Clinico-Laboratory Characteristics and 120 Days Outcome of CNS Vasculitis among Systemic Lupus Erythematosus Patients Admitted to a Tertiary Care Hospital: A Real-Life Experience

Presenter:

Dr. Md. Moniruzzaman

Junior Consultant (Medicine)

National institute of neurosciences and hospital, Dhaka

Background

- Systemic lupus erythematosus (SLE) is an autoimmune, chronic inflammatory disorder affecting multiple organs including the central nervous system (CNS). However, the involvement of medium to small cerebral arteries like the anterior, middle, posterior, and its branches in the CNS is rare, less than 1% of SLE[1]

1.Rodrigues M, Galego O, Costa C, et al. Central nervous system vasculitis in systemic lupus erythematosus: a case series report in a tertiary referral center. Lupus. 2017;26(13):1440-1447.

- CNS vasculitis is one of the catastrophic complications of SLE that may present like stroke, seizure, cranial nerve palsy, altered sensorium, psychosis, and cognitive dysfunction
- Various pathological mechanisms like an immune complex deposition, thrombosis, and accelerated atherosclerosis lead to ischemia or micro-hemorrhage in the brain parenchyma

1.Rodrigues M, Galego O, Costa C, et al. Central nervous system vasculitis in systemic lupus erythematosus: a case series report in a tertiary referral center. Lupus. 2017;26(13):1440-1447.

Rationale

- Diagnosing CNS vasculitis in SLE can be difficult due to its rarity and atypical presentation. It affects small to medium-sized vessels in the brain, spinal cord, and cranial nerves, leading to distinct neuroimaging characteristics.
- Diagnosing neuropsychiatric manifestations in SLE is important as those frequently occur in childbearing age, have high disease severity, are difficult to treat, and carry a poor prognosis
- We present our clinical experience for a better understanding of clinical presentations, imaging characteristics, and treatment outcomes of CNS vasculitis

Methods

Study Design: Prospective

Number of participants: Seventeen (17)

Study Center: National Institute of Neurosciences and Hospital, Dhaka

Inclusion Criteria: All confirmed cases of SLE who had Neuropsychiatric manifestations

ACR/EULAR nomenclature of CNS lupus 2003 [2]

Classification Criteria 2019 for SLE [3]

Objective:

To describe clinical and lab findings, radio-imaging characteristics, and treatment outcome at 120 days from enrollment

2.Nived O, Sturfelt G, Liang MH, De Pablo P. The ACR nomenclature for CNS lupus revisited. Lupus. 2003;12(12):872-876.
3.Aringer M, Costenbader K, Daikh D, et al. 2019 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Systemic Lupus Erythematosus. Arthritis Rheumatol. 2019;71(9):1400-1412.

Results

- Total patients 17
- Previously diagnosed cases were 6
- 4 were found in partial remission before the new clinical presentation.
- The mean age (±SD) of the patients was 24.8 (±8) years, and 15 were female (F:M- 7.5:1)
- The median (IQR) duration of SLE was 3 (1-4.7) months.
- Upon admission, the patient's SLEDAI (SD) score was 24 (9)

Demographic and Clinical Characteristics

Trait	Frequency (%)
Age (mean, SD)	24.8 (8) years
Sex	
Female	15 (88.2%)
Male	2 (11.7%)
Headache	11 (64.7%)
Convulsion	7 (41.2%)
Hemiparesis	4 (23.6%)
Paraparesis	1 (5.8%)
Psychosis	3 (17.6%)
Chorea	1 (5.8%)
Altered level of consciousness	1 (5.8%)
Papillitis	2 (11.7%)
Retinal vasculitis	1 (5.8%)

Laboratory characteristics

Trait	Mean (SD) / Median [IQR]
Hb (gm/dl)	10.7 (2.2)
Neutrophil count (/mm3)	5418 (1743)
Platelet (/mm3)	127757 (54570)
ESR (mm in 1st hour)	53 (22)
Creatinine (mg/dl)	0.9 (0.3)
SGPT (U/L)	38 (11)
Random glucose (mmol/l)	7.3 (3.4)
APTT (second)	35.3 (5.7)
D-dimer (<0.5 ng/dl)	1.3(0.4)
Anti-dsDNA (<25IU/ml)	63.40 [40.1-105.8]
Complement 3 (0.7-1.7 g/L)	0.17 [0.06-0.35]
Complement 4 (0.15-0.45 g/L)	0.41[0.17-0.93]

