

## Brachial plexitis: an unusual presentation in sickle cell disease

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### ABSTRACT

Brachial plexitis is defined as an inflammation of the brachial plexus. There are two entities of the disease: idiopathic, which is generally considered to be immune-mediated, and genetic. The disease manifests as the acute onset shoulder pain, weakness of the involving arm ± sensory loss. Brachial plexitis is also known as Parsonage–Turner syndrome and hereditary neuralgic amyotrophy. Diagnosis is made with the help of history, physical exam, and imaging. Conservative management is the mainstay of treatment. There has not been any proven treatment for the condition though some cases have been treated empirically with steroids. We present a case of 61-year-old woman with sickle cell anemia who presented with right upper extremity weakness and MRI findings of brachial plexitis.

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Brachial plexitis; sickle cell disease; lower motor neuron

## 1. Introduction

The brachial plexus is a network of nerves that control the sensory and motor functions of the upper extremity. It is formed by the lower four cervical nerve roots (C5–C8) and first thoracic nerve root (T1). It is subdivided into root, trunks, division, and cords. Brachial plexitis is characterized by the acute onset of motor ± sensory loss of one or more nerves originated by the brachial plexus. It is described with different nomenclature as Parsonage–Turner Syndrome, hereditary neuralgic amyotrophy, acute brachial plexitis, idiopathic brachial plexitis, idiopathic brachial neuritis, paralytic brachial neuritis among others [1]. We present a case of 61-year-old woman with sickle cell disease (SCD) who presented with the acute onset of right upper extremity weakness and sensory loss that was diagnosed with brachial plexitis with the help of MRI and NCS/EMG studies.

## 2. Case presentation

This is a case of 61-year-old right-handed Caucasian woman with a known history of HbSS disease, being diagnosed on Gene testing, presented to the hospital with the complaint of acute onset of right upper extremity weakness and loss of sensation. Physical exam was remarkable for complete plegia in the flexors and extensors of the right arm, forearm, wrist, and fingers. There was also no volitional movement in the abductors and adductors of the right shoulder and fingers. Scapular winging was evident on the right side. There was loss of pinprick, light touch, vibration, and proprioception in all dermatomes on the right upper extremity. Biceps, triceps, and brachioradialis reflexes were absent, even

with re-enforcement. The remainder of the neurologic exam was unremarkable. Investigations showed for Hemoglobin of 7.4 gm/dl, Hematocrit: 23.0, and platelet count: 143. Reticulocyte count: 1.10 and peripheral blood film showed sickle RBC, rest was unremarkable. Other investigations were unremarkable including ESR, CRP, and autoimmune profile including ANA, ANCA, anti-ds-DNA, RF, anti-Ro, anti-La, lupus anticoagulant, anticardiolipin Ab, complement levels. Lyme disease, syphilis, hepatitis E, and HIV were also ruled out.

MRI cervical spine and chest showed diffuse inflammation of the brachial plexus as shown in [Figure 1](#). Electromyography and Nerve conduction studies confirmed the diagnosis of brachial plexitis. After ruling out sickle cell crisis, which was diagnosed clinically, patient was discharged home on tapering steroid treatment with outpatient follow-up. Patient returned for follow-up after multiple attempts of reaching her. On follow-up visit, her neurologic functions were back to normal as power of 5/5 in all muscle groups in the right upper extremity, sensations intact to pinprick, temperature, vibration, and deep tendon reflexes of 2+ in biceps, triceps, and brachioradialis.

## 3. Discussion

Brachial plexitis is an uncommon cause of upper extremity weakness. The incidence is 1.64 cases among 100,000 with a male predominance. It is usually divided into two categories as either idiopathic or hereditary. Twenty-five percent to 55% cases are preceded by viral or bacterial infections. Commonly associated infections include smallpox, *Borrelia burgdorferi*, coxsackievirus, influenza, typhoid, HIV, and CMV. Fifteen percent of



