IDCases 33 (2023) e01809

Contents lists available at ScienceDirect

IDCases

journal homepage: www.elsevier.com/locate/idcases

Case report

Recurrent acute-onset of chronic inflammatory demyelinating polyneuropathy after COVID-19 vaccination

Tyler Grantham *, Shahkar Khan, Jai Behgal, Taqi Rizvi, Allison Glaser

Staten Island University Hospital, 475 Seaview Ave, Staten Island, NY 10305, United States of America

ARTICLE INFO

Keywords:
Covid-19
Covid-19 vaccination
Chronic Inflammatory Demyelinating Polyneuropathy
CIDP
CIDP exacerbation

ABSTRACT

This is the case of 54-year-old male with a past medical history of Chronic Inflammatory Demyelinating Polyneuropathy (CIPD) who was found to have an acute exacerbation of CIPD shortly after receiving his 1st COVID 19 booster (3rd dose of vaccination series) and was successfully treated with intravenous immunoglobulin (IVIG) and then was found to have another acute exacerbation of CIDP 6 months later after receiving his 2nd COVID 19 booster (4th dose of vaccination series) that required intubation and long term tracheostomy. CIPD is an acquired immune-mediated polyneuropathy that mainly affects the peripheral nerve roots nerves. It typically presents with relapsing/remitting, or progressive symmetrical muscle weakness and sensory involvement and can cause decreased respiratory effort. COVID-19 is mainly a respiratory disease, but it has been associated with a wide variety of neurological conditions. Although there have been several findings of acute inflammatory demyelinating polyneuropathy in association with COVID-19, CIDP exacerbation as a result of COVID-19 has rarely been seen in the literature. Furthermore, CIDP exacerbation as a result of COVID-19 vaccination is even less frequently seen.

Background

Chronic Inflammatory Demyelinating Polyneuropathy (CIPD) is an acquired immune-mediated polyneuropathy that mainly affects the peripheral nerve roots nerves. It typically presents with relapsing/remitting, or progressive symmetrical muscle weakness and sensory involvement, however, in atypical forms of the disease, it can present with asymmetrical weakness and/or sensory involvement. The disease itself has an immunological basis with the involvement of both the cellular and humoral immunity systems. The diagnosis of CIDP is usually made with the European federation of Neurological Societies and the Peripheral Nerve Society criteria which have both clinical, radiological, and electrodiagnostic criteria [1]. There is no clear genetic cause identified for CIDP.

COVID-19 is mainly a respiratory disease, but it has been associated with a wide variety of neurological conditions. The mild end of the symptom spectrum includes anosmia, ageusia, headache, myalgias. The more severe end of neurological manifestations includes altered consciousness, encephalitis, demyelination, neuropathy, and stroke. In addition, the Astra-zeneca vaccine has been found to be associated with Guillain-Barre Syndrome (GBS) [2]. In this case report, we present a case of a 55 year-old male with known CIDP presenting with signs and symptoms consistent with an exacerbation of his CIDP four days post the 3rd booster shot of the Pfizer Vaccine and then again on the day of the 4th booster.

Case presentation

This is a case of a 54-year-old male with a past medical history of CIDP, Arnold Chiari syndrome type 2, hydrocephalus, spina bifida (myelomeningocele), neurogenic bladder with suprapubic catheter and recurrent urinary tract infection, chronic sacral ulcer, depression, and obsessive-compulsive disorder. The patient was diagnosed more than ten years prior when he presented with bilateral upper and lower limb weakness and pain with waxing and waning symptoms. The patient progressed and became wheelchair-bound with baseline 1–2/5 strength in the lower extremities and 3–4/5 strength in the upper extremities with exacerbations in symptoms every 5–6 months. Electrodiagnostic and nerve conduction studies were consistent with severe multifocal demyelinating motor polyneuropathy with no signs to suggest superimposed myopathic polyneuropathy.