All patients have ANA (+) with titer >1:80, cytoplasmic pattern,

- **Negative lupus anticoagulant and anticardiolipin Ab,**
- **Negative C & P ANCA, and**
- **Negative Anti-SSA & SSB antibody,**
- **Normal protein C, S & antithrombin III**

Neuroimaging Characteristics (MRI Brain & Spinal cord, MRA of Brain & Neck vessels, and FFA of orbital vessels)

Trait	Frequency (%)
Micro hemorrhage	6 (35.2)
Hemorrhagic Infarct	4(23.6)
Ischemic infarct	4 (23.6)
Beaded appearance	7 (42)
Occluding thrombus in distal MCA	3 (17.6)
Dysplastic aneurysm	1(5.8)
Hypertintesnse lesion in brain & spinal cord	3 (17.6)
Optic Nerve head Vasculitis	2 (11.7)
Retinal vasculitis	1 (5.8)
Acute ischemic optic neuropathy	1 (5.8)

All patients have undergone MRV with contrast and were found normal

Retinal Imaging

Figure 1: Retinal photograph with FFA- optic nerve head vasculitis

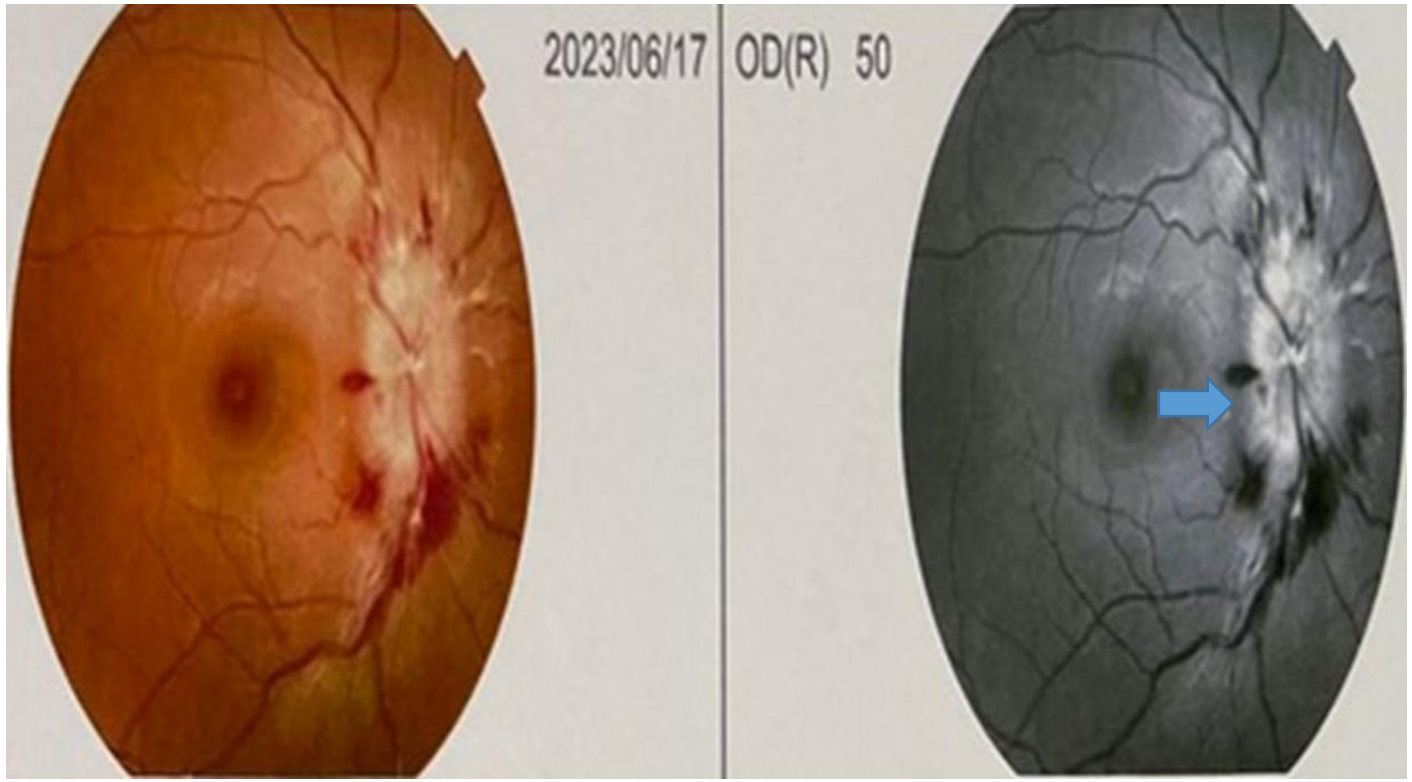
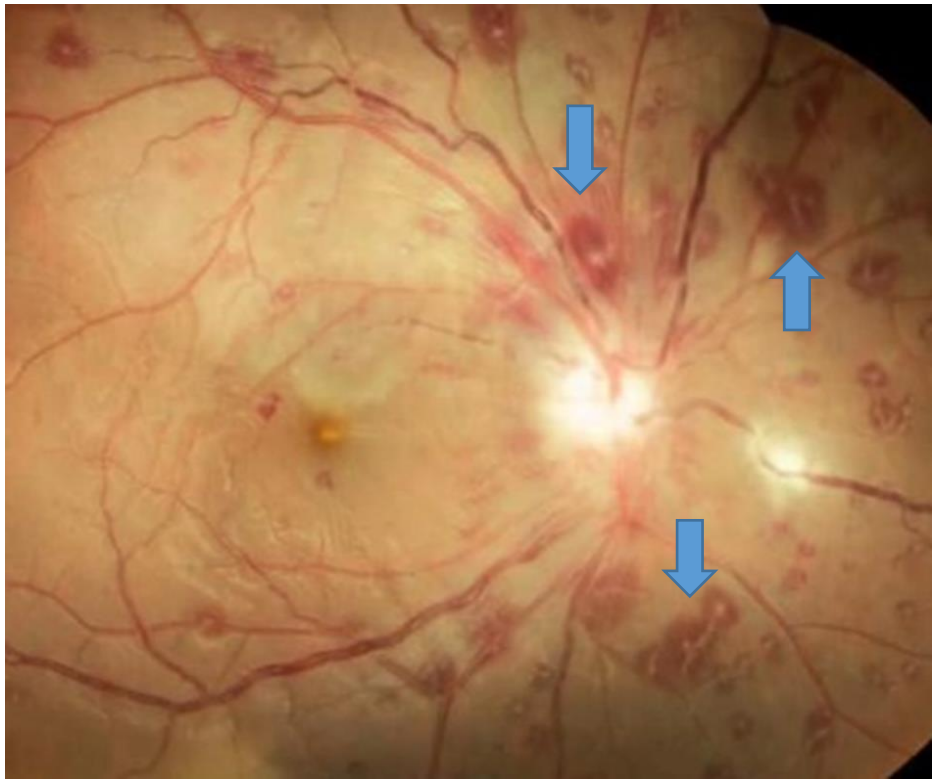


Figure 2: Retinal vasculitis



MRI Brain (GRE & DWI sequence)

Fig 1: Multiple areas of diffusion restriction (infarct) with blooming lesions (hemorrhage) in the GRE sequence

