Annals of Case Reports
Papacharalampous A, et al. Ann Case Rep: 9: 101733
www.doi.org/10.29011/2574-7754.101733
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Case Report

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Cerebellar Abscess Following Craniocervical Junction Decompression for Chiari I Malformation in a Patient with Rheumatoid Arthritis: A Case Report.

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Citation: Papacharalampous A, Kouyialis T, Stylianou P (2024) Cerebellar Abscess Following Craniocervical Junction Decompression for Chiari I Malformation in a Patient with Rheumatoid Arthritis: A Case Report. Ann Case Report. 9: 1733. DOI:10.29011/2574-7754.101733

Received: 25 March 2024, **Accepted:** 30 March 2024, **Published:** 02 April 2024

Abstract

'Chiari Malformations' are a group of pathologies involving the Craniocervical junction. The most common is Chiari I malformation (CIM), which is characterised by a downward dislocation of the cerebellar tonsils through the foramen magnum. Symptomatic patients and those with syringomyelia are candidates for decompression of the Craniocervical junction. A variety of complications have been described in the literature following this procedure. We present the case of a cerebellar abscess following Craniocervical junction decompression for CIM in a patient with rheumatoid arthritis (RA), which, in our knowledge, is the first described in the literature. We believe that RA and its treatment were major contributors to this complication, mainly due to the immune impairment and the subtle infectious clinical manifestations that they cause. This case led us to modify the perioperative management of patients with RA who undergo spinal surgery in our department. We suggest that strict infection prevention measures should be taken by spine surgeons who treat patients with RA and a high suspicion index for infection should be maintained.

Keywords: Chiari I Malformation; Rheumatoid Arthritis; Cerebellar Abscess; Craniocervical Junction Decompression.

Introduction

The term 'Chiari Malformations' describes a group of entities involving the Craniocervical junction. It is divided in four subgroups: Chiari I, II, III, IV [1]. Chiari I malformation (CIM) is the most common and it is characterised by a downward dislocation of the cerebellar tonsils through the foramen magnum [1]. CIM has a prevalence of approximately 1/1000 in the general population for symptomatic patients, while people meeting imaging criteria may be up to 1% [2,3]. Due to the extended use of imaging technology, CIM is a common incidental finding [4,5].A caudal displacement of the cerebellar tonsils through the foramen magnum of 5mm or more is usually sufficient for the radiological diagnosis of CIM [6,7], while some support that lower values of dislocation may also be indicative of CIM [6,8,9]. Syringomyelia coexists in approximately 50-75% of patients [8]. Headache is the most common symptom, usually occipital/ upper cervical, often exacerbated by Valsalva maneuver [1,10,11]. A variety of other symptoms and signs may also manifest, due to dysfunction of the cerebellum, cranial nerves, brainstem and spinal cord [10]. Surgical intervention is indicated for symptomatic patients and for those with syringomyelia even in the absence of symptoms. The extent of the cerebellar tonsils caudal dislocation is also taken into consideration [1,8]. Surgery aims to allow normal CSF flow to the cervical subarachnoid space, by expanding the posterior cranial fossa, thereby recreating the cisterna magna [8].The procedure

performed is decompression of the Craniocervical junction, during which, a piece of the occipital bone and the posterior arch of the atlas is removed, usually followed by duroplasty [1]. Common complications of the procedure are CSF leak, pseudomeningocele, meningitis (aseptic or bacterial), neuronal or vascular damage [1]. Less common complications are wound infection, epidural/subdural hematoma of the posterior cranial fossa, acute hydrocephalus, respiratory dysfunction, postoperative headache, neurological deficit, trigeminal neuralgia, intraventricular bleeding, respiratory dysfunction, embolization, death [12,13].