The patient presented to our Emergency Department (ED) for

* Corresponding author.
E-mail address: tgrantham@northwell.edu (T. Grantham).

https://doi.org/10.1016/j.idcr.2023.e01809
Received 13 March 2023; Received in revised form 20 May 2023; Accepted 22 May 2023
Available online 29 May 2023

worsening upper and lower extremity weakness and poor inspiratory effort with negative inspiratory force (NIF) score of 15 cm of water, vitals including heart rate, blood pressure, oxygen saturation were within normal limits, Arterial blood gas (ABG) showed pH 7.49, pCO2 39 and pO2 79. The patient had already completed the Pfizer COVID-19 vaccine series and received the Pfizer booster dose four days prior to initial presentation. This patient had no history of COVID-19 infection. The patient was placed on non0invasive mechanical ventilation and admitted to step-down unit for further management. The patient had a negative nasopharyngeal COVID-19 polymerase chain reaction (PCR) result and denied any fever, cough, chest pain, abdominal pain, nausea, vomiting, diarrhea, and constipation. During this admission, laboratory data revealed a normal complete blood count, basic metabolic panel, and hepatic function panel. CT scan without contrast demonstrated no evidence of intracranial hemorrhage, territorial infarct or mass effect, chronic findings of Arnold Chiari malformation type 2 were seen, which were unchanged from the previous scan. MRI lumbar spine demonstrated no acute changes. The patient was diagnosed with CIDP exacerbation and received intravenous immunoglobulin (IVIG) 400 miligram/kilogram for 5 days, which led to improvement in the weakness of upper and lower limbs, with a return to baseline respiratory function and no longer requiring non-invasive mechanical ventilation. Upon discharge he had an increase in NIF score to 76 cm of water. The patient was discharged with a plan to receive IVIG 1 g/kg every month to maintain remission of CIDP exacerbation.

The patient presented to the ED approximately 6 months later with respiratory distress. The patient had received the second Pfizer COVID-19 booster vaccine earlier that same day. His vital signs were temperature of 102 degrees Fahrenheit, heart rate of 154 beats per minute, blood pressure of 105/55, and respiratory rate of 16, oxygen saturation was 88 % on non-rebreather mask with 15 Liters of minute of oxygen. ABG showed pH 7.37, pCO2 53 and pO2 309. Notable laboratory values on this admission were aspartate transaminase of 160, alanine aminotransferase of 154, alkaline phosphatase of 353, and a urinalysis that was positive for leukocyte esterase and moderate amount of bacteria. CT scan of the Chest, Abdomen and Pelvis revealed bilateral dependent lung dense consolidations and additional scattered patchy ground glass and consolidative opacities suspicious for aspiration pneumonia and osteomyelitis of the right ischial tuberosity underlying chronic sacral decubitus ulcer. Broad spectrum antibiotics were immediately started and the patient was intubated and mechanically ventilated as the patient had persistent desaturations despite increases in oxygen requirements as well as decreased ventilatory drive. The patient was suspected to have an acute flare of CIDP leading to aspiration pneumonia. The neurology service was consulted and recommended IVIG treatment for 5 days. The patient ultimately underwent a tracheostomy for long term mechanical ventilation. The patient's neurological examination improved throughout his hospital stay, and the patient was discharged to a nursing home with mechanical ventilation and continued plan of monthly IVIG infusions.

Discussion

CIDP is often considered to be the chronic counterpart of acute inflammatory demyelinating polyradiculoneuropathy (AIDP), the most common form of GBS. Although similar in presentation and clinical features, CIDP is distinguished from AIDP by chronicity and responsiveness to medical therapy whereas AIDP is self-limited. Typical CIDP is characterized by symmetrical weakness in proximal and distal muscles that develops progressively and increases gradually over 8 weeks or longer, which is necessary to make diagnosis [3]. About one third of patients present with a course that is relapsing and remitting in nature with partial or complete recovery between recurrences [4]. Given the strong response to medical treatment including corticosteroids, intravenous immunoglobulins, and plasma exchange, CIDP is believed to be driven by a humoral immune response [5].

GBS is classically known to be associated with preceding infections such as Campylobacter and Cytomegalovirus (CMV). Additionally, instances of GBS have presented after vaccination. This was initially highlighted after the 1976 swine flu vaccine campaign which showed approximately one additional case of GBS for every 100,000 people who got the swine flu vaccine [6]. Other case reports have shown GBS developing after other vaccinations including influenza, meningococcal and recombinant zoster [7–9]. However, the association has been heavily debated in the medical literature as studies have failed to find an association between vaccination and GBS risk and the overall risk is thought to be small [10].

Although AIDP typically presents with an inciting infection or injury, CIDP has not been strongly correlated with an inciting event and the definitive cause remains unknown. One review article looked at the antecedent events within 1–42 days before onset of CIDP in a cohort of 411 patients from an Italian CIDP database. Infection was reported by 12 % of patients and vaccination was reported by 1.5 % of patients [11]. It has also been reported that patients in CIDP can experience worsening of symptoms after certain vaccines such as the flu vaccine [12].