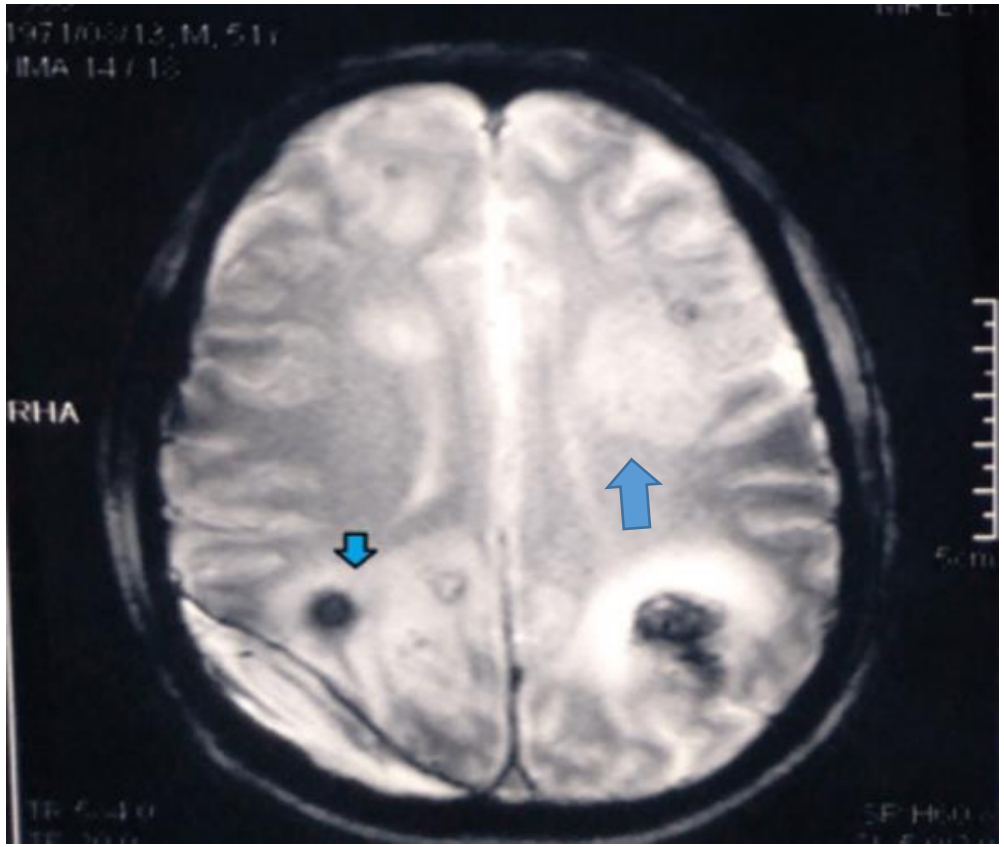
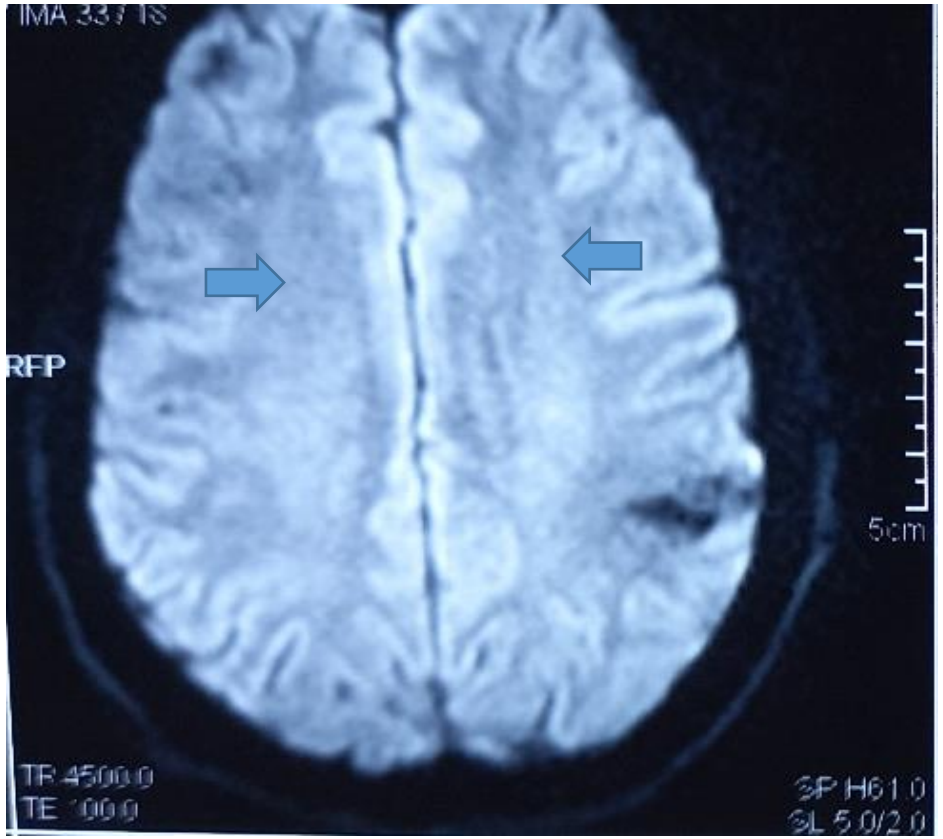
