Cribriform plate dehiscence with encephalomeningocele revealed by recurrent meningitis: A case report

La déhiscence de la lame criblée avec encéphaloméningocèle révélée par une méningite récurrente: A propos d'un cas

Najla Soyah[1], Hajer Ben Belgacem[1], Donia Brahem[1], Nadia Mama[2], Raoudha Kebaili[1], Samia Tilouche[1], Marwa Abdelberi[1], Nesrine Jaballah[1], Jihene Bouguila[1], Lamia Boughamoura[1].

1. University of Medecine Sousse. Department of Pediatrics Farhat Hached hospital. Sousse. Tunisia
2. University of Medecine Sousse. Department of radiology. Sahloul hospital. Sousse. Tunisia

Abstract

Introduction: Meningitis is a potentially life threatening illness. It requires prompt diagnosis and treatment. Recurrent meningitis needs detailed investigations to identify the underlying cause.

Observation: We report a case of recurrent pneumococcal meningitis in a 9-year-old boy with an underlying congenital skull base abnormality. Brain computed tomography (CT) scan showed no obvious skull base defects. A magnetic resonance imaging (MRI) of the brain revealed a dehiscence of the cribriform plate with encephalomeningocele. The patient underwent an endoscopic repair of the bony defect and had not developed any new infections ever since.

Conclusion: This case highlights the need to investigate recurrent bacterial meningitis with CT scan and MRI of the brain and skull base. Repair of these congenital skull base defects are mandatory to prevent the recurrence of meningitis.

Key words: Cribliform plate dehiscence; Encephalomeningocele; Recurrent meningitis

Résumé

Introduction: La méningite est une maladie potentiellement mortelle. Elle nécessite un diagnostic et un traitement rapide. La méningite récurrente nécessite des investigations détaillées pour identifier la cause sous-jacente.

Observation: Nous rapportons le cas d'une méningite récurrente à pneumocoque chez un garçon de 9 ans présentant une anomalie congénitale de la base du crâne. La tomodensitométrie (TDM) cérébrale n'a montré aucun défaut évident de la base du crâne. Une imagerie par résonance magnétique (IRM) cérébrale a révélé une déhiscence de la lame criblée avec encéphaloméningocèle. Le patient a eu une réparation endoscopique du défaut osseux et depuis, il n'a développé aucune nouvelle infection.

Conclusion: Ce cas clinique met en évidence la nécessité d'explorer les méningites bactériennes récurrentes par une TDM et une IRM cérébrale et de la base du crâne. La réparation des défauts congénitaux de la base du crâne est obligatoire pour éviter la récidive des méningites.

Mots clés: Déhiscence de la lame criblée ; Encéphalomeningocèle ; Méningite récurrente

Introduction

Bacterial meningitis is a severe and life-threatening infection (1). It is associated with a high rate of complications and neurologic sequelae despite advances in antibiotic therapy and intensive care measures (1). Recurrence of bacterial meningitis is rare in children with an estimated incidence of 1.3% amongst all cases of bacterial meningitis (2). Recurrence of bacterial meningitis is defined as the re-emergence of signs and symptoms of meningitis at least three weeks after a sterile culture of the cerebrospinal fluid (CSF) if the pathogen is the same, or a new episode caused by a different pathogen (3).

A single episode of meningitis is often due to blood borne bacteria (4). However, in recurrent meningitis possible predisposing factors have to be investigated. Predisposing factors for recurrent bacterial meningitis can be divided into anatomical abnormalities, immunodeficiencies,

Correspondance
Hajer Ben Belgacem
University of Medecine Sousse. Department of Pediatrics Farhat Hached hospital. Sousse. Tunisia
Email: hajerbenbelgacem01@gmail.com

and chronic parameningeal infections (2,5). Structural approach and early diagnosis of an underlying pathology are crucial to prevent further episodes and improve the overall outcome for the affected individual (4,6). Anatomical abnormalities (congenital and/or acquired) are the most common predisposing factors for recurrent bacterial meningitis (4,7).

