically and are more responsive to anthelminthic medications than the subarachnoid forms.

Escobar classified the parenchymal NCC into four pathological stages: vesicular, colloidal vesicular, granular nodular, and calcified nodular.[11] Imaging findings and clinical presentation at each stage reflect underlying changes in the disease process and host response.[12] Patient presentation ranges from asymptomatic to encephalitis, depending on the disease stage.[3,13-15] The importance of calcified granulomas with perilesional oedema as foci of seizures and epilepsy in populations where cysticercosis is endemic is newly recognized. It indicates that treatment with anti-inflammatory agents could control or prevent epilepsy in these patients.[16]

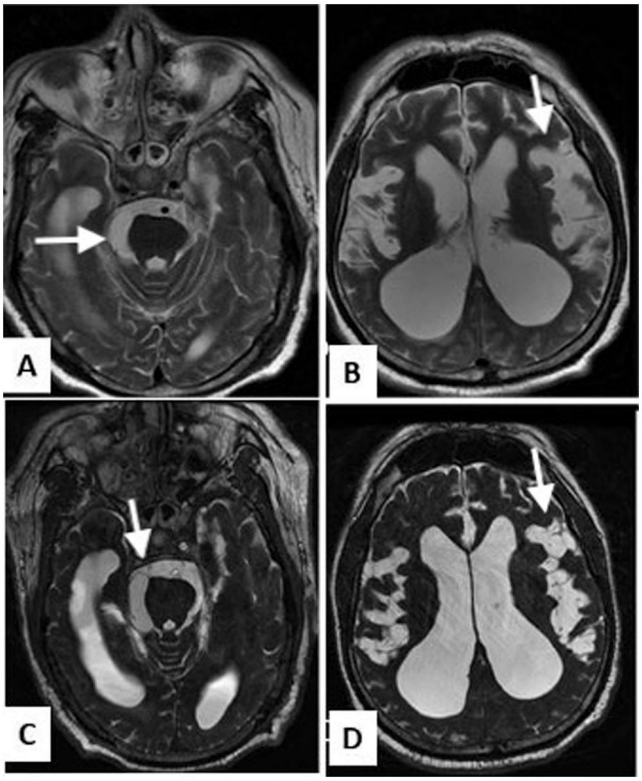

Figure 4. Axial T2-weighted image (A) shows dilated ambient cistern with an indentation on the pons. Axial T2 image (B) shows dilated lateral ventricles with widened sulcal spaces in the temporoparietal region. Axial FIESTA—Fast Imaging Employing Steady-state Acquisition image (C) revealed a cystic lesion in the ambient cistern, causing indentation on the pons. In addition to the widening of sulcal spaces, the axial FIESTA—Fast Imaging Employing Steady-state Acquisition image (D) shows thin septations.

Conversely, racemose NCC develops due to the segmentation of cysticercosis cellulosae with the budding of new cysts in clusters. These cysts grow slowly and expand in CSF spaces such as ambient cistern, quadrigeminal cistern, and Sylvian fissure. They are significantly larger than the parenchymal forms owing to the ample availability of the CSF spaces (Figures 1-5). They lack the usual evolutionary stages of the parenchymal NCC. Scolex, which represents rostellum and body of a larvae, degenerates as cysts progress and finally becomes calcified in the calcified nodular stage. A scolex is considered as a hallmark feature of parenchymal NCC and is frequently absent in racemose NCC as it degenerates.[17] Additionally, calcification, which is seen in the later stages of parenchymal NCC, and peripheral enhancement in vesicular and colloid vesicular stages of parenchymal NCC are also not seen in racemose NCC. However, sometimes, the racemose NCC can incite inflammation of surrounding tissues and may show enhancement (Figures 2 and 5). This inflammatory response can also cause vasculitis of the basal perforators, leading to infarcts.[18,19]

On conventional MRI, these lesions appear isointense to the CSF on all pulse sequences. Occasionally, they may show hyperintense signal on T1WI and FLAIR compared to the CSF due to their protein content.