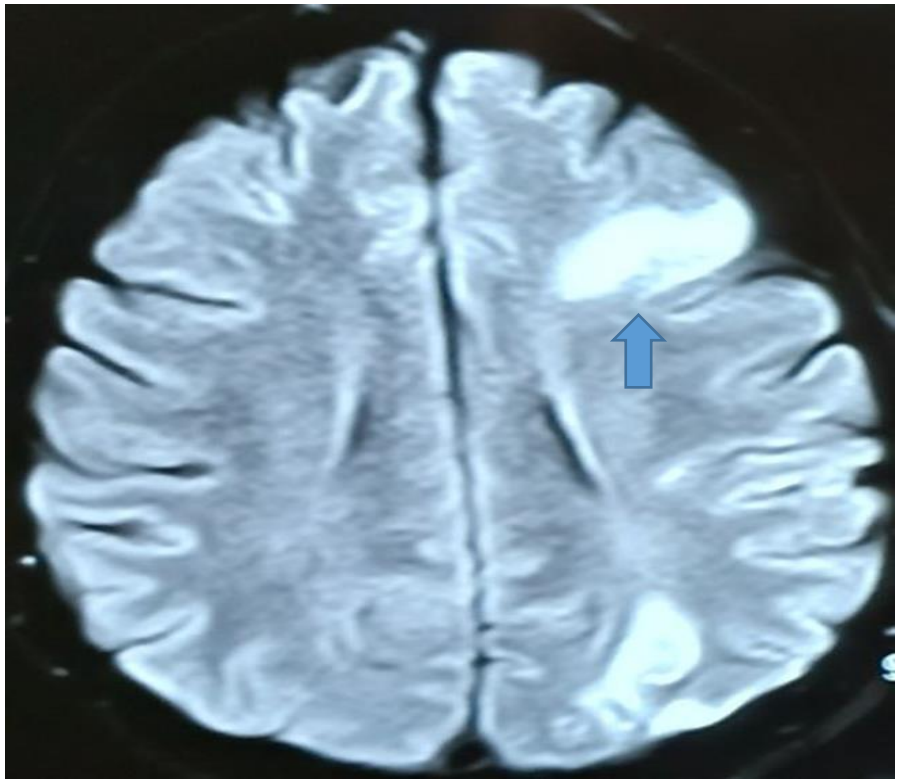


