Case Records of the Severn Valley

CLINICAL NEUROSCIENCES

A Weakness That Hasn't Read the Textbook

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ABSTRACT

A 56-year-old lady was admitted for an elective assessment of dysphagia and a droopy neck to a secondary referral outpatients Gastroenterology Unit. During a pre-procedure assessment, a nurse noticed a slight dyscoordination and unilateral preference. On further enquiry, the patient reported a distal weakness in her left upper and lower limb, and a mild facial droop with no speech disturbance. An initial diagnosis of a Cerebrovascular Accident (CVA) was made and the patient was transferred to a tertiary referral centre.

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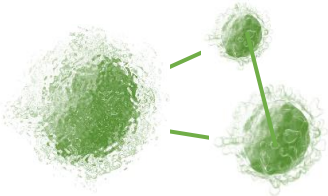

PRESENTATION

The patient reported a progressive weakness that was isolated to the proximal part of the distal muscles on both left leg and arm, that she noticed about seven months ago. She had progressive mobility problems as she could now barely walk a distance of 10 meters.

On top of her neurological problems, she suffered from a profound dysphagia, a feeling of stiff oesophagus, as she needed to take several swallows for every bite of her food.

She did not report any signs of meningism, malignancy, seizures, mylagias, or cramping, she was not in pain and her musculoskeletal history was unremarkable. There was no history of dyspnoea or signs of heath failure.

The lady saw her GP regularly for a treatment of hypertension, which was well controlled. She has no history of diabetes or hyperlipidaemia, and this is her first admission to the hospital. The remainder of the past medical and surgical history was unremarkable.

She recalls her dad having a similar problem, but since it was in the pre-NHS era, he did not get any diagnosis or treatment at that time, with a myocardial infarct as a cause of death.

The genogram representing the familial distribution of symptoms is presented in Fig.1

DRUGS: Ramipril 5mg *po od,* NKDA

She has no history of smoking, EtOH or drugs abuse. She lives alone with her dog and is well supported by five children who take turns in visiting her and helping her with the activities of daily living.

She heard that she may have had a stroke or a haemorrhage and is quite stressed about that, as she used to be a health visitor on a stroke ward in the community hospital, and she did not want to be transferred over there.

ON EXAMINATION

The patient appeared comfortable at rest: observations stable, apyrexial.

Cardiovascular system:
Blood Pressure: 135/90 mmHg
Pulse: 76 beats per minute, regular
Heart Sounds: *S1-VVVvvv-S2——–S1 + (/)*
[ejection systolic murmur, no added sounds]
Capillary Refill Time: <2s
Pulses: Present, normal (radial, brachial, carotid, abdominal, femoral, temporal, dorsalis paedis)

Respiratory:
Breath Sounds: vesicular, Air Entry: L=R, nothing of a note

Gastrointestinal:
Soft Non- Tender, Bowel Sounds + (present)

Neurological:
Cranial Nerve VIII – slight facial droop on the left side. Remainder of cranial nerves (N)

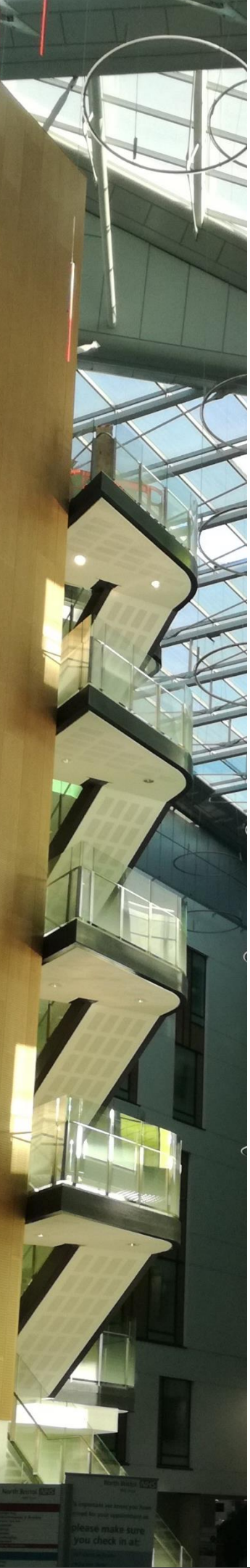

TABLE 1 Summary of the upper and lower limb neurological examination. *N=normal, L=left, R=right.*

UPPER LIMB		(L)	(R)
Inspection		Muscle wasting	N
Tone		UP	N
Power (MRC Oxford)	Shoulder elevation	5	5
	Shoulder depression	5	5
	Elbow flexion	2	5
	Elbow extension	4+	4+
	Wrist flexion	2	4+
	Wrist extension	4-	4+
	Finger flexion	4-	4+
	Finger extension	4-	4+
Sensation	C_4	N	N
	C_5	N	N
	C_6	N	N
	C_7	N	N
	C_8	N	N
	T_1	N	N

LOWER LIMB		(L)	(R)
Inspection		Muscle wasting	N
Tone		UP	N
Power (MRC Oxford)	Hip elevation	5	5
	Hip depression	5	5
	Knee flexion	4+	4+
	Knee extension	2	4+
	Ankle dorsiflexion	2	4+
	Ankle plantarflexion	4-	4+
	Big Toe flexion	4-	4+
	Big Toe extension	4-	4+
Sensation	L_1	N	N
	L_2		
	L_3		
	L_4		
	L_5		
	$S_{1/2}$		

Coordination, proprioception, and reflexes were (N)

TABLE 2 Blood workup and imaging investigations, along with the summary of the results.

Investigation	*Summary of Results*
Full blood count	Slightly elevated white cell count, otherwise normal
Magnesium, Calcium, Phosphate	(N)
ESR + PV	slightly elevated ESR, PV (N)
Creatinine, eGFR, Potassium Sodium	(N)
Clotting Panel	(N)
Liver Function Tests	(N)
Thyroid Function	(N)
Vitamin D levels	(N)
Rheumatoid factor	POSITIVE
ANA	NEGATIVE (normal)
CK level	MODERATE ELEVATION
Glucose	(N)
Paraneoplastic screen	(N)
Myositis-specific antibodies	NEGATIVE (normal)
Anti-ACh antibodies	NEGATIVE (normal)

INVESTIGATIONS

CT Head: Normal brain parenchyma, no intracranial bleeds, no focal lesions, CSF spaces within normal range; small global involutional change consistent with age.
CT myography: Selective atrophy of the forearm and knee flexors, otherwise unremarkable. The upper and lower limb examinations are summarised in Table 1.

The blood results are presented in Table 2.

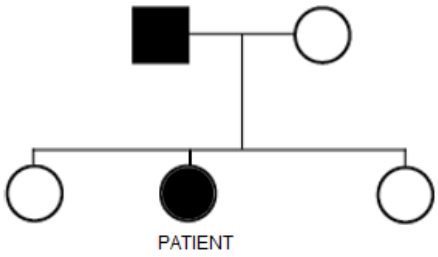

FIGURE 1. The genogram of familial distribution of the symptoms, individuals affected are marked in black. The patient from the case is labelled underneath. Square represent males and circles represent females.

NB: The hint is in the graphic on the first page...

QUESTIONS

1. What are the differential diagnoses?
2. What further tests would you consider?
3. What is the proposed treatment?

ANSWERS

Let us know your answers via email on *clinneusoci@neurologicalsociety.org*

The corresponding author will be replying to you individually; the answer will be revealed in the next issue of the *Clin Neurosci*

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