In the wake of the 2019 coronavirus disease epidemic, GBS has been found to be a relatively common neurological complication of COVID-19. One systematic review showed that there is evidence that suggests COVID-19 may be associated with GBS as it highlighted 73 patients reported across 52 publications that developed GBS following COVID-19 infection. Although most patients showed respiratory or systemic manifestations of COVID-19, asymptomatic cases were also described [13].

CIDP exacerbation as a result of COVID-19 has rarely been seen. There are only 2 documented case reports in the literature that discuss a CIDP exacerbation immediately following COVID-19 infection, both with a history of CIDP diagnosed many years prior [14,15]. Another case report features a case of recurrent GBS likely secondary to COVID-19 infection. The patient had two prior GBS flares from viral illness that were completely resolved, separated by multiple years and subsequently developing a third flare concomitantly with COVID-19 infection [16].

One case study shows an episode of acute-onset CIDP in the setting of COVID-19 infection 7 months prior and COVID-19 vaccination with ChAdOx1 nCoV-19 17 days prior [17]. This was the first documented finding of CIDP exacerbation following COVID-19 vaccination. Our case displays CIDP exacerbation following COVID-19 vaccination, the first occurrence after the third dose in vaccination series and of the BNT162b2 vaccine. Our patient had a previously established diagnosis of CIDP and received the vaccine four days prior to this exacerbation. There was no evidence of recent illness or any other obvious inciting event. The patient was then suspected to have exacerbation on the same day as the fourth dose in the vaccination series, however, given the alternative sources of infection it is difficult to isolate the booster as the clear source. However, the infectious disease service found the osteomyelitis to be chronic in nature and not an acute source of infection. Also, it is thought that the aspiration event may be secondary to the CIDP flare.

It is difficult to say definitively that there is an association between CIDP exacerbation and COVID-19 disease given that CIDP is not strongly correlated with any particular inciting events. There is evidence that vaccination may cause CIDP exacerbation or worsening of symptoms, however, the occurrence is few in numbers and the data is limited. Given the mass vaccination and the coming series of boosters to follow, this is a particular finding of interest. Although speculative, given the recent timing of the patient's vaccine and the presumed humoral based pathogenesis of CIDP, it cannot be conclusively ruled out. More incidence of this finding will have to be present to elucidate a clearer association.

CRediT authorship contribution statement

Tyler Grantham: writing of discussion, editing of paper, data collection, data analysis. **Shahkar Khan:** writing of introduction, data collection, data analysis. **Jai Behgal:** writing of case description, data

collection, data analysis. **Taqi Rizvi:** writing of case description, data collection, data analysis. **Allison Glaser:** Editing of manuscript.

Funding

None.

Ethical approval

Approved.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Declaration of Competing Interest

All authors must disclose any financial and personal relationships with other people or organizations that could inappropriately influence (bias) their work. Examples of potential conflicts of interest include employment, consultancies, stock ownership, honoraria, paid expert testimony, patent applications/registrations, and grants or other funding.

References

[1] Van den Bergh PY, Hadden RD, Bouche P, et al. European Federation of Neurological Societies/Peripheral Nerve Society guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy: report of a joint task force of the European Federation of Neurological Societies and the Peripheral Nerve Society - first revision [published correction appears in Eur J Neurol. 2011 May;18(5):796]. Eur J Neurol. Vol. 13(no. 3); 2010, p. 356–63. ⟨https://doi.org/10.1111/j.1468-1331.2009.02930.x⟩.

[2] Oo WM, Giri P, de Souza A. AstraZeneca COVID-19 vaccine and Guillain-Barré Syndrome in Tasmania: a causal link? J Neuroimmunol 2021;360:577719. https://doi.org/10.1016/j.jneuroim.2021.577719.

[3] Köller H, Kieseier BC, Jander S, Hartung HP. Chronic inflammatory demyelinating polyneuropathy. N Engl J Med 2005;352(13):1343–56. https://doi.org/10.1056/NEJMra041347 [PMID: 15800230].

[4] McCombe PA, Pollard JD, McLeod JG. Chronic inflammatory demyelinating polyradiculoneuropathy. A clinical and electrophysiological study of 92 cases. Brain 1987;110(Pt 6):1617–30. https://doi.org/10.1093/brain/110.6.1617 [PMID: 3427403].

