Original Article

The patterns of clinical presentations of cerebellar syndromes among adult Sudanese patients

Abbashar Hussein, Adam Mohamed, Ali Younis, H.A.Eltoum, A.Sidig, K. Khalf alla, O Al adil, A Hammad, M. Malk, Ammar El Tahir, MOH Gadour.

ABSTRACT

Cerebellar syndromes are one of the commonest neurological diseases.

Objectives: To study the patterns of clinical presentations of cerebellar syndromes and to identify the possible causes.

Methods: This is a prospective hospital based, cross-sectional study. One hundred adult Sudanese patients with cerebellar syndromes were included in the study during the period from January 2006 – January 2007.

Results: The most common age group affected was 18 – 25 years. Male to female ratio was 1.5: 1 unsteadiness on walking was the most common symptom (83%).

Gait-ataxia was the most common sign (83%). Cerebrovascular disease was the most common aetiology (25%).

Conclusion:

Cerebellar syndromes are not rare in Sudan. However, they were diagnosed more commonly at the central regions of the country probably because of more awareness of patients and better facilities for diagnosis.

The age of onset, the male predominance, the presentation and clinical findings were not different from reported literature. This also goes for the common causes apart from alcohol which is a strikingly rare as a cause in this study and could be accounted for the implementation of Elshariya (Islamic laws) Laws in Sudan.

Keywords: ataxia, dysmetria, disdiadochokenesis, decomposition, nystagmus, dysarthria.

Cerebellar syndromes are clinical terms used to designate manifestations of damage of the cerebellum regardless of origin[1,2]. There are many disorders that affect the cerebellum leading to the cerebellar dysfunction and cerebellar syndromes. Cerebellar syndromes are not uncommon in clinical practice, although there are no available data about prevalence in Sudan. They affect both sexes and occur at any age. Cerebellar disorders present with a variety of clinical features include, ataxia, dysmetria, disdiadochokenesis, decomposition of movement, hypotonia, nystagmus, dysarthria or tremor.

Corresponding Author: Dr Abbashar Husien. Department of Medicine, Facualty of Medicine, University of Khartoum, Sudan. E. mail: abbashar99@yahoo.com

Some patients may have difficulties in walking with repeated falling, sphincteric disturbances, disturbance of sight or hearing, or rarely cognitive dysfunction. Although there are many cerebellar syndromes, two distinguishable cerebellar syndromes (midline and hemispheric syndromes) are common[3]. The midline cerebellar syndromes are characterized by imbalance, while hemispheric syndromes are characterized by incoordination. Cerebellar syndromes have a variety of causes with variable onset and courses, and some causes are aggressive and can lead to disability and even death. Generally cerebellar disorders make a great burden on the family and the heath care system especially when the presentation is early.Saddly, the impression is that the patients are presenting late when the diagnosis and treatment become more demanding, difficult, expensive and less rewarding. The

diagnosis of cerebellar syndromes is mainly clinical, based on detailed history, physical and neurological examination. Investigations such as neuro-imaging, electrophysiological study, CSF and blood test for specific conditions are useful in supporting the diagnosis, and detection of the causative agents, also other mimicking conditions can be excluded[4, 5].

Objectives: The objectives of this study were to study the patterns of clinical presentation of cerebellar syndromes among adults Sudanese patients in Al-Shaab Teaching Hospital, from January 2006 to January 2007and to identify the possible causes of cerebellar syndromes.

Methods

This is a descriptive prospective, hospital based, cross sectional study. It was conducted at Al Shaab Teaching Hospital which is a specialized tertiary hospital, located in the centre of Khartoum town. There are two neurological units with 43 beds and two neurosurgical units with 50 beds, there are three intensive care units, two neurology referred clinic and three neurosurgery referred clinics each week. The study population included was 100 patients with cerebellar syndromes, referred or admitted to Al Shaab Hospital in the period from January 2006 to January 2007. All the patients were newly diagnosed and they were not on treatment. All the patients were adult Sudanese; those below 18 years of age were excluded. All patients gave their verbal consent to participate in the study. Ethical approval was received from the local ethics committee prior to the start of the study .The data were collected by the authors using structural questionnaire. The informers were either the patients or close relatives. A full details history proper systemic and neurologic examination was performed to each patient by the authors. The physical signs were grouped to cerebellar signs, non cerebellar neurological signs and systemic signs. Investigations done for each patient include CBC and ESR, RBS, CT scan or MRI of the brain, while the investigations done if needed included BFFM, LFTs, TSH, T4,VDRL, HIV screen, viral Serology, LP and CSF examination, serum vitamin (B12 or E) levels ,serum copper and ceruloplasmin, CXR, ECG, ECHO or electrophysiological study. The data were analyzed and the results were expressed into figures, tables or graphs using statistical package for social science (SPSS) and discussions, conclusions and recommendations were done.