When the ventricular system is involved, these cysts are frequently mobile and can cause acute obstructive hydrocephalus. Usually, they migrate from the lateral ventricle to the third ventricle and the fourth ventricle through the sylvius aqueduct.[20] It often manifests as a solitary intraventricular cyst, the fourth ventricle being the most common location. The cyst wall may occasionally be seen as a thin linear structure on T2-weighted imaging or demonstrate contrast enhancement. They can cause ventricular deformity, enlargement, and hydrocephalus.

Serological tests (enzyme-linked immunoelectrotransfer blot assay, enzyme-linked immunosorbent assay) that detect anticysticercal antibodies in serum or CSF inherently lack specificity or sensitivity, thereby precluding their reliability in isolation.[6] MRI is the primary diagnostic modality used for the detection of cysticercus lesions and their secondary complications.

A racemose NCC is more challenging to diagnose than a parenchymal NCC. These lesions are often missed on routine MRI of the brain owing to their signal, which is isointense to CSF and absence of contrast enhancement of the cyst wall.[21] In the absence of a high index of suspicion, these lesions can be simply dismissed as communicating hydrocephalus (if intraventricular) or mistaken for other extraaxial cystic lesions (arachnoid cysts, epidermoid cysts, etc.) owing to their multicystic appearance.[7,8] Hence, diagnosing an NCC, especially the racemose variant, is a multidisciplinary approach that involves the correct interpretation of the symptomatology, neuroimaging, and serology in the proper epidemiological context.

Fast Imaging Employing Steady-state Acquisition is a heavily 3D T2WI high-resolution gradient-echo sequence routinely used to evaluate the inner and middle ear's structures and cerebellopontine angle lesions. In this sequence, the residual transverse magnetization is refocussed to achieve a steady magnitude of transverse and longitudinal magnetization following a few repetition times (TRs). It significantly increases the conspicuity of lesions in cisterns, sulci, and intraventricular spaces. The millimetre-thin sections, short TE (limited signal loss resulting from magnetic susceptibility effects), and low flip angle (limited T1WI) allow superb spatial resolution and signal-to-noise ratio within a clinically feasible acquisition time.[22] CISS (constructive interference in steady state) and DRIVE (Driven Equilibrium Radio Frequency Reset Pulse) are similar sequences used by other MRI manufactures.

As opposed to the T2W sequence, tissue contrast in FIESTA sequences is determined by the T2/T1 ratio of the tissues. It accentuates the contrast between the CSF and the surrounding structures, including the lesion. In comparison with conventional FLAIR or T2 sequences, this sequence is significantly better at detecting NCC lesions in intraventricular and extraventricular CSF spaces by allowing better visualization of the cyst wall, internal septations, and internal scolex.[23]

As previously discussed, scolex is frequently absent in racemose NCC. However, anecdotal case reports have described the presence of scolex on SWI sequences. This case series also shows the presence of scolex in one patient.[21]

The clinical presentation of racemose cysts is pleomorphic and quite different from parenchymal cysticercosis. Studies suggest that classification of the racemose NCC into meningeal, intraventricular, and cisternal forms can help us predict the clinical progression in these patients.[10] The meningeal form refers to cases where the lesions are typically seen in the cerebral convexities. This form of racemose NCC results in raised intracranial pressure by causing meningitis and