Fig 2: Multiple micro infarcts with small vessels ischemia in DWI sequence

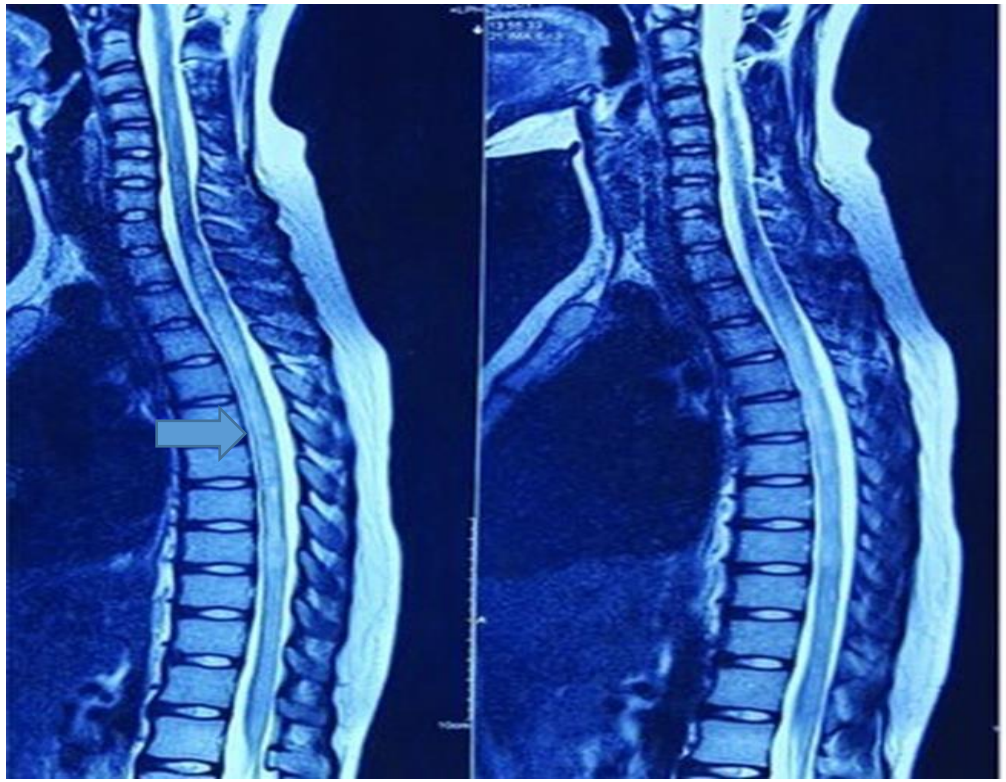


Hyper-intense lesions in MRI Brain & Spine

Cortical hyper-intense lesion in brain



Hyper-intense lesion in spine



MR Angiogram of Brain & Neck Vessels

Fig 1: Beaded appearance with luminal narrowing in the M1 branch of both MCA & vertebral artery

