Global Advanced Research Journal of Medicine and Medical Science (ISSN: 2315-5159) Vol. 4(7) pp. 347-350, August, 2015
Available online http://garj.org/garjmms

Full Length Research Paper

Neuromyositis: Clinical and Electrophysiological study of 8 cases

Halil Onder*, F.GökcemYıldız and Çagrı Mesut Temucin

Hacettepe University Hospital, Department of Neurology,Sıhhıye, Ankara, Turkey

Accepted 06 August, 2015

Concomitant involvement of peripheral nerve system in dermatomyositis and polymyositis defined as 'Neuromyositis' is a very rarely reported entity. Clinical presentations and pathogenesis of this entity is not well known. In addition, definite diagnostic criterias as well as even occurrence of this entity is still an argument subject for some authors. Here, via our 8 cases diagnosed both with dermatomyositis/polymyositis and neuropathic involvement, the existence of 'Neuromyositis' will be interrogated. We scanned the electrophysiological (EP) reports of dermatomyositis or polimyositis patients studied between 2000-2014 also for the key word 'neuropathy'. Clinical evaluation of these patients were performed retrospectively using hospital computer data system and file notes. EP reports of 8 patients with myopathy showed findings of neuropathic involvement (5 male). Two patients were diagnosed with dermatomyositis and 6 were diagnosed with polimyositis. All EP reports revealed axonal neuropathic involvement. We determined the coincidental presence of diseases like diabetes mellitus, systemic lupus erithematosus, collagen tissue diseases or history of drug usages (cycplatin, colchicum, etoposide) that could be responsible for neuropathy in 7 of them. Except one patient diagnosed with dermatomyositis and diabetes mellitus, not any of them had neuropathy related symptoms (sensorial deficit, distal paresis). For our opinion, in cooccurence of neuropathy and myopathy seconder neuropathy causes must be excluded. In EP studies of myopathic patients, axonal neuropathy findings can suggest the entity of 'Neuromyositis'. Nevertheless, to investigate the 'clear picture' of neuromyositislarger case studies involving concomitant nerve and muscle biopsies must be done.

Keywords: Neuromyositis, Electrophysiological, dermatomyositis, polymyositis

INTRODUCTION

The term 'Neuromyositis' indicating concomitant occurrence of polymyositis (PM) or dermatomyositis (DM) with neuropathy was first introduced by Senator in 1893. Although its long usage, its meaning has oftenly been interrogated. In other respects, polyneuropathy and myositis can occur concomitantly with the association of lupus, arteritis, visceral malignencies or collagen tissue diseases as well as neuropathy can occur as a consequence of an coincidental metabolic diseases (diabetes mellitus, vitamin deficiency, alcoholism) (Matsui et al. 2003). Also, possibility of muscular changes caused by neurological involvement and, on the other hand, neurogenic type manifestations caused by PM constitute another argument about definition and diagnosis of this entity. A French study group defined it as a clinic

*Corresponding Author Email: halilnder@yahoo.com; Telephone: +3123051807

Table 1. Demographicandclinicaldata of patients

Patient Number	Gender	Age	Diagnosis	Neuropathytype	Co-morbidities	Pre-EMG medicine	Prox. Str
1	F	69	PM	axonal	Sjogren S., gastrit, HT	steroid (iv, po), cyclofosfamid	mildparesis
2	M	51	DM	axonal	Sclc		nodata
3	F	52	PM	axonal	DM, Hepatosteatoz	steroid (iv, po)	mildparesis
4	F	70	PM	axonal	HT, OP, CAD	steroid (iv, po), cyclofosfamid	mildparesis
5	M	47	PM	axonal	Scleroderma	Scleroderma	mildparesis
6	F	65	PM	axonal	FMF, OP	colchicum	moderateparesis
7	F	65	PM	axonal	DM	steroid (po), IVIG, plz.	normal
8	F	80	DM	axonal	BreastCa.	steroid (po)	nodata

DM: dermatomyositis, PM: polimyositis, İv: intravenous, po: peri oral, HT: hyperension, S: syndrome, dm: diabetesmellitus, OP: osteoporosis, CAD: coronaryarterydisease, FMF: familialmediterraneanfever

opathological reality based on definite criteria of both primer muscle and nerve disease. They also made it fundamental to exclude a seconder cause (diabetes, alcoholism) after diagnosis of neuropathy (Laraki et al., 1994).

Signs of peripheral neruopathy can be found at clinical, electrophysiological (EP) and pathological examinations. Axonapathy is usually reported to be as the neuropathy type, however the existence of polyradiculoneuritis rarely has been shown to occur. On the contrary; in an EP study, slowed motor and sensory nerve conduction velocities a marker of demyelinization were suggested as the most important and common findings, nonetheless it was stated that there is not a consensus on EP findings of neuromyositis, and electrophysiological differentiation of PM and neuromyositis may consume a very difficult deal (Laraki et al., 1993).