RESULTS: Sixty patients (60%) were males. Age distribution ranged between 18–73 years with mean age of 38.18 years (table 1).

Table 1: Age distribution

Age group	No.	Male	Female
18-25	28	16	12
26-33	16	10	6
34-41	15	10	5
42-49	13	6	7
50-57	5	3	2
58-65	14	10	4
66-73	9	5	4
Total	100	60	40

Twelve patients (12%) had onset of symptoms before 18 years of age. Forty patients (40%) were from Khartoum State and 19 (19%) from Gazera State (table 2).

Table 2: Areas of origin of patients

Areas of origin	No.	Male	Female
Khartoum State	40	28	12
Gazera State	19	13	6
Northern region	14	6	8
Eastern region	12	6	6
Western region	11	5	6
Southern region	4	2	2
Total	100		

Twenty patients (20%) had hypertension. Diabetes and epilepsy were found in 15 patients each while jaundice and cough were found equally in seven patients. Only 2 patients (2%) had visual loss. Eight patients (8%) had family history of cerebellar disease. Thirty patients (30%) tend to smoke cigarettes, 20 (20%) used to consume alcohol.

Eighty three patients (83%) had unsteadiness on walking, 79 patients (79%) had tremor on movement, 74 (74%) had unsteadiness on standing, 12 (12%) had unsteadiness on sitting and 71 (71%) had disturbance of speech. The onset of symptoms was acute in 20, sub-acute in 25 and gradual in 55 patients. Only seven patients (7%) had episodic course. The majority (55%) of patients had progressive course, 27% had static course and 11% had static course. Ataxic gait occurred in the vast majority (83%) of patients. intention tremor past-pointing (dysmetria) and dysdiadochokinesia were found in 81 (81%) each. Dysarthria, nystagmus, pendular reflexes and hypotonia were detected in 71 53, 31 and 29 patients respectively. MRI and CT scan of the brain revealed that cerebellar demyelination and plaque formation were the commonest radiological findings where as cerebellar abscess is the least one to be seen (table 3).

Table 3: Brain MRI or CT-scan findings

MRI or CT-scan findings	N0	M	F
Cerebellar infarction	13	10	4
Cerebellar hemorrhage	11	7	4
Cerebellar demylinetions and plaque formation	19	3	16
Primary cerebellar tumor	15	11	4
Secondary cerebellar tumor	2	1	1
Viral encephalitis	7	5	2
Cerebellar abscess	1	1	zero
Cerebellar degeneration + cerebellar atrophy	15	12	3
No abnormal findings	17	10	6
Total	100		

Macrocytic anaemia was detected in 15 patients (15%), and falciparum malaria was positive in 7 (7%) patients (table 4). Cerebrovascular diseases contributed to one fourth of the patients whereas alcoholic cerebellar disease and Kearn's Sayre syndrome form the least aetiological causes of cerebellar syndromes (table 5).

Table 4: Blood test findings

Blood test findings	No.	Male	Female
Macrocytic anaemia	15	10	5
High ESR	25	15	10
Positive blood film for malaria	7	5	2
Low T_4 and high TSH	2	0	2
Abnormal LFT	10	6	4
Low ceruloplasmin	1	1	zero
Low serum copper	2	2	zero
High blood sugar	15	10	5
HIV positive	2	1	1
HSV positive	2	1	1
No abnormalities	19	9	10
Total	100		

Table 5: Aetiology of cerebellar syndromes

Blood test findings	No.	Male	Female
Macrocytic anaemia	15	10	5
High ESR	25	15	10
Positive blood film for malaria	7	5	2
Low T_4 and high TSH	2	0	2
Abnormal LFT	10	6	4
Low ceruloplasmin	1	1	zero
Low serum copper	2	2	zero
High blood sugar	15	10	5
HIV positive	2	1	1
HSV positive	2	1	1
No abnormalities	19	9	10
Total	100		

Discussion

Cerebellar syndromes are clinical neurological disorders, characterized by imbalance, incoordination and disturbance of involuntary eye movements. It may be due to disorders affecting the cerebellum, its main connections or both. The most common causes include cerebrovascular, demyelinating disorders, drugs, toxins and alcohol. Rare causes include tumours, infections, degenerative hereditary disorders, idiopathic cerebellar atrophy and metabolic disorders[6-8].