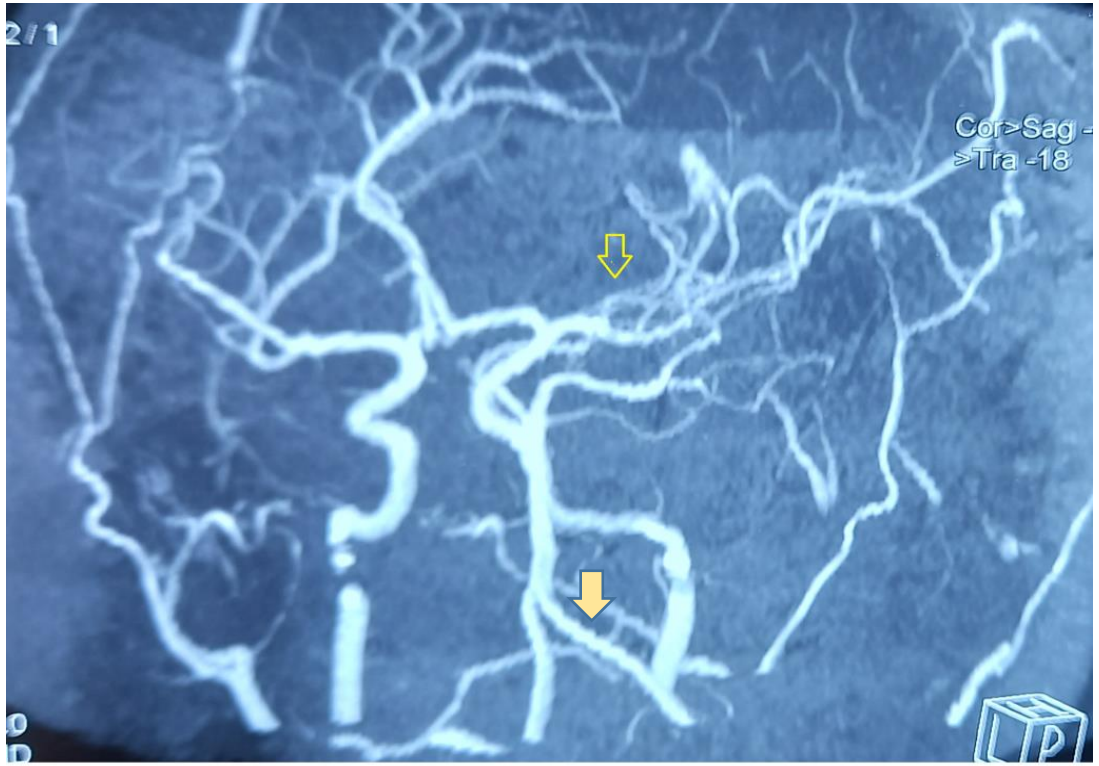
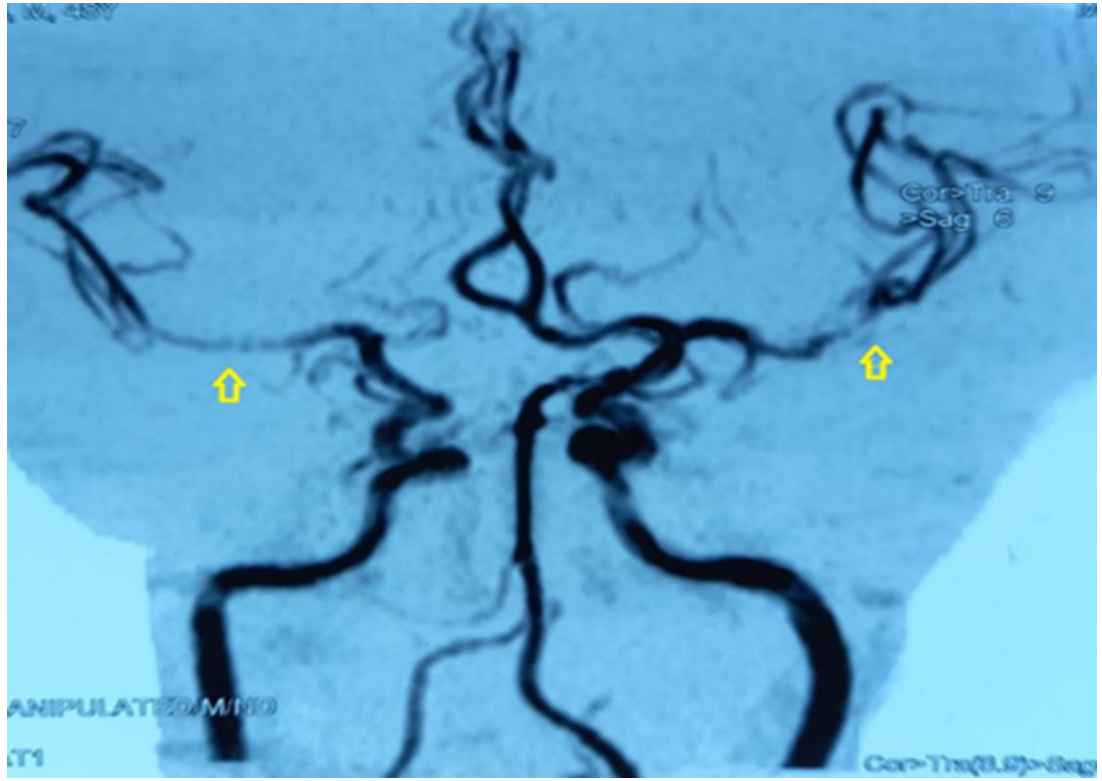
