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Case Report

Primary CNS Lymphoma Presenting with Brachial Plexopathy

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Abstract

A patient had a traumatic right rotator cuff, few months later presented with weakness. Brachial plexopathy was diagnosed based on electrodiagnostic studies. Further investigations discovered a primary CNS lymphoma.

Keywords: *CNS Lymphoma; Brachial Plexopathy*

Introduction

- Optimal assessment of brachial plexopathy requires a thorough history for the precise localization and subsequent investigations with electrodiagnostic studies to reach the accurate diagnosis.
- Electrodiagnostic evaluation of a plexus injury should be systematic and comprehensive to provide the maximum precision for localization, extent of involvement and severity of injury.
- It is important to emphasize on the screening for malignancy in plexopathies as it is one of the differential diagnoses.
- The ultrasound is an alternative imaging modality for evaluation of plexus and roots if MRI can't be obtained.

Case Report

A 61 -year-old gentleman presented with a gradual onset of right upper extremity weakness eight weeks prior to presentation. The Weakness continued to worsen progressively with difficulties dressing himself initially and became completely disabled and has no functional movement of his right upper extremity from shoulder down. It was associated with numbness in the right thumb, index and middle fingers with some shooting numbness from his elbow down to the wrist on the dorsal aspect of the forearm. His symptoms started almost 6 months after his initial traumatic injury to his right shoulder that resulted in a rotator cuff tear. There was no neck pain or radicular type pain. There was no history of flu-like symptoms prior to the onset of his symptoms. He did not have any other neurologic signs or symptoms in other limbs. He did not have speech difficulties or any memory problems.

His past medical history is significant for diabetes mellitus, hypertension and ischemic heart disease with implantable cardiac defibrillator.

His examination showed a normal mental and language functions. The cranial nerves were intact except for a right Horner's syndrome. The muscle bulk was significantly decreased in his right upper extremity with wasting of his forearm muscles, biceps and triceps. There was a significant shoulder girdle muscle wasting including some aspects of the pectoralis, supraclavicular, infraclavicular, trapezius and rhomboid muscles. There was a flaccid weakness of the right upper extremity and he was able to wiggle his fingers and his index, middle and ring fingers with almost no activity at the little finger. There was an impaired sensory function in the right upper extremity affecting his entire hand and arm up to his shoulder with some relative sparing on the upper outer aspect of his arm. His reflexes were absent in the right upper extremity and normal in the left arm and in the bilateral legs.

He has had electrodiagnostic studies performed three times, initially it showed denervation changes affecting all muscles distal to the right shoulder joint with possible relative sparing of the lower trunk. A follow-up study revealed significant denervation changes in all muscles tested in the right upper extremity with some evidence of denervation in shoulder girdle muscles as well as in the right trapezius muscle which suggested a poly neuritis that spread further than the brachial plexus. The CT of the cervical spine showed only moderate left-sided neuroforaminal stenosis at C3 - C4 and C4 - C5. The Ultrasound of the right brachial plexus revealed a diffuse right brachial plexopathy with relative sparing of the C8 nerve root. The nerves were diffusely thickened in C5, C6, C7 and C8 as well as T1 when compared to the opposite side. Because of his previous ICD the MRI could not be obtained. The Ultrasound of the right shoulder showed a full thickness tear of the subscapularis, supraspinatus and infraspinatus tendons. There was fatty infiltration of both supraspinatus and infraspinatus and teres minor muscles.

The CSF study was sent and showed an elevated protein and white cells with the lymphocytic predominance at 79%. The CSF cytology showed occasional small cells with atypical indented or cleave nuclei in lymphocytes. Spinal fluid flow cytometry showed clonal B cells. The bone marrow aspirate and biopsy were negative. A bone scan and SPECT study revealed an abnormal blood pooling and hyperaemia in right arm, wrist and almost all the small joints of the right hand. He has had a CT of his chest, abdomen and pelvis with contrast which revealed significant atrophy and fatty infiltration in the right shoulder and upper hemithorax. There was no overt masses or lymphadenopathy that was seen. The viral serology was nonreactive for hepatitis B or C and HIV was negative.