[5] Peltier AC, Donofrio PD. Chronic inflammatory demyelinating polyradiculoneuropathy: from bench to bedside. Semin Neurol 2012;32(3):187–95. https://doi.org/10.1055/s-0032-1329194.

[6] Schonberger LB, Bregman DJ, Sullivan-Bolyai JZ, Keenlyside RA, Ziegler DW, Retailliau HF, et al. Guillain-Barre syndrome following vaccination in the National Influenza Immunization Program, United States, 1976–1977. Am J Epidemiol 1979;110(2):105–23. https://doi.org/10.1093/oxfordjournals.aje.a112795.

[7] Vellozzi C, Iqbal S, Broder K. Guillain-Barre syndrome, influenza, and influenza vaccination: the epidemiologic evidence. Clin Infect Dis 2014;58(8):1149–55. https://doi.org/10.1093/cid/ciu005 [Epub 2014 Jan 9. PMID: 24415636].

[8] Centers for Disease Control and Prevention (CDC). Update: Guillain-Barré syndrome among recipients of Menactra meningococcal conjugate vaccine–United States, October 2005–February 2006. MMWR Morb Mortal Wkly Rep; Vol. 55(no. 13); 2006, p. 364–6. [PMID: 16601664].

[9] Goud R, Lufkin B, Duffy J, et al. Risk of Guillain-Barre syndrome following recombinant zoster vaccine in medicare beneficiaries. JAMA Intern Med 2021;181(12):1623–30. https://doi.org/10.1001/jamainternmed.2021.6227.

[10] Haber P, Sejvar J, Mikaeloff Y, DeStefano F. Vaccines and Guillain-Barré syndrome. Drug Sal 2009;32(4):309–23. https://doi.org/10.2165/00002018-200932040-00005 [PMID: 19388722].

[11] Doneddu PE, Bianchi E, Cocito D, Manganelli F, Fazio R, Filosto M, et al., Italian CIDP Database Study Group. Risk factors for chronic inflammatory demyelinating polyradiculoneuropathy (CIDP): antecedent events, lifestyle and dietary habits. Data from the Italian CIDP Database. Eur J Neurol 2020;27(1):136–43. https://doi.org/10.1111/ene.14044 [Epub 2019 Aug 9. PMID: 31325350].

[12] Kuitwaard K, Bos-Eyssen ME, Blomkwist-Markens PH, van Doom PA. Recurrences, vaccinations and long-term symptoms in GBS and CIDP. J Peripher Nerv Syst 2009;14(4):310–5. https://doi.org/10.1111/j.1529-8027.2009.00243.x [PMID: 20021573].

[13] Abu-Rumeileh S, Abdelhak A, Foschi M, Tumani H, Otto M. Guillain-Barré syndrome spectrum associated with COVID-19: an up-to-date systematic review of 73 cases. J Neurol 2021;268(4):1133–70. https://doi.org/10.1007/s00415-020-10124-x [Epub 2020 Aug 25. PMID: 32840686; PMCID: PMC7445716].

[14] Abu-Rumeileh S, Garibashvili T, Ruf W, Fangerau T, Kassubek J, Althaus K, et al. Exacerbation of chronic inflammatory demyelinating polyneuropathy in concomitance with COVID-19. J Neurol Sei 2020;418:117106. https://doi.org/10.1016/j.jns.2020.117106 [Epub 2020 Sep 2. PMID: 32911389; PMCID: PMC7462872].

[15] Van Looy E, Veenker L, Steyaert A, Leenders J, Malfroid G, De, et al. COVID-19-induced exacerbation of chronic inflammatory demyelinating polyneuropathy. J Neurol 2021;268(9):3129–31. https://doi.org/10.1007/s00415-021-10417-9 [Epub 2021 Feb 19. PMID: 33606069; PMCID: PMC7892325].

[16] McDonnell EP, Altomare NJ, Parekh YH, Gowda RC, Parikh PD, Lazar MH, et al. COVID-19 as a trigger of recurrent Guillain-Barré syndrome. Pathogens 2020;9(11):965. https://doi.org/10.3390/pathogens9110965 [PMID: 33228253; PMCID: PMC7699516].

[17] Suri V, Pandey S, Singh J, Jena A. Acute-onset chronic inflammatory demyelinating polyneuropathy after COVID-19 infection and subsequent ChAdOx1 nCoV-19 vaccination. BMJ Case Rep 2021;14(10):e245816. https://doi.org/10.1136/bcr-2021-245816 [PMID: 34607818; PMCID: PMC8491284].