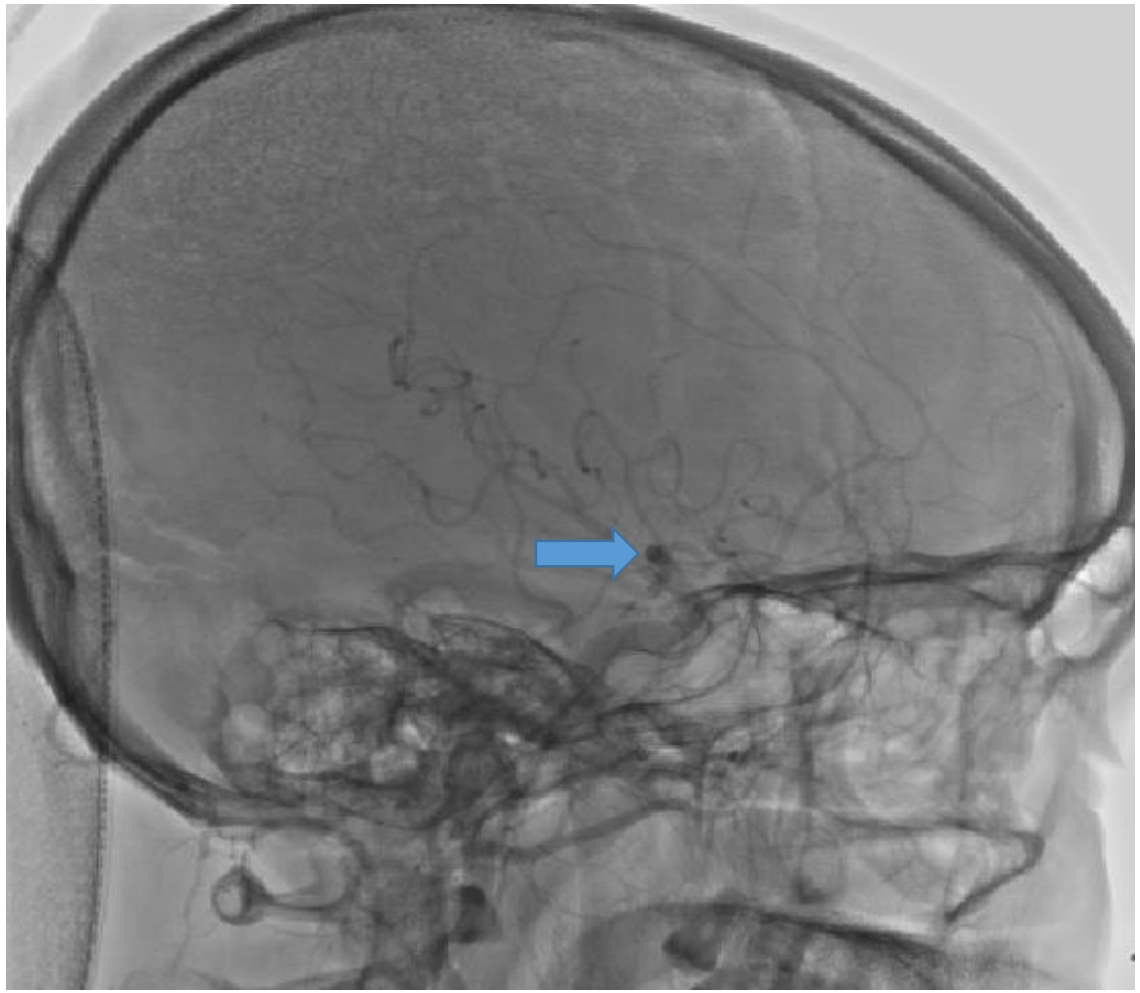