In this case, we report a 9-year-old boy with recurrent meningitis secondary to cribliform plate dehiscence with an encephalomeningoceles.

Observation

A 9-year-old boy was admitted to the pediatric department complaining of fever over the last two days associated with headache, nuchal pain and recurrent vomiting. His medical history was insignificant except for a Streptococcus pneumoniae meningitis episode he had four years earlier after an upper respiratory tract infection. The patient recovered well following treatment with intravenous antibiotics. The child did not receive a vaccine for S. Pneumoniae and there was no history of head injury, otorrhea, rhinorrhea or recurrent infections involving any other system.

On examination, the child was febrile at 39°C and lethargic. The central nervous system examination revealed neck stiffness with positive Kernig's and Brudzinski's sign (8). There was no focal neurological deficit and examination of the cranial nerves including the fundi was normal. Other systemic examinations did not reveal any abnormality.

Laboratory findings showed leukocytosis (17500 /mm3, normal range: 5000-11000/mm3), with an increase in polynuclear cells (10800/mm3, normal range: 1800-8000/mm3) and a high C-reactive protein level (140 mg/l, normal value < 6 mg/l). Suspicion of meningitis led to perform a lumbar puncture. CSF analysis identified a cell count of 1500/ml (normal range: 0-5 /ml) with 90% neutrophilic preponderance; an elevated protein concentration (1.2 g/l, normal range: 0.15-0.45 g/l) and a low glucose level of 0.83 mmol/l (normal range: 2.8-3.3 mmol/l) along with a simultaneous blood glucose level of 6.9 mmol/l (normal range: 3.5 et 6.1 mmol/l). Gram-positive diplococci were identified on CSF Gram staining. The CSF findings were compatible with bacterial meningitis. Antibiotics with cefotaxime (300 mg/kg/day) and vancomycin (60 mg/kg/day) were given immediately. The CSF culture grew Penicillin-sensitive Streptococcus pneumoniae.

Since this was the second episode of bacterial meningitis, complete immunological testing was performed, and the results were normal. The investigation included human immunodeficiency virus serology; serum immunoglobulin levels; complement classical and alternative pathway tests; and the presence of spleen in ultrasound. On otorhinolaryngological examination, no obvious CSF leak was demonstrated. Brain computed tomography (CT) scan showed no abnormalities. A magnetic resonance imaging (MRI) of the brain was performed to identify a possible undetected skull base defect. The MRI revealed an anterior and medial defect of the cribriform lamina with meningocele of the left middle turbinate and a small encephalocele (Figure 1). Two-weeks after intravenous cefotaxime and vancomycin treatment, the patient was discharged home without neurologic sequelae. A planned surgical repair of the defect was done through intranasal approach, and the patient had not developed any new infections since then.

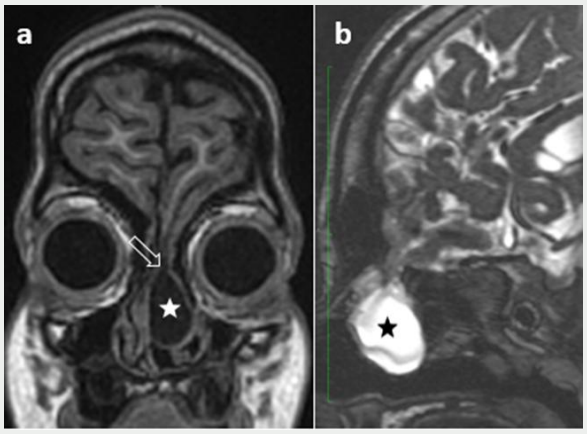

Figure 1. MRI of the brain

(a) 3D T1 coronal reconstruction

(b) 3D constructive interference in steady state (CISS) sagittal reconstruction: An anterior and medial defect of the cribriform lamina with meningocele of the left middle turbinate (star) and a small encephalocele (arrow)