Case Report

A 58-year-old female was referred to our neurosurgical department, with the diagnosis of CIM. Her symptoms included persistent occipital headaches, dizziness, gait instability and weakness of upper and lower extremities. Physical examination revealed spastic quadriparesis and gait instability in tandem gait test (heel-to-toe). A brain MRI revealed CIM with 13 mm caudal dislocation of the cerebellar tonsils (Figure 1). Further imaging of her spinal cord did not reveal the presence of syringomyelia. The rest of her medical history was positive for rheumatoid arthritis (RA) treated with hydroxychloroquine (200mg 1x1 pos) and occasionally methylprednisolone (16mg 1x2 pos) and analgesics PRN (diclofenac, tramadol, naproxen, meloxicam), dyslipidemia (atorvastatin 10mg 1x1 pos), and depression (paroxetine 20mg 1x1 pos). Based on the patient's symptoms and diagnosis, a decision was made for surgical intervention. For better operative planning, a CT angiography of the brain and neck was performed, aiming to identify the exact course of the vertebral arteries, thus avoiding a possible intraoperative damage due to anatomical variations. The patient underwent posterior cranial fossa craniectomy and laminectomy of C1 followed by duroplasty. She was discharged on the third postoperative day. The wound was routinely inspected a week after the procedure. Stiches were removed on the 14th postoperative day. At one-month follow-up, the patient reported complete resolution of the headaches. Physical examination revealed improvement of strength in upper and lower extremities and improvement in the gait test. At four-month follow-up, a further improvement of her symptoms and signs occurred and a new brain MRI revealed adequate decompression of the Craniocervical junction. The scan also revealed a fluid collection with peripheral contrast enhancement on the dorsal part of the C1-2 vertebrae, below the position of the occipital craniectomy, with edema and contrast enhancement on the surrounding tissues (Figure 2). Differential diagnosis was between inflammatory collection (abscess) and organized hematoma. Laboratory findings were unremarkable: white blood cells= 8300/μL, neutrophils= 4810/μL, ESR= 45 mm/hr (normal range: 0-15), CRP= 1.66 mg/dl (normal range: <0.5). The increased ESR and CRP were attributed to RA. Furthermore, the patient was afebrile, and she did not complain of local pain. Taking these into consideration, the possibilities of an inflammatory collection seemed low and no intervention followed. At 11 months following the procedure, the patient returned with a new onset of headaches and gait instability. She immediately underwent a new brain MRI, which revealed a fluid filled cavity with imaging characteristics of an abscess in the lower part of the right cerebellar hemisphere, in addition to the fluid collection that was previously observed (Figures 3&4). Laboratory findings were white blood cells: 7800/μL, neutrophils: 3060/μL, ESR: 32, CRP: 7.4. Given the high possibilities for a cerebellar abscess a reoperation was performed. During surgery, a granulomatous tissue was identified at the area of the previous cervical epidural collection and a cerebellar abscess. The granulomatous tissue was debrided and the abscess was drained. Samples were taken for cultures and antibiogram and empiric antibiotic therapy was initiated (Vancomycin 1gr BD and Meropenem 2grs TDS). Results from the cultures, failed to isolate any pathogen. During her hospital stay she showed significant improvement, and she was discharged on the 23rd postoperative day. The first follow-up was performed one month after her discharge and the second four months later along with a new MRI scan of the brain. At her second follow up, she reported complete resolution of her symptoms and imaging showed no signs of abscess or collection (Figure 5).

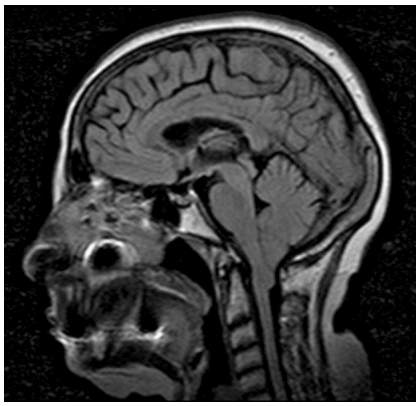

Figure 1: Preoperative MRI scan showing CIM with 13mm caudal dislocation of the cerebellar tonsils through the foramen magnum.

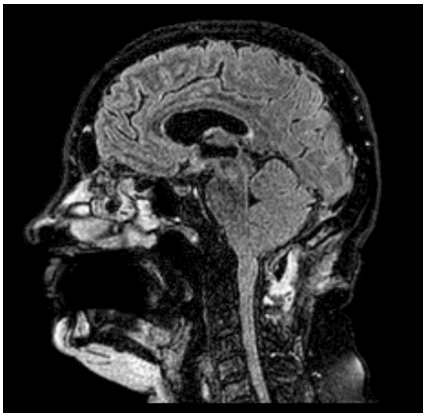

Figure 2: Postoperative MRI scan showing adequate decompression of the foramen magnum along with a sub-occipital fluid collection.

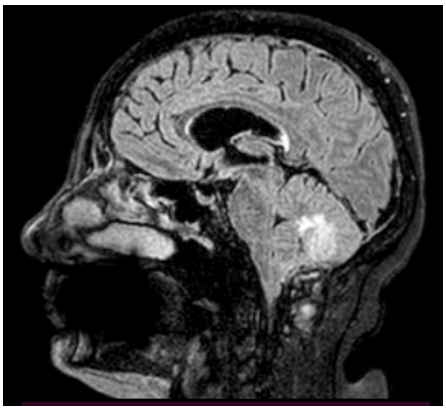

Figure 3: MRI scan showing a lesion with characteristics of abscess in the right lower hemisphere of the cerebellum (FLAIR sequence).

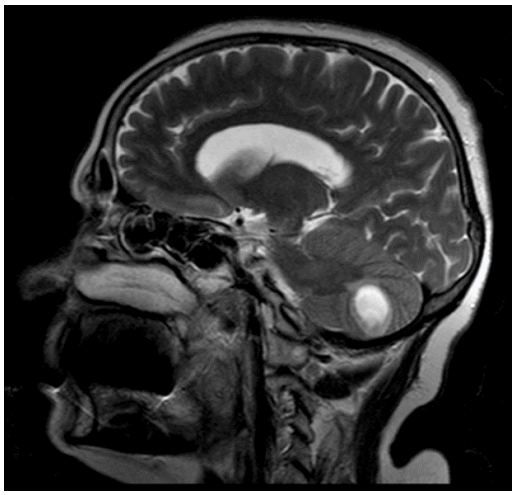

Figure 4: MRI scan showing a lesion with characteristics of an abscess in the right lower hemisphere of the cerebellum (T2W sequence).