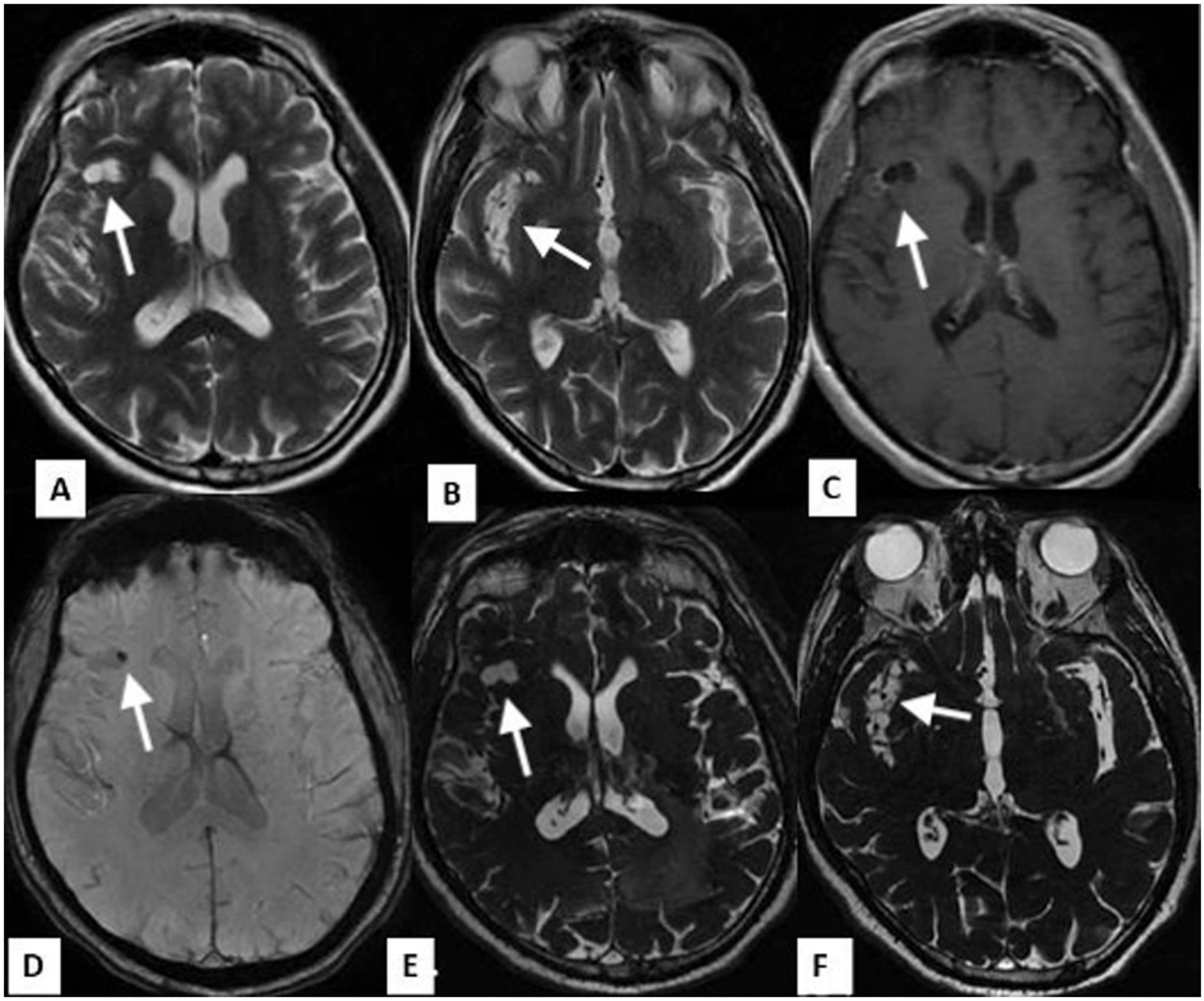

Figure 5. Axial T2-weighted images show a cystic lesion in the right Sylvian fissure (A) with asymmetric prominence of the right Sylvian fissure (B). Axial T1 image (C) showing smooth peripheral enhancement. The susceptibility-weighted image (D) shows a blooming focus in the cystic lesion, representing a scolex. Axial FIESTA—Fast Imaging Employing Steady-state Acquisition (E, F) images better visualize the cystic lesion and additionally demonstrate multiple thin septations in the widened right Sylvian fissure.