Fig 2: Luminal narrowing present in both M2 segment of MCA



DSA of Cerebral Vessels in Lupus Nephritis Patient

Fig: Dysplastic aneurysm in Pcom artery



Treatment Outcome

- Treatment was administered to all patients in the form of IV Methyl Prednisolone and IV Cyclophosphamide along with Hydroxychloroquine, followed by oral prednisolone and either Azathioprine or Mycophenolate mofetil.
- It was found that all of our patients achieved remission at 4 months with a SLEDAI of 2 [1-4.5]

120 Days Outcome

Trait	Result
SLE DAI on admission (Mean, SD)	24 (9)
SLE DAI at 120 days (Median, IQR)	2 [1-4.5]
Complete remission (frequency, %)	12 (70.5)
Partial Remission (frequency, %)	5 (29.4)
Herpes zoster	1 (5.8)
Upper respiratory tract infection	1 (5.8)

Conclusion

- Diagnosis of CNS Vasculitis is very challenging
- CNS vasculitis can be the first presentation of SLE
- Early diagnosis and initiation of immunosuppressive treatment is the key to good clinical outcome

Acknowledgement

- Dr. S K Jakaria been syeed
- Dr. Reaz Mahmud
- Dr. Uzzwal Mallick
- Dr. Sabrina Rahman
- **Prof. Md. Mujibur Rahman**
- Department of Radiology of NINS


THANK
YOU