TITLE

Post Polio Syndrome: A Case Report

ABSTRACT

Post Polio Syndrome (PPS) is a sequelae of poliovirus infection that causes weakness in previously infected polio patients. The diagnosis is one of exclusion and includes the following: 1) a prior episode of poliomyelitis with residual motor neuron loss, 2) a period of at least 15 years or more after the acute onset of polio with neurologic and functional stability, and 3) a gradual onset of new weakness and abnormal muscle fatigability that has persisted for at least one year. While the exact etiology is unknown, the prevalence of PPS has increased as patient's who have previously survived polio are getting older [6]. In this report, we will examine a patient coming to his primary care provider for evaluation of worsening weakness over the past 3 years and discuss general characteristics, general evaluation and potential treatment plans. The goal of this paper is to shine a light on the need for further research and investigation in a growing patient population.

INTRODUCTION

Poliomyelitis is an infection of the motor neurons in the anterior horn of the spinal cord and brain stem caused by the poliovirus. The polio virus is a neurotropic enterovirus that, while eradicated in most parts of the world, is still endemic throughout South Asia. The subject of this paper is Post Polio Syndrome (PPS), a potential sequelae of the poliovirus that is becoming more apparent as previously infected individuals are getting older [7]. PPS diagnosis requires the following: 1) a prior episode of poliomyelitis with residual motor neuron loss, 2) a period of at least 15 years or more after the acute onset of polio with neurologic and functional stability, and 3) a gradual onset of new weakness and abnormal muscle fatigability that has persisted for at least one year. The diagnosis of PPS is also one of exclusion, meaning that this diagnosis may only be considered after having ruled out more common causes of muscle weakness [9]. The main cause of PPS is still widely unknown. Multiple studies have looked at the potential causes including reactivation of a latent form of the virus, progression of motor neuron degeneration and induction of autoimmunity causing inflammation [3-4]. However, these investigations have all been found to be inconclusive to this point.

CASE

The patient is a 61 year old male with a past medical history of poliovirus infection, BPH, and hyperlipidemia that is coming in for worsening weakness in his legs. When he was 2 years old living in Pakistan, the patient was diagnosed with polio. His family told him that he had flu-like symptoms, including fever and fatigue, along with muscle weakness. He says that weakness was mostly in his legs when he was younger, but did not notice it after he was 5. The patient noticed weakness again in his legs bilaterally that began 3 years ago. He feels that the weakness has been increasing ever since then. There was no inciting event that he can recall. He has experienced falls over the past 3 years but stated the falls were only after his weakness got worse, which is why he began using a cane. He denied specific trauma to his lower back or pain in that area. The patient reports trouble getting up from a chair and sitting back down. He also noticed that he gets more tired after standing for long periods of time. He says that he is able to walk but he uses a cane which he did not bring with him to the clinic today. He denies pain in any area, fever, chills, nausea, vomiting, diarrhea, constipation, loss of sensation in any area, paresthesia, chest pain or SOB. He has never been hospitalized and the only procedure he underwent was a

colonoscopy which was normal. He denies tobacco use, alcohol use and illicit drug use. He is sexually active with his wife and has never had a previous diagnosis of a STD. Family history significant for type 2 diabetes in his mother and unspecified heart disease in his father. Physical exam was significant for limited ambulation, waddling gait, hypotonicity in lower extremities bilaterally except for feet, ⅕ strength in bilateral hips and knees with atrophy of quadriceps and calves bilaterally, deep tendon reflexes absent in lower extremities bilaterally and waddling gait. Strength and reflexes intact with normal findings in upper extremities bilaterally. CN intact. Remainder of the physical exam was normal. The patient's CBC from the previous visit had been normal aside from slightly elevated LDL cholesterol. Patient has lumbar X ray from a previous encounter with an internal medicine doctor who was evaluating his weakness. X ray showed prominent levoscoliosis, mild degenerative disc disease and reverse spondylolisthesis of the L3-L4 region. The patient was given a referral for neurology in order for full evaluation and an MRI, EMG and nerve conduction study test were ordered to rule out other potential causes of his weakness. A referral to PM&R was also ordered due to the high likelihood that the patient will require intensive rehabilitation. Given the patient's socioeconomic status and other social determinants of health, he has been unable to fulfill the order thus far.

DISCUSSION

Based on review from current literature, PPS is a very rare side effect of poliovirus infection. It occurs in approximately 20-30% of polio survivors [1]. There is an absence of reinnervation of the anterior horn cells caused by the initial polio infection. As mentioned in the introduction, there are multiple theories investigating the cause for PPS.