Based on CSF study a primary CNS lymphoma versus secondary lymphoma with neurological involvement was suggested. To clarify this a restaging with CT chest, abdomen, pelvis and head and neck was repeated which did not show any masses or systemic lymphadenopathy to suggest the patient's symptoms.

The patient was diagnosed with primary CNS lymphoma. He was treated with four cycles of MATRix chemotherapy and achieved a complete clinical remission with normal CSF, bone scan and restaging CT scans. He was left with the right upper extremity weakness and followed in the hand clinic.

Muscle (Right)	SA	MUP configuration	R
Rhomboids	2+	0	0
Trapezius	2+	Large	Mildly reduced
Supraspinatus	2+	0	0
Infraspinatus	2+	0	0
Deltiod	3+	0	0
Biceps	3+	0	0
Triceps	2+	Large	Mildly reduced
Pronator teres	3+	Large	Mildly reduced
EDC	2+	Large	Severely reduced
EIP	2+	Large	Mildly reduced
FDI	2+, CRD	Large	Moderately reduced
APB	3+	Large	Moderately reduced
Brachioradialis	3+, CRD	0	0
ADM	3+	Large	Moderately reduced
Pectoralis Major	3+	Large	Mildly reduced

Table 1: *Needle EMG study.*

APB: Abductor Pollicis Brevis; ADM: Abductor Digit Minimi; EDC: Extensor Digitorum Communis; EIP: Extensor Indicis Proprius; FDI: First Dorsal Interosseous; SA: Spontaneous Activity; MUP: Motor Unit Potential; CRD: Complex Repetitive Discharge; R: Recruitment Patterns.

CMAP	DL (ms)	A(mV)	CV(m/s)
R Median (APB)	3.67	8.7	45.8
R Ulnar (ADM)	2.4	10	51
R Radial (EIP)	2.67	5	46.2
L Median (APB)	3.4	9.6	50

SNAP (antidromic)	DL (ms)	A(pV)	CV(m/s)
R Median (Dig II)	2.67	5.9	52.5
R Ulnar (Dig V)	2.5	50	56
R Radia (Dig I)	-	-	-
R LAC	1.6	8.54	62
R MAC	1.6	11.8	59.4

Table 2: *Motor and sensory nerve conduction studies.*

CMAP: Compound Muscle Action; SNAP: Sensory Nerve Action Potential; A: Amplitude; CV: Conduction Velocity; DL: Distal Latency.

Discussion

In our patient the trauma caused the pain and the rotator cuff syndrome. Months later, he started to have the upper extremity weakness which lead to the diagnosis of brachial plexopathy based on the presentation and electrodiagnostic studies and confirmed by the ultrasound of the involved plexus. Further investigations including CSF studies lead to the diagnosis of primary CNS lymphoma.

Brachial plexopathy is a term used to describe dysfunction of the brachial plexus. Symptoms of brachial plexopathy may include pain, motor deficits and/or sensory deficits in the distribution of the nerves that comprise the brachial plexus (C5 - T1), that is, shoulder, arm, wrist and hand. On physical examination, atrophy and hyporeflexia may also be present in addition to weakness and sensory loss [1]. Common causes of brachial plexopathy include trauma, tumors and its associated complications [2] (i.e. Pancoast tumor), infiltration or inflammation (i.e. brachial plexitis or Parsonage-Turner syndrome) and exposure to toxins. In addition to a comprehensive clinical evaluation, optimal assessment of the brachial plexopathy requires the performance of ancillary studies. Of these, electrodiagnostic examination is by far the most helpful. Although an extension of the neurologic examination, it has several advantages over the latter, including the ability to localize and characterize the lesion, evaluate muscles not easily assessed clinically (e.g. anconeus), recognize minimally affected muscles that seem normal clinically, prove continuity when visible muscle movement is lacking, recognize remote lesions no longer appreciable clinically and estimate lesion severity for current and future comparative studies. By integrating requisite anatomic, pathophysiologic and neuromuscular knowledge with detailed clinical assessment and the results of ancillary studies, the examining physician can make an accurate diagnosis and prognosis. The lesion must be localized and characterized. This ability requires an understanding of the relevant anatomy, as well as a familiarity with disorders affecting the brachial plexus [3].

