

## Group A Streptococcus Meningitis Following Arnold-Chiari Correction Surgery: A Case Report

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### Abstract

Group A *Streptococcus* (GAS) can cause invasive disease but rarely causes meningitis. Here we report a three-year-old girl with a GAS meningitis after Arnold-Chiari type I corrective surgery. Six days after surgery she presented fever, headache, vomiting and surgical wound inflammatory signs. A cerebrospinal fluid (CSF) collection with a fistulous path to skin was identified and GAS was found in blood and CSF cultures. Despite targeted antibiotherapy, due to lack of improvement and difficult management of the CSF fistula, she needed to be submitted to several surgical corrections and increased antibiotherapy spectrum. Despite the low risk in acquiring bacterial meningitis after neurosurgery, these procedures may be a direct route for infection. Surgery complications like a fistula are a known risk factor for infection and although uncommon, GAS meningitis may be difficult to manage and can cause serious morbidity.

**Key Words:** Arnold-Chiari type I malformation surgery, Group A *Streptococcus* invasive disease; Group A Streptococcus meningitis.

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## 1- INTRODUCTION

Group A *Streptococcus* (GAS) also known as *Streptococcus pyogenes*, a gram-positive coccoid-shaped bacterium that grows in chains, is associated with a wide spectrum of infections (1, 2). The only known GAS reservoirs are the human skin and mucous membranes and it usually causes a mild disease like pharyngitis and cellulitis (1-3). Less commonly, it causes invasive disease, including necrotizing soft tissue, toxic-shock syndrome, bacteraemia, respiratory tract infection and rarely meningitis, with significant associated morbidity and mortality. Invasive disease has a global incidence estimated around 2-3.5 cases of infection per 100,000 people with a case-fatality rate of 30 to 60%, despite GAS being uniformly susceptible to various classes of antibiotics including penicillin (1, 2). Although there are some reported cases of GAS meningitis, it is a rare causative agent of bacterial meningitis, accounting for 1% or less of all cases (4-7).

Arnold-Chiari (AC) malformations are a group of deformities of the posterior fossa and hindbrain (cerebellum, pons and medulla oblongata). They are classified based on morphology and severity of anatomic defects, AC I to IV (8). Type I is the least severe and it is characterized by one or both abnormally shaped cerebellar tonsils that project at least 5 mm below the foramen magnum, measured by McRae Line. Decompressive surgery is the main treatment for symptomatic AC type I that cannot be managed by conservative measures, with clinical and imaging surveillance. It re-establishes the CSF flow across the craniovertebral junction and relieves pressure on the cerebellum and hindbrain, by decompressing the posterior fossa (8, 9). We report a case of GAS meningitis following AC type I corrective surgery.

## 2- CASE REPORTS

A three-year-old girl symptomatic since one-year-old, presenting with headache and vomiting with mild physical activity was diagnosed with AC malformation type I (cerebellar tonsils projecting 9 mm below the foramen magnum), without hydrocephalus, syringomyelia or other malformations. In February 2020 she was admitted in the Paediatric Intensive Care Unit of Centro Materno-Infantil do Norte - Centro Hospitalar Universitário do Porto, Portugal, following post corrective AC surgery – suboccipital craniectomy, excision of the posterior C1 arch and duraplasty with neuropatch. During the surgery all preventive measures to guarantee asepsia were taken and no complications or intercurrents were reported. On day six (D6) after surgery, she started fever, headache, vomiting, surgical wound swelling and CSF and purulent material drainage by the surgical wound.

On examination she had a Glasgow coma score of 15 and no abnormalities were present in neurological examination. She had normal blood pressure, heart rate, respiratory pattern and rate, and the remaining physical exam was unremarkable. Cranial computed tomography (CT) showed intracranial hypertension with mild hydrocephalus and a 20x30x66 mm collection inferior to cerebellum compatible with a CSF collection and a fistulous path to skin with a subcutaneous infectious process (**Figure.1**).

Analytically presented leucocytosis (35330/ $\mu$ l) with neutrophilia (93%), thrombocytosis (579 000/ $\mu$ l) and 419.82mg/L protein-C-reactive (PCR). Empirical antibiotherapy with vancomycin and ceftazidime was initiated. She was submitted to a second surgery to clear the wound and correct the CSF fistula with a blood patch. On D7 after first surgery, multisensitive GAS was identified in blood and CSF culture, ampicillin and clindamycin were started, suspending the previous antibiotherapy. She maintained fever,

sporadic vomiting and headache. At this point, Cranial CT scan showed a persistent epidural CSF collection at the previous surgical approach area, with no other major complications. On D12, due to persisting symptoms, a lumbar puncture was performed, showing an amicrobial CSF with pleocytosis and a predominance of polymorphonuclear cells. On D14, due to loss of CSF through the surgical wound, a new surgical review was made with removal of the old patch and placement of a new duraplasty with cervical fascia. A lumbar drain was positioned due to hydrocephalus. At this time, CSF was cloudy (5744/mm<sup>3</sup> leucocytes, 4642 polymorphonuclears and 1.71mg/dl proteins), and culture was negative. Blood work showed: 11310/ul leucocytes count and a 30.4mg/L PCR. The antibiotherapy was changed to vancomycin and meropenem. She became feverless 2