One of the theories resembles the same pathophysiology as shingles. It is believed to be a reactivation of the virus causing further degeneration of motor neurons and preventing reinnervation. The virus does not necessarily reactivate in everyone diagnosed with polio but it has never appeared in patient's who have been vaccinated and protected against it. However, patients that have been vaccinated via OPV that have experienced symptoms of polio should be monitored for recurrence of the disease as well. It stands to reason that the potential for PPS is high if the weakened virus from OPV was strong enough to cause an initial infection to begin with [1]. The other theories include an autoimmunity affecting the motor neurons triggered by the previous poliomyelitis. The evidence supporting this includes the presence of elevated protein and oligoclonal bands seen in CSF in some of the PPS patients [5]. The last major theory surrounding PPS is that it is the progression of motor neuron degeneration originating from the time of the original infection. All three of these theories lack sufficient evidence for the medical community to confidently attribute the cause of PPS to any one of these mechanisms specifically [11].

Our patient has not gone under specific testing in order to rule out more common diagnoses such as ALS, polyneuropathies, etc. Therefore, our patient could not be formally diagnosed with PPS yet. However, when taking into consideration the patient's history, physical exam findings and progression of symptoms, PPS appears to be the most likely diagnosis at this time. The recommendation for the patient was for further evaluation from another institution with the necessary equipment i.e. nerve conduction studies, EMG and MRI. The MRI is used to rule out compression of the spinal cord. The lack of evidence of sensory neuropathy means that the spondylolisthesis is less likely the cause of the patient's weakness. In addition, the spondylosis noted in the X ray would also present with some form of back pain with potential radiation of the pain down the legs. However, the patient is strictly complaining of weakness.

The CBC ordered in the clinic was mostly normal aside from elevated LDL cholesterol. There is no evidence of a chronic infection due to a lack of an elevated white count.

A focus on the differentials should be considered when examining a patient with suspected PPS. The first is ALS. The patient's symptoms do not appear to line up with the classic presentation of ALS given that he does not have a constant linear progression of symptoms and the weakness has not moved past his legs in the past 3 years. Further investigation with nerve conduction studies to evaluate for nerve potentials is required to truly rule out ALS [5]. There is also a lack of upper motor neuron signs in this patient which decreases the likelihood that the patient's condition is ALS. The other condition high on the differential is lumbar spondylolisthesis causing compression of the motor nerve roots. Patients can experience weakness in their legs but they also typically have sensory symptoms as well. They would also experience relief with lumbar flexion and worsening of their symptoms with lumbar extension [5]. However, this patient did not feel relief with any positional changes. The last diagnosis to consider on the differential is another form of demyelinating polyneuropathy. As mentioned in the evidence above, this differential was ruled down due to the presentation of the patient's case. However, much like ALS, a nerve conduction study is needed to properly evaluate the conduction velocity. The patient's lab values and clinical presentation make the cause being myositis or hypothyroidism less likely than PPS.

The management of PPS includes aerobic intensity exercise along with physical exercise using work-stop programs. The evidence of these programs are consolidated to several small uncontrolled studies due to the small patient population and lack of funding and research in this field [2]. In addition, due to the prevalence of polio in certain endemic areas seen in Figure 2. It is important to globally promote and secure access for inactivated vaccines for all conditions, including the salk vaccine (the inactivated version of the polio vaccine) [8,10]. The effectiveness of three doses of the polio vaccine is 99-100% according to the latest studies conducted by the CDC [1]. Eradication of polio globally will subsequently eradicate the condition known as PPS.

CONCLUSION

This case brings to light a vulnerable population that has not been discounted due to the virtual eradication of the parent condition. Despite polio no longer being a threat to a majority of the world, those who have previously been infected are surviving and 20-30% are experiencing this rare sequelae [1]. PPS causes muscle weakness and atrophy in individuals that greatly impact their quality of life without the proper diagnosis and treatment. If providers ignore the prevalence of PPS, they are ignoring a growing population of patients due to the survival rate of those previously diagnosed with polio. This is best exhibited by the decrease in death rate in polio seen in Figure 1. Not only is it important to recognize these patients, but to promote preventive medicine through vaccines. Vaccination has become a wide debate with the COVID-19 pandemic and people's views have become polarized. However, the public should not only focus on the acute condition, but the risk of long term effects of these conditions as well. This is why this case should not only serve as a signal for more research on a growing PPS patient population, but also on the necessity for vaccines as protection in the short and long run.

APPENDIX

Figure 1 [12]