Discussion

Recurrent bacterial meningitis in children is a rare disease since the intracranial structures are protected by bone and dura mater (9). Bony cranium is the most important barrier for the spread of infection into the intracranial structures (9). Dura mater is the second barrier reacting strongly in the face of an infection (9). A structural defect in these barriers facilitates the spread of infection to the subdural areas (9,10). The most common cause of recurrent meningitis is the presence of a persisting communication between the subarachnoid space and the air-filled cells of the temporal bone or the paranasal sinuses (9,10). The presence of bony defects underlying intact soft tissues should be considered and searched for meticulously (9). After a review of 114 publications on recurrent bacterial meningitis, Tebruegge and cutis (4) concluded that anatomic defects (acquired and congenital) were the greatest risk factors (59% of cases), followed by primary and secondary immunodeficiency (36% of cases).The findings of symptomatic or clinical CSF leak from the nose strongly suggests the possibility of occult skull base malformations (11). However, CSF rhinorrhea is difficult to diagnose in young children and infants and it is frequently misdiagnosed because CSF rhinorrhea can be indistinguishable from normal nasal discharge (12). Rhinorrhea and otorrhea should be tested for β-2 transferrin, which has high specificity for a CSF leak (12,13). However, β-2 transferrin is a protein also found in vitreous humor and inner ear perilymph in addition to CSF (6,13).

Congenital skull base encephalomeningocele is a herniation of intracranial contents through a skull base defect with an incidence of about 1/11.500 of live births

(2). Basal encephaloceles could be transethmoidal, spheno-ethmoidal, and trans-sphenoidal (2,14). It might remain unrecognized at birth since the herniated content exists within the nasal or paranasal cavity and cannot be observed on physical examination (10). It may also present as feeding or respiratory difficulties in the neonatal age, as nasal obstruction at any age in childhood, or as recurrent meningitis at any age (2,4). Interestingly, rhinorrhea in this group appears to be rare and occurs primarily after surgical interventions such as biopsy or polypectomy (2,4).

Prompt antibiotic treatment is essential for any case of suspected bacterial meningitis and improves outcome (12). Initial selection of antibiotics is made empirically prior to the availability of definite culture results, based on the incidence and susceptibility pattern in the population (12). The organism that causes recurrent meningitis may be the same as that in a previous attack or it may be different (6,12). At the second meningitis episode, the CSF culture revealed S. Pneumoniae, which is the infectious agent in more than 50% of patients with recurrent bacterial meningitis (4–6).The presence of a positive bacterial pathogen can often orient towards the possible etiology (11). Pneumococcus and Hemophilus suggest cranial dural defects (2,6). Escherichia coli and other gram negative bacilli suggest spinal dural defects, and Neisseria meningitidis suggests complement deficiency (3,6,11).

When a structural defect is suspected, CT or MRI are first-line imaging examinations (15). Thin section cranial CT offers a relatively easy, reliable, and non-invasive method of delineating anatomical defects in recurrent meningitis (16). Axial cranial computed tomography may fail to identify defects in the basal ethmoidal area and cribriform plate and thus give false reassurances, whereas coronal thin sections show detailed anatomy of the anterior cranial fossa and identify most skull defects (16). Brain three dimensional (3D) multi-detector CT imaging is particularly useful for investigating precise bone structural abnormalities in the auditory ossicles or skull base (17). MRI cisternography using a steady-state free precession technique allows for multiplanar reformats and facilitates the localization of the actual site of the CSF leak and detection of encephalocele or meningoencephalocele (18). Herniation of brain parenchyma or meninges through the bone defect could be easily visualized on MRI images and it allows an accurate depiction of the olfactory and optic tracts and hypothalamic-pituitary system (15,16). If conventional imaging is non-diagnostic and the clinical suspicion for a CSF fistula remains, radionuclide cisternography should be pursued (13,15).Invasive radiological examinations should be implemented as a second-line step (15). Radionuclide cisternography involves the intrathecal administration of the radiotracer, followed by acquisition of the images (18). The accumulation of the radiotracer in the nasal cavity or nasopharynx points to the presence of a CSF fistula (15,18). Identification of the CSF leak is essential for surgical repair. Nasal endoscopy has been the treatment of choice for more than a decade for the repair of anterior cranial fossa defects (19). Endoscopic repair of cribriform CSF leaks enjoys a very high success rate with few complications, recurrence rates, and morbidity (19,20).