Electrodiagnostic evaluation of a plexus injury should be systematic and comprehensive to provide the maximum precision for localization, extent of involvement and severity of injury [4].

The exact number of brachial plexus injuries that occur each year is difficult to ascertain; however, with the advent of increasingly extreme sporting activities and high energy motor sports, as well as the increasing number of survivors of high-speed motor vehicle accidents, the number of brachial plexus injuries continues to rise throughout the world. Most of these injuries occur in males aged 15 to 25 years. Based on his experience with 1,068 patients with brachial plexus injuries during an 18-year span, Narakas developed his rule of "seven seventies". He reported that approximately 70% of traumatic brachial plexus injuries occurred secondary to motor vehicle accidents; of these, approximately 70% involved motorcycles or bicycles. Of the cycle riders, approximately 70% had multiple injuries.

Overall, 70% had supraclavicular lesions; of those, 70% had at least one root avulsed. At least 70% of patients with a root avulsion also have avulsions of the lower roots (C7, C8, or T1). Finally, of patients with lower root avulsion, nearly 70% will experience persistent pain [5].

In a prospective study showed that 1 in 4 patients who had rotator cuff tears with shoulder muscle atrophy on presentation had an associated peripheral neuropathy of the severe degrees of atrophy may be more likely to have an associated peripheral neurologic injury. The most common lesion was brachial plexopathy for a prevalence of 16% [6]. All patients should undergo careful neurologic screening as an essential part of their workup. In selected patients in whom there is suspicion for neurologic injury, electrodiagnostic testing is warranted when the diagnosis remains in doubt [6].

It is known that the MRI is the mainstay of plexus imaging, providing superior definition of features of intraneural anatomy as well as localizing pathologic lesions in conditions where electrophysiologic and physical findings are nonspecific [7]. Only in the past decade has neuromuscular ultrasound been incorporated into the diagnosis and characterization of brachial plexus pathology [8]. When used in conjunction with electrodiagnosis, may mitigate these diagnostic dilemmas [8]. It replaced the MRI in our case.

Primary CNS lymphoma is an uncommon, aggressive non-Hodgkin lymphoma confined to the CNS in the absence of systemic disease. Diagnosis can be made by brain biopsy, CSF analysis, or vitreous fluid analysis. Primary CNS lymphoma is typically diffuse large B cell in histology. Evaluation of extent of disease should be performed before initiation of therapy. Initial induction treatment includes high-dose methotrexate-based chemotherapy. Several studies have demonstrated improved outcome using consolidative whole-brain radiation therapy or high-dose chemotherapy and autologous stem cell transplantation. Neurologic complications can result from direct or indirect effects of leukemia and lymphoma or may be treatment-induced [9]. In the literature review, few reported cases with primary CNS lymphoma progressed to brachial plexopathy due to either chemotherapy or radiotherapy or both [10].

Conclusion

Optimal assessment of brachial plexopathy requires a thorough history for the precise localization and subsequent investigations with electrodiagnostic studies to reach the accurate diagnosis. Electrodiagnostic evaluation of a plexus injury should be systematic and comprehensive to provide the maximum precision for localization, extent of involvement and severity of injury. It is also important to emphasize on screening for malignancy in plexopathies as it is one of the differential diagnoses. The ultrasound is an alternative imaging modality for evaluation of plexus and roots if MRI can't be obtained.

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