days after (D16). She kept having multiple episodes of drain malfunction and extra-drain leakage needing several drain removals and replacements. On D25 she needed another surgical reintervention of duraplasty reinforcement, due to recurrence of surgical wound CSF drainage and hydrocephalus – assumed as a consequence of previous meningitis. Lumbar drain was removed at D32. Multiple CSF harvests were carried out, in which cultures were sterile. On D34 a peritoneal ventricular shunt with a programmable valve was placed, resulting in a progressive reduction of the CSF collection, without recurrence of the surgical wound CSF fistula. She became asymptomatic with no evident neurological deficits. Six months after surgery, the surgical wound was healed and she was symptomless.



**Fig.1:** TC scan showing a fistulous path to skin with inflammatory process (arrow).

### 3- DISCUSSION

The patient described had AC type I malformation. Most patients are usually asymptomatic during their first years of life, while others present symptoms related to elevated intracranial pressure, cranial neuropathies, brainstem cerebellar compression or myelopathy. The most common presentation is a headache localized to the suboccipital or neck area that is exacerbated by activities that increase intracranial pressure, as presented by our patient. Nausea, vomiting, sensory changes, motor deficits and vertigo are sometimes described (8-10). AC malformations are frequently accompanied by brainstem, cerebellum, and craniocervical junction anatomic abnormalities (9, 11). As previously stated, decompressive surgery is the main treatment for recurrent symptomatic patients. Neurosurgery complications can occur in 16.4% of all paediatric neurosurgical interventions, including CSF leak, new neurological deficit, early shunt or endoscopic third ventriculostomy blockage, shunt infection, haemorrhage and wound infection (12, 13).

Neurosurgery procedures penetrate the dura mater, and therefore may introduce bacteria in CSF or meninges or cause a fistula of the meninges which may be a direct route for infection (14). Despite the overall low risk in acquiring bacterial meningitis after surgery, it cannot be disregarded. Meningitis following craniotomy occurs in 0.3 - 8.6% of all patients and it depends on indication for surgery, underlying medical conditions, and local implementation of infection-control measures (12, 14, 15). The presence of a fistula or wound infection is a well-known risk factor for meningitis. Drake et al described meningitis in 13% of all CSF leakage and 10% of reported wound infections related to neurosurgery (12). Our patient presented a CSF collection related to the neurosurgery

intervention and a fistulous path to skin, with an associated subcutaneous infectious process. These findings could increase the risk of bacteria migration related to the infection process, to the meninges and CSF, causing meningitis and all the associated complications. Most cases of bacterial meningitis occur in the first days after surgery, however, some cases have been reported more than 10 days after (14). Thereafter, due to multiple CSF wound leakage episodes, the patient had to be submitted to several surgical corrections of the CSF fistula, with suboptimal results. Post-neurosurgical meningitis is caused mainly by cutaneous organisms such as coagulase negative staphylococci, *Staphylococcus aureus* and sometimes non-cutaneous as *Enterobacteriaceae*, *P. aeruginosa*, *Acinetobacter spp.*, *Streptococcus pneumoniae* but rarely GAS, as reported in this patient (15, 16).

Antibiotic therapy primarily applied to the patient was aimed at these common agents. As stated before, GAS is susceptible to penicillin, which is the reason why ampicillin was given to the patient, despite her being under vancomycin and ceftazidime. CSF was sterile in 6 days, although maintaining an inflammatory CSF. Due to the persistence of fever, occasional vomiting and the CSF fistula, it became necessary to increase antibiotic spectrum, changing to vancomycin and meropenem with good results. Moreover, post-neurosurgical inflammatory processes often have a non-infectious aetiology, presenting fever and CSF with inflammatory characteristics (15).

Outcome of GAS meningitis is less well known than other bacterial meningitis due to its low incidence and report, though it seems to be related to high morbidity. GAS meningitis seems to be associated mainly with neurological sequelae, although none has been recognized in the case we reported (17). It has been described that in 46% of children with

GAS meningitis, hearing loss, epilepsy, neurodevelopmental delay and behavioural problems can occur (18). In addition to the necessary follow-up for a post-op AC patient, follow-up of hearing and neurologic development is strongly suggested for these kinds of patients.

#### 4- CONCLUSION

Despite the low risk in acquiring bacterial meningitis after neurosurgery, these procedures may be a direct route for infection. Surgery complications such as fistula or subcutaneous infection are known risk factors and may require surgical reintervention. GAS meningitis is uncommon and may be difficult to manage. It can cause serious morbidity and be associated with important neurological sequelae. Long-term follow-up is necessary to assess GAS meningitis outcomes.

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