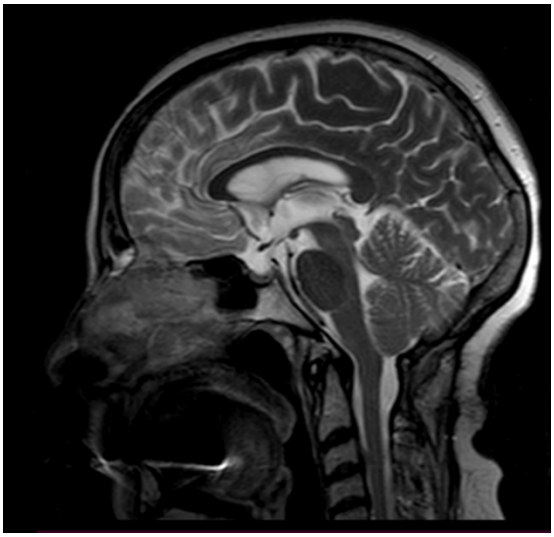

Figure 5: Postoperative MRI scan after reoperation for the drainage of the abscess.

Discussion

A variety of postoperative complications have been described in the literature following surgical management of CIM [1,7-37]. To our knowledge, this is the first reported case of a cerebellar abscess following a craniocervical junction decompression for Chiari I malformation, both in adult and in pediatric patients. A possible mechanism that led to the formation of the abscess may be the presence of the suboccipital epidural collection identified at the first scan that was subsequently infected. An ascending infection to the inferior cerebellum probably followed, that led to the formation of the cerebellar abscess. Patient's comorbidities and medication taken at home (RA treated with hydroxychloroquine and methylprednisolone) possibly blunted immune response, thereby facilitating infection of the collection and expansion of the infection to the cerebellum. Another possible explanation is that the first collection was infectious from the beginning, by a pathogen of low virulence. This would explain the slow course of the infection. A blunted immune response, along with the analgesics and NSAIDs she was using (PRN) for RA, may also explain the limited clinical manifestations until the infection had further expanded. It is widely accepted in the literature that routine laboratory tests have limited contribution in the diagnosis of an intracranial abscess and normal laboratory results cannot exclude its presence [38]. This may also explain the unremarkable initial laboratory findings in our case. In addition, negative cultures from the samples taken intraoperatively are in agreement with the fact that 14%-34% of samples taken from brain abscesses result in negative cultures [38]. A variety of possible mechanisms have

been described as the reason for the increased infection rate in RA [39,40-43]. Dysfunction of the innate and acquired immune response is a key factor [40]. Neutropenia and dysfunction of neutrophils, either autoantibody- mediated or medication-mediated, causes an impaired innate response [40,45]. On the other hand, 'ageing' of the immune system observed in RA as well as in other autoimmune disorders, leads to a malfunctioning acquired immune response [40]. Moreover, several genetic polymorphisms found in patients with RA, may also be responsible for this, thus, the increased susceptibility to infections [40]. Furthermore, medication used for the management of RA, like glucocorticoids, cytokine inhibitors and other Disease Modifying Antirheumatic Drugs (DMARDs) may compromise immune response and increase infection rate [40,43,45] and it is therefore advisable that such patients undergo modification of their medical regime with the aid of a Rheumatologist, aiming to decrease infection risk, while maintaining disease control [42,46]. Lastly, RA patients usually carry several comorbidities, like obesity and diabetes that are identified factors which independently increase infection rate [40]. This experience has led us to modify the perioperative management of patients with RA undergoing spinal surgery in our department. Apart from the perioperative modification of the regimen of certain DMADs, in collaboration with a Rheumatologist, we also follow strict infection prevention measures, some of them, inspired by the retrospective study of El-Kardi et al. (2019) [44]. Patients are advised to wash thoroughly with a disinfectant soap the day before surgery. Prior to the incision, the area is carefully prepared first three times with chlorhexidine, followed by three times with povidone iodine solution. Doors of the operative theatre remain closed throughout the operation. During the procedure, the preparation of the tissues is performed with minimum use of coagulation so as to achieve hemostasis without severely affecting tissue perfusion that increases susceptibility to infections. Prior to suturing, the surgical field is irrigated with three litres of normal saline and the wound is covered with vancomycin powder. Wound dressing is not changed during first 48 hours, unless if wet. After discharge, the caregiver is educated for the proper care of the surgical wound at home and the patients undergo more frequent follow-ups.

Conclusion

Patients with RA are vulnerable to postoperative infections and those undergoing spinal surgery are definitely no exception [41,43]. Keeping this in mind, strict infection prevention measures should be taken. Treating physicians should familiarise themselves with the mechanisms leading to a higher infection rate in these patients and should keep in mind that symptoms and signs of a possible infection may be subtle, as described above. Finally, each team treating RA patients should keep a high index of suspicion for infection with even minor relevant symptoms and signs taken into consideration, and a low threshold for proceeding with laboratory or imaging studies.

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