A RARE CASE OF POLYARTERITIS NODOSA PRESENTING AS MONONEURITIS MULTIPLEX
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Abstract: Polyarteritis nodosa (PAN) is a systemic necrotizing vasculitis that typically affects medium-sized muscular arteries and occasionally affects small muscular arteries, resulting in secondary tissue ischemia. The kidneys, skin, joints, muscles, peripheral nerves, and GI tract are most commonly affected, but any organ can be involved. However, the lungs are usually spared. We present a case with neural and peripheral vascular, cardiac and renal manifestation. 

Keyword: mononeuritis multiplex, polyarteritis nodosa, vasculitis, periarteritis nodosa, pancreatitis nodosa

INTRODUCTION: Polyarteritis nodosa, also known as periarteritis nodosa, is a rare multisystem disease characterized by a necrotizing arteritis of small and medium-sized arteries. The most common sites affected are the skin, joints, kidneys, gastrointestinal tract, and peripheral nerves.

CASE REPORT: An 18 year old boy was admitted with complaints of diminished sensation over both feet and difficulty in walking and difficulty in breathing. 5 weeks back he developed a "numb, stabbing" persistent pain in his left foot. A week later, an identical pain developed in his right foot. One week before referral both his feet "went floppy." Few days before admission, this weakness progressed to the extent that he could not walk unaided. He also felt short of breath and had a dry cough. His past medical history was unremarkable except for hypertension which was diagnosed a month back.

EXAMINATION: Patient was tachypneic, illnourished with gross wasting of both upper and lower limbs. 

VITALS: SIGN: PR-115 beats/min BP-190/100 mmHg. RR- 28 breaths/min, oxygen saturation 89% on air, and temperature 36.8°C.

CVS - S1 S2+ pan systolic murmur heard at the apex.

RS - nvs+ crepts+

P/A- soft

CNS- HIGHER MENTAL FUNCTIONS- normal.

CRANIAL NERVES- 2ND CRANIAL NERVE- optic atrophy in right eye and hypertensive retinopathy changes in left eye. other cranial nerves normal

MOTOR SYSTEM: BULK - severe wasting of both upper and lower limb and involving trunk muscles.

INVESTIGATION:

• CBC: Hb - 9gms%, platelet count -1,70,000, WBC-11000
• RFT - urea -32mgs, creatinine -1.2mgs
• Blood sugar – 98mgs
• ELECTROLYTES - normal
• CRP - 62 mg/l
• ECHO - dilated left heart chambers.
• ANA - positive 1 in 10 dilutions – speckled pattern. ANTI DS DNA-NEGATIVE P ANCA -POSITIVE
• LFT- normal.
• VIRAL MARKERS - HbsAg, Anti HCV, and HIV are negative.
• USG ABDOMEN - increased cortical echos with normal sized kidneys.
• DOPPLER BOTH LIMB VESSELS - normal.
CT ANGIOGRAM shows no evidence of major vessel vasculitis including renal artery.

NERVE CONDUCTION STUDY - sensory motor axonal neuropathy.

RENAL DOPPLER STUDY - shows tardus parvus pattern in mid and lower pole suggested distal renal artery stenosis.

SURAL NERVE BIOPSY - diagnosed as medium vessel vasculitis - panarteritis nodosa.

DISCUSSION: Polyarteritis nodosa (also known as "Panarteritis nodosa and "Periarteritis nodosa") is a vasculitis of medium & small-sized arteries, which become swollen and damaged from attack by rogue immune cells. Polyarteritis nodosa was described in 1866 by Kussmaul and maier so it is also called as Kussmaul disease or Kussmaul-Maier disease. PAN doesn't affect pulmonary arteries although bronchial vessels may be involved. It is uncommon disease mainly affects fourth and fifth decades of life slightly male predominance. Non specific signs and symptoms are hallmarks of PAN. Fever wt loss and malaise are present most of the cases. Vague symptoms such as weakness, malaise, head ache, abdominal pain and myalgia that can be rapidly progresses to a fulminant illness. In renal involvement most commonly manifests as hypertension, renal insufficiency or haemorrhage due to microaneurysms. Pt had diagnosed by ACR criteria 3 out of 10 criteria:
- Weight loss of > 4 kg since beginning of illness
- Livedo reticularis
- Testicular pain or tenderness
- Myalgias, weakness, or leg tenderness
- Mononeuropathy or polyneuropathy
- Development of hypertension
- Elevated BUN or creatinine unrelated to dehydration or obstruction
- Presence of hepatitis B surface antigen or antibody in serum
- Arteriogram demonstrating aneurysms or occlusions of the visceral arteries
- Biopsy of small or medium-sized artery containing granulocytes

Our patient presented as mononeuritis multiplex in the course of hospital stay patient had multiple complications. nerve biopsy confirmed the diagnosis. ACCORDING TO ACR CRITERIA 6 OUT OF 10 CRITERIA IS FULFILLED. Patient was treated with steroids and cyclophosphamide. patient remarkably improved.

REFERENCE:
- Lightfoot et al 1990; Hasaniya and Katzen 1993
- HARRISON'S PRINCIPLE OF INTERNAL MEDICINE
- KELLY'S TEXTBOOK OF RHEUMATOLOGY