Interestingly, studies have reported a more favorable outcome and a lower mortality rate in patients who suffer from recurrent bacterial meningitis than in those who suffer from one episode (6,21).This can be explained by several factors. First, patients with recurrent meningitis might recognize symptoms of meningitis, promoting early medical attention (6). Patients with recurrent meningitis presented early with less severe symptoms compared with the non-recurrent cases (6). In a disease as deadly as bacterial meningitis, early antibiotic treatment is one of the most important factors for favorable outcome (6,22).Second, episodes are often predisposed by a CSF leakage, which contributes to a better prognosis due to the relatively benign pathophysiology (23). Third, patients with recurrent meningitis might have a less severe inflammatory response, potentially caused by immuneocompromise and inborn errors of innate immunity (6). Bacterial meningitis is a complex disease with outcomes driven by the host inflammatory response (22). Even, the outcome of recurrent meningitis episodes appears to be favorable; long-term neurological sequelae could be identified in a large number of patients after bacterial meningitis (6). This provides further incentive to actively search for underlying conditions to prevent further recurrent episodes.

The prevention of recurrent bacterial meningitis requires the detection and elimination of risk factors, including surgical repair of CNS defects and age appropriate vaccinations (23, 24). Patients susceptible to recurrent bacterial infections should receive immunization against pneumococcus, meningococcus and hemophilus (24). There is no evidence to support the use of antibiotic prophylaxis in patients with a cerebrospinal fluid leak (24). In a randomized controlled trial, Ratilal et al. (25) failed to identify a benefit of prophylactic antibiotics in preventing meningitis in patients with CSF leaks and skull base injuries.

Conclusion

Recurrent pneumococcal meningitis may be due to an underlying anatomical skull base defect, and a multidisciplinary team, which includes otolaryngology, neurosurgery and pediatrics, is necessary to successfully treat the disease. The present case report highlights the importance of detailed imaging in the form of CT scan of both temporal bone and anterior skull base as well as the value of an MRI study if the CT scan is normal. It is mandatory that all skull base fistulae be repaired to prevent further recurrences.

References

1. Sfaihi L, Kamoun F, Kamoun T, Alloulou H, Mezghani S, Hammemi A, et al. Bacterial meningitis in children: Epidemiological data and outcome. La Tunisie médicale. 2014;141-6.
2. Alsaeed G, Alsaeed I, Alsaeed M. Recurrent Bacterial Meningitis in

Paediatrics. American Journal of Pediatrics. 2019;5 (3):98.