In our study, 28 patients (28%) lie in the age group (18–25 years), this high percentage of cerebellar syndrome among this age group can be explained by the fact that these are the

active sector in the community, so they are more exposed to risk factor like trauma, than other age groups, also the high presence of diseases such hereditary cerebellar ataxia, metabolic, infections and demyelinating disorders among these age group might have contributed to that. This high percentage is similar to literature[7, 8]. Male predominance (1.5:1) may be because of the same reasons besides being more smokers and alcohol consumers. The better education, finical status, awareness of population and availability of diagnostic tools and health facilities at the central parts of Sudan - Khartoum and Gazera State- have probably played a role detection of the disease and hence the high percentage of cerebellar syndrome found in these areas when compared to East, West and North Sudan. Unsteadiness on walking, tremor on movements, disturbance of speech were the most common presenting features in this study similar to what was described by Holmes and others[7-13]. Also the clinical cerebellar findings were in concordance with the literature[9-12].

The brain MRI or CT scan had a big role in detecting most causes of the cerebellar syndrome (like focal lesions) in our study; this was similar to what was mentioned in the literature[6-8]. Diabetes mellitus, hypertension, epilepsy, multiple sclerosis were the commonest predisposing diseases associated with cerebellar syndromes, while chronic liver diseases, tuberculosis and lung cancer were the less common predisposing factors, this is in agreement with other researchers[14,15]. Cerebellar disorders were less commonly seen in the families in our study population, this indicated that the hereditary disorders were rare causes; this goes with reports[16,17].Alcohol consumption and smoking as risk factors were seen in a considerable number of our study population, this was found to be similar to what was mentioned in other studies world wide[13]. Cerebellar vascular diseases were the commonest cause of cerebellar syndrome and this is consistent with the literature[18,19]. Demyelinating lesions especially multiple sclerosis is the second common cause of cerebellar syndrome in our study population, presenting with typical cerebellar features, which is similar to what was reported by other researchers[6-8,20]. However, this is different from Bruce's findings[7,21]. Unlike reports from literature but similar to what was mentioned in America, tumours of the posterior cranial fossa affecting the cerebellum or it's connections is a 3rd common cause of cerebellar syndrome in our study population[7,8,12,20]. Infections such as viral encephalitis (cerebellitis) were found to be the 4th common cause of cerebellar syndrome in our study population, this differed from what was mentioned in the literature[7,8,20], but similar to a recent American [22-25], India and Sir-Lanka studies[15-17]. The high incidence of infections in our study population may be due to poor hygiene, poverty and illiteracy or may be due to endemicity of some infections like malaria and lack of prophylaxis such as vaccines. Drug toxicity (antiepileptic drugs) is the 5th common cause of cerebellar syndrome in our study population, this high percentage is similar to what was mentioned in the literature[6-8,20,26]. Hereditary cerebellar ataxia and idiopathic cerebellar atrophy and metabolic disorders like hypothyroidism and Wilson's disease are least common causes of cerebellar syndromes, these are similar to what was mentioned in the literature[27-31]. Mitochondrial disorders like Kearns's Sayre syndrome is a very rare cause of cerebellar syndrome among our study population[32,33]. Alcoholic cerebellar disease was found to be a strikingly rare cause of cerebellar syndrome in our study population, which is different from reports from other parts of the world[7,8,20,34,35]. The presence of low alcoholic cerebellar disease in our study population can be explained by decreased alcohol consumption in our community during the last twenty years due to implementation of Elshariya (Islamic laws) Laws.

Conclusion

Cerebellar syndromes are not rare in Sudan. However, they were diagnosed more commonly at the central regions of the country probably because of more awareness of patients and better facilities for diagnosis.

The age of onset, the male predominance, the presentation and clinical findings were not different from reported literature. This also goes for the common causes apart from alcohol which is a strikingly rare as a cause in this study and could be accounted for the implementation of Elshariya (Islamic laws) Laws in Sudan.

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