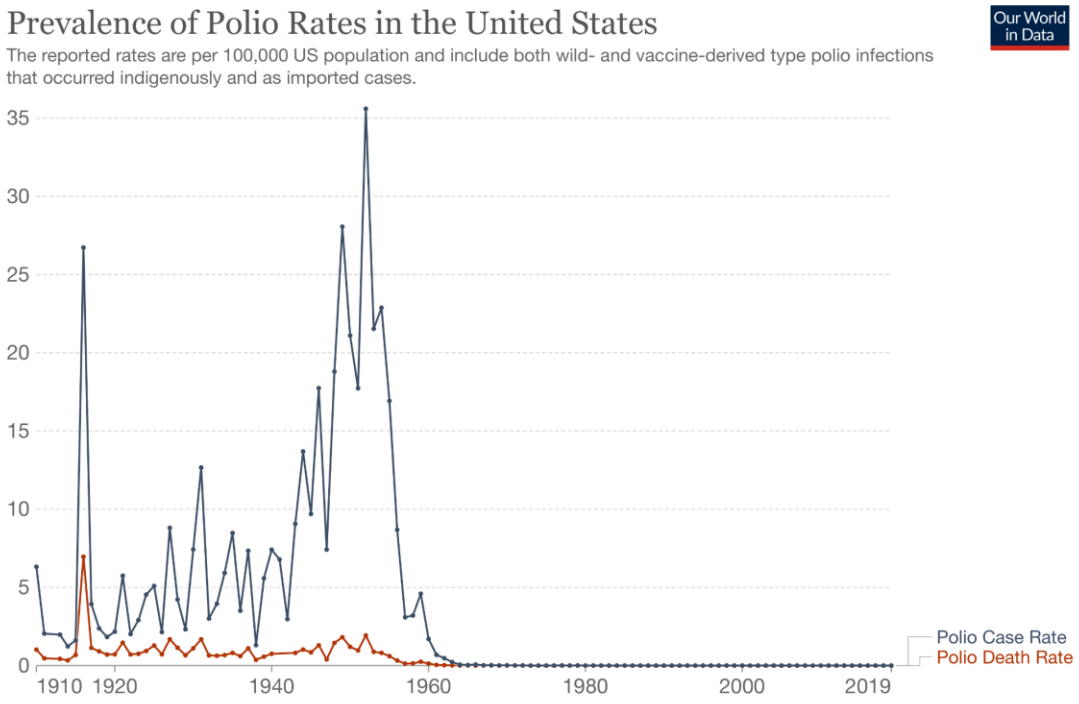
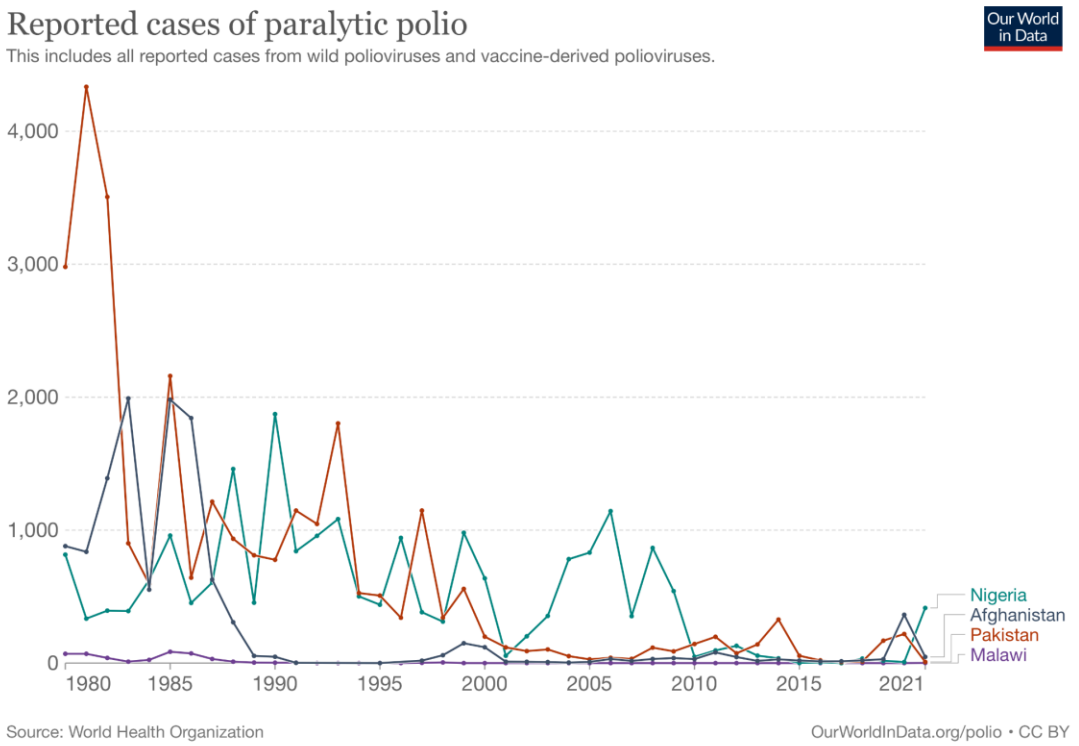

Prevalence of Polio Rates in the United States
The reported rates are per 100,000 US population and include both wild- and vaccine-derived type polio infections that occurred indigenously and as imported cases.
Our World in Data
35
30
25
20
15
10
5
0
1910
1920
1940
1960
1980
2000
2019
Polio Case Rate
Polio Death Rate
Source: Our World In Data based on US Public Health Service; US Center for Disease Control; and WHO
OurWorldInData.org/polio/ • CC BY

Figure 2 [12]


Reported cases of paralytic polio
This includes all reported cases from wild polioviruses and vaccine-derived polioviruses.
Our World in Data
4,000
3,000
2,000
1,000
0
1980
1985
1990
1995
2000
2005
2010
2015
2021
Nigeria
Afghanistan
Pakistan
Malawi
Source: World Health Organization
OurWorldInData.org/polio • CC BY

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