3. Masri A, Alassaf A, Khuri-Bulos N, Zaq I, Hadidy A, Bakri FG. Recurrent meningitis in children: etiologies, outcome, and lessons to learn. Child's Nervous System. 2018; 34(8):1541-7.
4. Tebruegge M, Curtis N. Epidemiology, etiology, pathogenesis, and diagnosis of recurrent bacterial meningitis. Clinical Microbiology Reviews. 2008; 21(3):519-37.
5. Janocha-Litwin J, Simon K. Recurrent meningitis--a review of current literature. Przeglad Epidemiologiczny. 2013; 67(1):41-5, 125-9.
6. Ter Horst L, Brouwer MC, van der Ende A, van de Beek D. Recurrent Community-Acquired Bacterial Meningitis in Adults. Clinical Infectious Disease 2021; 73(9):e2545-51.
7. Tuygun N, Tanir G, Aytekin C. Recurrent bacterial meningitis in children: our experience with 14 cases. The Turkish Journal Pediatrics. 2010; 52(4):348-53.
8. Tracy A, Waterfield T. How to use clinical signs of meningitis. Archives of Disease in Childhood Education and Practice Edition.2020;105 (1):46-9.
9. Bektas D, Caylan R, Bahadir O, Caylan R. Occult anterior skull base defect without rhinorrhea as a cause of recurrent meningitis. Surgical Neurology. 2007; 68(1):50-2.
10. Li X, Liu HZ, Pang L yu, Wen X, Sun SZ. A Retrospective Study of Recurrent Bacterial Meningitis in Children: Etiology, Clinical Course, and Treatment. Computational and Mathematical Methods in Medecine. 2022; 2022: 3681871.
11. Morgenstern Isaak A, Bach Faig A, Martínez S, Martín-Nalda A, Vázquez Méndez E, Pumarola Segura F. et al Recurrent meningitis due to anatomical defects : The bacteria indicates its origin. Anal of Pediatria. 2015; 82(6):388-96.
12. Lee E, Yum MS. Recurrent Bacterial Meningitis in Pediatric Patients with Anatomical Defects. Annals of child neurology.2011;19(2):101-107
13. Rosenberg J, Galen BT. Recurrent Meningitis. Current Pain and Headache Reports. 2017; 21(7):33.
14. Dewan R, Anuradh S, Garg S, Kochhar R, Negi M, Nischal N. Encephalcoele- A Cause of Recurrent Meningitis. jk sciences. 2015; 17(1):54-56.
15. Masuoka S, Miyazaki O, Takahashi H, Tsutsumi Y, Hiyama T, Kitamura M, et al. Predisposing conditions for bacterial meningitis in children: what radiologists need to know. Japanese Journal of Radiology. 2022;40 (1):1-18.
16. Carrol ED, Latif AH, Misbah SA, Flood TJ, Abinun M, Clark JE, et al. Recurrent bacterial meningitis: the need for sensitive imaging. The BMJ. 2001; 323(7311):501-3.
17. Akimoto T, Morita A, Shiobara K, Hara M, Minami M, Shijo K, et al. Surgically Cured, Relapsed Pneumococcal Meningitis Due to Bone Defects, Non-invasively Identified by Three-dimensional Multi-detector Computed Tomography. Internal Medicine. 2016; 55(24):3665-9.
18. Hiremath SB, Gautam AA, Sasindran V, Therakathu J, Benjamin G. Cerebrospinal fluid rhinorrhea and otorrhea: A multimodality imaging approach. Diagnostic and Interventional Imaging. 2019; 100(1):3-15.
19. Wilson M, Snyderman C. Endoscopic Management of Developmental Anomalies of the Skull Base. Journal of Neurological Surgery. 2018; 79(1):13-20.
20. Mantravadi AV, Welch KC. Repair of Cerebrospinal Fluid Leak and Encephalocele of the Cribriform Plate. Atlas of Endoscopic Sinus and Skull Base Surgery. 2019. 223-232.
21. Adriani KS, van de Beek D, Brouwer MC, Spanjaard L, de Gans J. Community-acquired recurrent bacterial meningitis in adults. Clinical Infectious Disease .2007;45(5):46-51.
22. Van de Beek D, Brouwer M, Hasbun R, Koedel U, Whitney CG, Wijdicks E. Community-acquired bacterial meningitis. Nature Review Disease Primer. 2016;(2):16074.
23. Ter Horst L, Brouwer MC, van der Ende A, van de Beek D. Community-acquired Bacterial Meningitis in Adults With Cerebrospinal Fluid Leakage. Clinical Infectious Diseases. 2020;70 (11):2256-61.
24. Rimmer J, Belk C, Lund VJ, Swift A, White P. Immunisations and antibiotics in patients with anterior skull base cerebrospinal fluid leaks. The Journal of Laryngology Otology. 2014; 128(7):626-9
25. Ratilal BO, Costa J, Pappamikail L, Sampaio C. Antibiotic prophylaxis for preventing meningitis in patients with basilar skull fractures. Cochrane Database Systimic Review. 2015;(4):CD004884.