**Figure 1.** Inflammation of the brachial plexus shown as hyperintense areas as pointed by arrows.

the cases have reported recent history of vaccination [1]. Other causes include trauma, post-surgical, pregnancy, and strenuous exercise [1]. A case has been reported with herpes zoster infection [2]. Radiation has also been reported as an attributed factor as 25 cases reported during the period of 1967–1980 [3]. The hereditary form is an autosomal dominant condition and believed to be due to deficiency in the protein septin family [1]. The most common initial symptom is the acute onset of shoulder-based pain which is present in 95% followed by weakness. Weakness either coincides with the onset of pain or occurs in a delayed fashion. It is unilateral in 66% of the cases and 54% involve the right side. Most common muscles group involved are deltoid, biceps, triceps, spinatus, and serratus anterior. Sensory symptoms are present in 78% of the cases of which paresthesia and hypoesthesia are the most common [1]. The condition has also been described in the pediatric population with a similar presentation as in adults [4]. In children, acute flaccid myelitis may mimic brachial plexitis clinical manifestations.

Brachial plexitis is a diagnosis of exclusion. It is diagnosed with the help of history, physical exam, imaging, and EMG studies. There is no laboratory test to diagnose the condition. In the acute phase, MRI shows hyperintensities in the T2 image as a consequence of edema from nerve demyelination. Electromyography is the best study to demonstrate demyelination and to be performed after 3 weeks of symptoms onset to demonstrate abnormality [1]. According to the American College of Radiology appropriateness criteria, MRI brachial plexus with and without IV contrast is usually the appropriate test in patient with acute or chronic, non-traumatic, or traumatic, with or without the history of malignancy. In patients with a history of malignancy are

unable to undergo MRI, FDG-PET scan is the next best study while CT scan can be performed in other patient population [5]. CSF studies are nonspecific for the diagnosis of brachial plexitis [6].

Treatment of brachial plexitis requires a multidisciplinary approach involving physical therapy, pharmacologic, non-pharmacologic, and in some case even surgical cases. *Cochrane review* provides no treatment to improve the neurologic outcome and prognosis of the disease [7]. *Van Alfen and Van Engelan* suggested early recovery with 2 weeks of oral steroid treatment with taper. The steroids were used at 1 mg/kg dose [8] as treated in our patient. Pain in some cases is treated with the non-inflammatory, anti-inflammatory drugs, or opioids. Patients with the history of recent viral infection or postherpetic neuralgia should be treated with antivirals [8]. A recent randomized controlled trial on neuropathic pain outlined the evidence-based algorithm which suggests tricyclic antidepressants (amitriptyline, nortriptyline, desipramine, imipramine), opioids (CR oxycodone, methadone, morphine), tramadol, gabapentin, and pregabalin are all beneficial in providing pain relief [9]. *Kim JG and Chung SG* reported a case of herpetic brachial plexopathy treated with the ultrasound-guided corticosteroid injection [2]. In refractory cases, surgical treatment includes neurolysis, nerve grafts, and nerve transfer [7].

SCD is known to cause neurological deficits. The most common being symptomatic strokes or silent infarcts. While rare, peripheral nerve involvement has also been involved. This is usually due to vaso-occlusion of the vessels leading to neurological deficits and pain. There are limited case reports describing SCD associated neuropathies and therefore treatment. In one report of a young woman presenting with flaccid paralysis and sensory loss of her left

lower extremity [10], patient was treated with exchange transfusion therapy and symptoms resolved in 48 h.

Our case presents a 61-year-old woman with complete plegia of the Right arm and sensory loss. Our patient presented with the complete involvement of the brachial plexus and there was no preceding viral infection in her case. As *van Alfen and Van Engelen* suggested 2 weeks of oral steroid course taper [8], we treated our patient with steroid as well though the challenging part was the history of SCD. Steroids are known to precipitate sickle crisis in sickle cell patients. Hemoglobin electrophoresis was performed which demonstrated no crisis stage and patient was discharged with steroids with close outpatient follow-up with neurologist and hematologist. Outpatient physical therapy and occupational therapy were prescribed as well.

#### 4. Conclusion

As per the available data, there has been a good understanding of the disease process, but the treatment options are mainly limited to physical therapy and pain management. Different treatment options as oral steroids, or even intraarticular steroids have been tried but none has been studied to enough extent.

#### Disclosure statement

No potential conflict of interest was reported by the authors.

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