adhesions. It can also present with focal neurological deficit due to entrapment of the cranial nerves in the inflammatory exudate.[14] The intraventricular form refers to cysticercus within the ventricular system. The cisternal form refers to racemose lesions within the basal cisterns. These forms can occlude the CSF pathway causing acute episodes of ventriculomegaly and a mass effect. They can also cause hydrocephalous as a sequalae of ependymitis resulting from the death of larvae.[24] The blockage of CSF pathways resulting in raised intracranial tension and hydrocephalous is an important determinant in the prognosis of these patients. We propose the following imaging-based classification system to help prognosticate these patients and decide the management.

- Type 1A—Meningeal form without hydrocephalous.
- Type 1B—Meningeal form with hydrocephalous.
- Type 2A—Intraventricular form without hydrocephalous
- Type 2B—Intraventricular form with hydrocephalous.
- Type 3A—Cisternal form without hydrocephalous.
- Type 3B—Cisternal form with hydrocephalous.
- Type 4A—Present in more than 1 location without hydrocephalous
- Type 4B—Present in more than 1 location with hydrocephalous
- The suffix + P to be used in the presence of associated parenchymal NCC.

The common differential diagnosis for subarachnoid NCC includes arachnoid cysts, neuroglial cysts, and epidermoid tumours. Like NCC, arachnoid cysts are located in the subarachnoid space. However, they are unilocular and devoid of septations and scolex. The neuroglial cyst can be intra- or intraparenchymal, with the former being more common. Like an arachnoid cyst, they lack septations. Ruptured hydatid cysts can also be a differential for racemose NCC, especially in areas with an endemic population. However, as hydatid cysts are primarily intraparenchymal before rupture, there may be some tell-tale evidence of their intraparenchymal location before they rupture into extraaxial CSF spaces. Moreover, positive hydatid serology will further be supporting evidence.[7,8] However, for the detection of all of these

cystic lesions in the CSF space, the FIESTA sequence is quite promising since it provides excellent contrast between the lesions and the CSF space.

Conclusion

Racemose NCC should be considered in the differential diagnosis of multiloculated, clustered cystic lesions involving extraaxial CSF spaces.

We recommend routine inclusion of the FIESTA sequence in the evaluation of cases of obstructive hydrocephalus or any cystic lesion involving the extraaxial CSF spaces in an endemic region. Not only can this sequence demonstrate septations within the lesion as involving the subarachnoid spaces, but it may also help in identification of the cyst in an intraventricular location that otherwise remains occult on other MRI sequences.

Learning points

- Racemose NCC is distinct from parenchymal variant. It infiltrates CSF spaces like basal cistern, Sylvian fissure, and around brain stem.
- It often lacks scolex and does not show enhancement.
- It should be considered in the differential diagnosis of multiloculated cystic lesions in extraaxial CSF spaces
- A heavily 3D T2WI high-resolution gradient-echo sequence accentuates the contrast between the CSF and the surrounding structures, thereby improving lesion visibility.

Funding

None declared.

Conflicts of interest

None declared

Informed consent statement

Written informed consent was obtained from the patients for publication of this case report, including accompanying images.

