CASE REPORT

An unusual case of *E coli* meningitis in a patient with Marfan's syndrome

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SUMMARY

Spontaneous non-traumatic *Escherichia coli* meningitis is very rare in adults. We report a case of a 48-year-old woman with Marfan's syndrome with *E coli* meningitis. Apparently, the relation between an increased risk of meningitis and Marfan's syndrome is not well known. This patient was discharged on intravenous antibiotic therapy after a diagnosis of *E coli* meningitis without looking for the cause by imaging studies previously. Her blood cultures were negative ruling out haematogenous spread. Our work-up revealed extensive dural ectasia with intrasacral meningoceles extending into the pelvis possibly acting as a portal of entry for the bacteria into the brain from the gastrointestinal tract.

BACKGROUND

Patient's with Marfan's syndrome are likely more prone for meningitis from atypical organisms like *Escherichia coli* as they may have sacral meningoceles and dural ectasias in the pelvic region allowing intra-abdominal bacteria into the central nervous system through developmental defects. These patient's should receive imaging of the spine and pelvis for potential ports of entry to prevent recurrence. Also, possible surgical options need to be considered to prevent life-threatening meningitis recurrence.

CASE PRESENTATION

A 48-year-old woman with Marfan's syndrome presented to the emergency room with persistent headaches. She originally presented to another facility with left-sided back pain and fever and was diagnosed to have *E coli* urinary tract infection with no pyelonephritis. Almost 2 days after hospital admission started to have high-grade fever, neck stiffness and altered behaviour. Lumbar puncture revealed 1400 white blood cells (WBCs) with 83% neutrophils, with a glucose of 4 mg/dl and protein of over 200 mg/dl. Blood cultures were negative and cerebrospinal fluid (CSF) cultures grew *E coli*. She was started on ceftriaxone at a dose of 2 g intravenously every 12 h with a plan of treating for 2 weeks as CSF cultures were consistent with *E coli* meningitis and was discharged. She was re-admitted because of persistent headaches, nausea and vomiting on the fourth postdischarge day to our facility. She was continued on ceftriaxone with the addition of vancomycin. Repeat lumbar puncture showed 513 WBCs with 69% granulocytes, a glucose of 86 mg/dl and protein of 163 mg/dl. Repeat blood cultures and CSF cultures were negative. An extensive work up was carried out to look

for possible portal of entry of *E coli* into the meningeal cavity.

INVESTIGATIONS

An MRI of lumbar spine with and without contrast showed prominent intrathecal enhancement and clumping of the nerve roots, consistent with meningitis. Also, extensive dural ectasia with intrasacral meningoceles extending into the pelvis and associated remodelling of the lumbar vertebral bodies were seen consistent with a known history of Marfan's disease. The anterior margin of arachnoid scar tissue seen in the pelvic retroperitoneum was in proximity to the small intestine and loops of large intestine. This area was seen intimately enhanced suggesting an inflammatory response. There was no evidence of air-fluid level within the CSF-filled spaces in the sacrum. The lumbar MRI was otherwise unremarkable.

CT abdomen and pelvis with and without contrast showed no acute process within the abdomen or pelvis. Other chronic findings as aforementioned, including findings related to dural ectasia in the setting of patient's history of Marfan's syndrome.

CT intravenous pyelogram did not reveal any abnormalities.

Abdominal ultrasound did not show any pyelonephritis.

Initial urine culture from another facility showed more than 1 00 000 *E coli* with negative blood cultures and CSF cultures grew *E coli*.

All blood cultures, CSF and urine cultures repeated during rehospitalisation at our facility were negative.

Also, CSF venereal diseases research laboratory (VDRL), CSF, herpes simplex virus, PCR, HIV serologies were negative.

Immunoglobulin levels were within normal limits.

DIFFERENTIAL DIAGNOSIS

Based on the CSF findings of elevated protein, low glucose and neutrophilic predominance, bacterial meningitis was high on the differentials. Other aetiologies like abdominal abscesses, pyelonephritis and immune deficiency states were also considered and ruled out.

TREATMENT

The patient was started on ceftriaxone intravenously and headaches gradually resolved. Since the MRI showed inflammation around the arachnoid scar tissue lying in proximity with the retroperitoneum,

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neurosurgery was consulted for possible surgical interventions to prevent recurrent meningitis from gut pathogens especially *E coli*. Since the risks of surgery outweighed the benefits, no intervention was performed. The patient was neurologically intact and hence a decision was made to consider the procedure for recurrences. Prolonged oral antibiotic suppressive therapy was offered to the patient, and she preferred to consider it if symptoms recurred.

OUTCOME AND FOLLOW-UP

The patient was symptom free after completing intravenous ceftriaxone for 2 weeks. She was advised to seek immediate medical attention if any recurrent signs of meningitis occurred for an early initiation of antibiotics.

DISCUSSION

Recurrent bacterial meningitis in an immunocompetent host is a rare phenomenon and generally poses a considerable diagnostic challenge especially if it is from a less-common organism like *E coli*. A systematic approach is needed to diagnose and prevent further episodes and improve the overall outcome. Most of the literature describes 'recurrent meningitis' as being two separate episodes of meningitis that are separated by a period of convalescence and complete recovery, where as recrudescence and relapse represent persistence of the 'original' infection from treatment failure. Work up should include looking for congenital inner ear malformations like Mondini dysplasia, asplenia, basal skull fractures, terminal complement deficiencies, X linked agammaglobulinemia, IgG subclass deficiency, IRAK-4 deficiency, HIV and parameningeal foci of infections.¹ Even a single episode of Gram negative enteric bacillary meningitis should raise a concern and search for CSF fistulas especially rectothecal fistulas, vertebral and meningeal anomalies and urinary tract malformations.¹ Recurrent Gram negative enteric bacillary meningitis is reported predominantly in patients with lumbosacral lesions like meningoceles and dermal sinuses, and these patients

should be evaluated by imaging studies and surgical interventions pursued if indicated.^{2 3} *E coli* is a Gram negative enteric organism associated with meningitis. In cases like this a detailed history and physical examination is very important to provide diagnostic clues. This patient had a history of Marfan's syndrome which is known to cause spinal anomalies, hence prompt work up was carried out in this regard.

Learning points

- *Escherichia coli* meningitis in an immunocompetent person with blood culture being negative requires evaluation of spinal column for possible meningoceles.
- Patient's with marfan's syndrome are at a risk for meningitis from gut pathogens especially *E coli* from meningeal abnormalities acting as a portal of entry.
- Surgical options should be explored for patients with recurrent meningitis from colonic pathogens with meningoceles or other anomalies amenable to surgery.^{2 3}

Contributors RVK and JM have contributed to the write up.

Competing interests None.

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