Here, we investigated 8 patients at whom concomitant myopathy and neuropathy were determined. Their demographic features, co-morbidities, medication history, and neurological examinations were separately presented. Also EP findings will be discussed.

MATERIALS AND METHODS

Reports of EP studies performed between 2000-2014 were scanned using hospital computer system (HCS).Reports consisting the terms ‘neuropathy’ in the findings tab of the report and additionally ‘dermatomyosit, polymyosit’ in the patient’s clinical tab were determined. Concomitant neuropathic involvement in DM or PM patients were defined at 8 patients’ EP study reports. All the patients were diagnosed with DM or PM according to clinical and laboratory examinations. Patients’ demographic features, co-morbidities, drug histories and laboratory findings were obtain edvia HCS patients’ files scanning's.

RESULTS

Of eight patients, five were female. Their age range were from 47 to 80. Age average was 62. Six patients were diagnosed with PM and two were diagnosed with DM. Their median age was 62 (range: from 47 to 80). At all of the patients, EP studies showed axonal neuropathic involvement. At the same time, 6 of 8 recordings showed findings compatible with active myopathy in addition to neuropathy. Blood creatinine kinase (CK) values measured nearest time before EP study varied from 139 to 915 ng/ml. Six of them had taken medical treatments such as steroid, intravenous immunglobulin due to myositis before EP study. Seven patients had comorbidities which could cause neuropathies. Two of them were malignencies (breast cancer, lung cancer), 3 of them were rheumatic diseases (Sjogren's syndrome, scleroderma, Familial Mediterranean fever) while 2 of them were diabetes mellitus. Tree patients had also drug usage history consisting of colchicum, cisplatin and etoposide.

Muscle biopsies were conducted on 5 patients. Three of them showed inflamatuarmyopathic changes, one of them mild differences in size between muscle fibers and other biopsy had showed normal muscle tissue. As a result, except one patient; all patients had a history of additional disease or drug usage that could be attributable as a cause of concurrent neuropathic involvement. At patient 8, a 80 year old female with a history of breast cancer, there was not any

Table 2. NerveConductionStudy

	Right Median				Right Ulnar				Right Peroneal		Right Tibial		Right Sural	
	Motor		Sensory		Motor		Sensory		Motor		Motor		Sensory	
Patient Number	**CV**	**A**	**CV**	**A**	**CV**	**A**	**CV**	**A**	**CV**	**A**	**CV**	**A**	**CV**	**A**
1	47	6.4	45	7.2	43	6.4	51	7.9	41	2	41	2.8	41	2.7
2	52	9.2	45	4.5	47	6.3	44	2.2	39	0.9	36	5.4	39	1
3	58	6.8	41	3.4	55	6.6	56	2.2	47	2.4	43	6.5	49	4.1
4	54	6.7	48	9	54	6.9	46	4.8	38	1.9	36	1.9	42	3.9
5	51	1.8	47	3	51	2.1	0	0	43	1.9	42	6.1	42	6.8
6	53	8.2	52	14	54	2.5	0	0	46	2.1	42	0.4	0	0
7	55	5.2	52	21	53	9	50	16	49	3.8	45	12.3	32	7.8
8	50	4.1	57	4.6	57	4.4	53	3.8	46	2.9	47	11.3	45	8.2

CV: conductionvelocity, A: amplitude

additional cause found for neuropathy process.

In all the patients, at least moderate response were achieved with only corticosteroid or additionally immunosuppressant therapies during clinical follow up.

DISCUSSION

The present study demonstrates clinical and EP findings of 8 patients which are diagnosed with concomitant neuropathy and myopathy existence.

On clinical examination of myopathy patients, polyneuropathy findings can usually be missed because of the unclear findings and symptoms (Laraki et al., 1993). Hence, polyneuropathy at these patients are usually reported to reveal in postmortem examination. In these reports, pathological features demonstrate heterogeneous features including both axonal degeneration and demyelination (Thomas et al., 1940; McEntee and Mancall, 1965; Barren and Fine, 1959). Except one patient neither of our patients complained any sensorial symptom in accord with the past studies. Only patient 4 had stocking-glove sensory defisit. However, in another EP study in a wide group of DM/PM patients 17 (50%) of 34 patients whose were evaluated having peripheral nerve involvement on NCS, were reported to have clinical signs and symptoms of peripheral neuropathy (Wang et al., 2010).