References

1. Bansal R, Gupta M, Bharat V, et al. Racemose variant of neurocysticercosis: a case report. *J Parasit Dis*. 2016;40(2):546-549.
2. Thakur K, Zunt J. Neurologic parasitic infections in immigrants and travelers. *Semin Neurol*. 2011;31(3):231-244.
3. Lerner A, Shiroishi MS, Zee CS, et al. Imaging of neurocysticercosis. *Neuroimaging Clin N Am*. 2012;22(4):659-676.
4. Del Brutto OH. Neurocysticercosis. *Handb Clin Neurol*. 2014; 121:1445-1459.
5. Del Brutto OH. Neurocysticercosis: a review. *ScientificWorldJournal*. 2012;2012:159821.
6. Bustos JA, Coyle CM. Brain calcification because of neurocysticercosis: a vast field to be explored. *Curr Opin Infect Dis*. 2020;33 (5):334-338.
7. Marcin Sierra M, Arroyo M, Cadena Torres M, et al. Extraparenchymal neurocysticercosis: demographic, clinicoradiological, and inflammatory features. *PLoS Negl Trop Dis*. 2017;11 (6):e0005646.
8. White AC, Jr, Coyle CM, Rajshekhar V, et al. Diagnosis and treatment of neurocysticercosis: 2017 clinical practice guidelines by the Infectious Diseases Society of America (IDSA) and the American Society of Tropical Medicine and Hygiene (ASTMH). *Clin Infect Dis*. 2018;66(8):e49-e75.
9. Kimura-Hayama ET, Higuera JA, Corona-Cedillo R, et al. Neurocysticercosis: radio-logic-pathologic correlation. *Radiographics*. 2010;30(6):1705-1719.
10. Mahale RR, Mehta A, Rangasetty S. Extraparenchymal (racemose) neurocysticercosis and its multitude manifestations: a comprehensive review. *J Clin Neurol*. 2015;11(3):203-211.
11. Escobar A, Aruffo C, Cruz-Sánchez F, et al. Neuropathologic findings in neurocysti-cercosis. *Arch Neurobiol (Madr)*. 1985;48 (3):151-156.
12. Zee CS, Segall HD, Boswell W, et al. MR imaging of Neurocysticercosis. *J Comput Assist Tomogr*. 1988;12 (6):927-934.
13. Venkat B, Aggarwal N, Makhaik S, et al. A comprehensive review of imaging findings in human cysticercosis. *Jpn J Radiol*. 2016;34 (4):241-257.
14. Sotelo J, Escobedo F, Penagos P. Albendazole vs praziquantel for therapy for neuro-cysticercosis. A controlled trial. *Arch Neurol*. 1988;45(5):532-534.
15. Hingwala D, Chatterjee S, Kesavadas C, et al. Applications of 3D CISS sequence for problem solving in neuroimaging. *Indian J Radiol Imaging*. 2011;21(2):90-97.
16. Nash TE, Garcia HH. Diagnosis and treatment of Neurocysticercosis. *Nat Rev Neurol*. 2011;7(10):584-594.
17. Garcia HH, Del Brutto OH. Taenia solium cysticercosis. *Infect Dis Clin North Am*. 2000;14(1):97-119, ix.
18. Levy AS, Lillehei KO, Rubinstein D, et al. Subaracnoid neurocysticercosis with occlusion of the major intracranial arteries: case report. *Neurosurgery*. 1995;36(1):183-188.
19. Hauptman JS, Hinrichs C, Mele C, et al. Radiologic manifestations of intraventricular and subarachnoid racemose neurocysticercosis. *Emerg Radiol*. 2005;11(3):153-157.
20. Govindappa SS, Narayanan JP, Krishnamoorthy VM, et al. Improved detection of in-traventricular cysticercal cysts with the use of three-dimensional constructive interfer-ence in steady state MR sequences. *AJNR Am J Neuroradiol*. 2000;21:679-684.
21. Neyaz Z, Patwari SS, Paliwal VK. Role of FIESTA and SWAN sequences in diagnosis of intraventricular Neurocysticercosis. *Neurol India*. 2012;60(6):646-647.
22. Suh DC, Chang KH, Han MH, et al. Unusual MR manifestations of Neurocysticercosis. *Neuroradiology*. 1989;31(5):396-402.
23. do Amaral LL, Ferreira RM, da Rocha AJ, et al. Ferreira Neurocysticercosis: evaluation with advanced magnetic resonance techniques and atypical forms. *Top Magn Reson Imaging*. 2005; 16(2):127-144.
24. Stern WE. Neurosurgical considerations of cysticercosis of the central nervous system. *J Neurosurg*. 1981;55(3):382-389.