EP study showed axonal neuropathy in all our patients. This data is compatible with findings of Wang et al. In their study with patients exhibiting comcomitant myopathy and neuropathy, neuropathic damage were reported to be predominantly distal axonal, most severe in lower limb nerves and sensory nerves (Wang et al., 2010). In contrast, nerve pathology data at DM patients from literature gives conflicting results. Some specimens remarked inflammatory cells around nerve fibers as well as small vessels while others did not affirm this. Nonetheless, endothelial vascular injury in peripheral nerve tissues is usually a demonstrated finding (Vogelgesang et al., 1995). In another study, vascular endothelial growth factor (VEGF) level in nerve tissue was evaluated to be high. As a result, they suggested a vasculitic process induced by overproduction of VEGF for DM patients complicated with peripheral neuropathy. Still, the pathogenesis behind neuropathy is unclear (Matsui et al. 2003). In our series, nerve biopsy was not conducted from any patient. Muscle biopsies were conducted from 5 of 8 patients. Two of them showed lenfositic infiltration, one showed myositic changes while the other patient's biopsy showed only mild diameter differences of muscle fibers and patient 5's specimen was evaluated as in normal ranges (which was attributed to technical failure of biopsy).

An important argument about this entity is that whether the neuropathy is due to nerve compression or to malnutrition from underlying chronic disease. Another aspect is possibility of another seconder cause for neuropathy (SLE, polyartheritis, metabolic disease) can be misdiagnosed (Wang et al., 2010). A study in

1994, which demonstrated 4 patients of neuromyoositis followed in an Internal Medicine department and the review of the literature suggested the diagnosis of neuromyositis, which can be expressed as a clinico-pathological reality, is based on the association of definite criteria of both primary muscle and nerve involvement excluding muscular abnormalities that could be the consequence of nerve involvement and vice versa. As a result when they searched literature according their restricted criteria for neuromyostis they could find only 6 cases corresponding to this definition. In 2 of this 6 cases, pathological specimens showed vasculitic samples that is much higher than observed in PM (Laraki et al., 1994). In our study, except one patient, all patients had at least one possible seconder causes which can be responsible from neuropathy. In only one patient, a 80 year old female, diagnosed with DM and breast cancer a seconder cause could not be found. Nevertheless, because of the retrospective study design a possibility of paraneoplasticneuropathy as a responsible factor for neuropathy could not be excluded. Another issue is that; the blood creatine kinase (CK) levels of the patients, investigated nearest time to the EP study, were not in very high levels which suggests at the time of EP study, the myositis were not in very active phase. Additionally, data of neurological examinations available from 6 patients revealed normal motor strength in one patient, mild proximal muscle weakness in 4 patients and moderate proximal weakness in only one patient demonstrating a well-controlled group of myositis. Concurrently, all the patients gave moderate to good response to only corticosteroid or immunosuppressant agents additionally.

Despite small number of patients in our report, we suggest that there is no correlation between the severity of myositis and neuropathy occurrence which may support our hypothesis (neuropathy is seconder to other coexistences) and decrease the possibility of myositis as a cause of underlying neuropathic involvement. Still, we think that these hypothesis have to be proven in further prospective case series.

CONCLUSION

As a result; in circumstances of these rare association, we suggest to exclude a seconder cause of neuropathy as a first-step investigation. Axonal neuropathy were determined in all our patients. We think that in nerve conduction studies motor and sensory nerve amplitude and fibrilations and giant MÜPs in needle electromyography can be an important sign for neuropathic involvement. However, our 7 cases do not correspond with 'clear picture' of neuromyositis mentioned by Vogelgesang et al. 1995 (Vogelgesang et al., 1995). Nonetheless, whether axonal neuropathy is a presentation of neuromyositis or a seconder cause still constitute an issue of debate. Further studies of larger case series including pathological specimens (both muscle and nerve biopsies) should be done to clarify to this argument.

REFERENCES

Barren KD, Fine DIM (1959). Neuromyositis. J. Nerv. Ment. Dis. 128: 497- 507.

Laraki R, Blétry O, Agbalika F, Bouche P, Godeau P (1994). (Do neuromyosites exist?). Ann Med Interne (Paris).145(2):88-97

Laraki R, Bletry O, Raguin G, et al (1993). 4 new cases of neuromyositis, one of them associated with HTLV-I infection. Rev. Neurol. (Paris). 149(4):283-288.

Matsui N, Mitui T, Endo I, et al (2003). Dermatomyositis with peripheral nervous system involvement: activation of vascular endothelial growth factor (VEGF) and VEGF receptor (VEGFR). Invasculitic lesions. Int. Med. 42:1233–1239

McEntee WJ, Mancall EL (1965). Neuromyositis: A reappraisal. Neurology 15: 69-75.

Senator H (1893). Uber acute Polymyositis und Neuromyositis. Deutsche MedWchnschr 19: 933-936,

Thomas D, Kinney MD, Mary M, Maher MD (1940). Dermatomyositis.A study of five cases. Am. J. Pathol. 16: 561-594,.

Vogelgesang SA, Gutierrez J, Klipple GL, Katona IM (1995). Polyneuropathy in juvenile dermatomyositis. J Rheumatol 22: 1369-1372,

WANG Yue, CUI Li-ying, CHEN Lin, LIU Ming-sheng, QI Xin, LI Ben-hong, DU Hua (2010). Nerve conduction studies in patients with dermatomyositis orpolymyositis. Chinese Med. J. 123(5):523-526