PECULIARITIES OF PRIMARY-PROGRESSIVE MULTIPLE SCLEROSIS IN THE RESIDENTS OF VOLYN REGION

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Abstract

The course of primary progressive multiple sclerosis (PPMS) among the residents of Volyn region was studied. It was established that the proportion of patients with PPRS is 7.95%. The typical characteristics of the cohort of the patients are polysymptomatic debut, in early thirties, with primarily involving the cerebellum and pyramidal systems, with a significant degree of disability and rapid progression of the disease. The data can be used in the planning of health and social care on the patients with PPMS.

Keywords: Primary-Progressive Multiple Sclerosis, course of disease.

Introduction. Multiple Sclerosis (MS) is an actual medical problem in Ukraine, as it holds the second position in the disabilities among diseases of the nervous system. Thus, according to the Health Statistics of the Ministry of Health of Ukraine, there were registered 20674 adult patients with MS in 2013. The prevalence rate of MS in Ukraine is 55.3 per 100,000 inhabitants [1, 4].

Each year the number of patients on the MS increases by an average of 1000-1200 people. In 2013, MS was firstly diagnosed in 1196 inhabitants of Ukraine. The incidence of MS in 2013 was 3.2 per 100.000 population. Volyn region is located in the north-western part of the country and has one of the highest incidence and prevalence rates in Ukraine [1].

Primary-progressive MS (PPMS) is a relatively rare form of MS, which occurs in approximately 10% of patients with MS. The absence of relapses makes certain difficulties in the diagnosis. Preliminary studies indicate the rapid increase in disability after onset of symptoms. None of the treatments showed significant slowing the progression of symptoms in patients with PPMS, including drugs approved for use in relapsing-remitting MS [4].

Extending possibilities in the treatment of MS, implementation in routine practice new generation drugs require evaluation of MS course and possibility of progression of the early stages

of the disease. Therefore, the study of typical clinical features of the disease is extremely important.

The purpose of the investigation was to study the peculiarities of PPMS on the basis of the MS registry in Volyn region.

Methods. During 2012-2013 a complete clinical examination of patients with MS and other demyelinating diseases was performed at first. We included into the registry MS patients who correspond to criteria №1 McDonald 2010 with Polman amendment [5]. In determining primary progressive course of the disease it was used definition of F.Lublin [3]. We used Functional System Scales and Expanded Disability Status Scale to assess the disability. The index of progression was calculated by the index ratio FS to disease duration in years [7]. Statistical analysis was performed using Microsoft Excel and Statistica 17.0 using standard descriptive statistics. We used the multiple surveillance Kaplan-Mayer curves to analyze the probability of achieving the events. Comparison of survival functions in the two groups was conducted using Cox F-test. Results were considered statistically significant at $p < 0.05$.

Results. Of the 629 patients registered in the Volyn regional registry 302 patients were examined from all districts. Primary-progressive course was identified in 24 (7.95%) patients (Tab. 1). PPMS prevalence is relatively lower than in other regions and individual countries.

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Table 1
Clinical and demographic characteristics of patients with PPMS

Index	PPMS
Age at the time of inspection, years	49.79±11.61
Women/men	13/11
Average age of first symptom	30.04±10.52
Average duration of the disease	15.5±8.45
Debut	
- Monosymptomatic	7 (29.17%)
- Polysymptomatic	17 (70.83%)

Among the examined women were dominated, but the difference was not statistically significant. These results coincide with data of other authors [2, 6]. Beginning at an early age - 30 years and the prevalence of polysymptomatic debut is typical for our cohort of patients. According to other studies, the typical age debut PPMS is 40 years [2, 7]. Early age of onset of illness is a feature of Volyn region.

The disability of the patients was due mainly to pyramidal and cerebellar symptoms (Tab. 2). Sensitive and visual disturbances were less observed. Patients with PPMS have severe impaired mobility. According to results of the survey, only 3 (12.5%) patients did not achieve EDSS score 6.0 at the time of examination. The patients also have a high rate of disease progression.

Table 2
The functional status of patients at the time of examination

Index	PPMS
FS scales:	
- Visual	0.87±0.89
- Brainstem	1.91±1.01
- Pyramidal	4.29±0.99
- Cerebellar	3.2±0.72
- Sensory	0.91±1.21
- Bowel/Bladder	2.25±1.25
- Cerebral	1.58±0.82
- Mobility	8.2±3.69
EDSS	7.04±1.3
Achieving EDSS 3,0	24 (100%)
Achieving EDSS 6,0	21 (87.5)
Index of progression	2.25±1.33

We evaluated the time to reach EDSS=6.0 for a PPMS comparing men and women (Fig. 1). We tried to find dependency of disability in patients with monosymptomatic and polysymptomatic onset of the disease (Fig. 2).

Fig. 1. Probability of achieving EDSS=6.0 depending on debut

Fig. 2. Probability of achieving EDSS=6.0 depending on gender

In our cohort of the examined, women reached EDSS=6.0 faster than men, but statistically significant difference was not found ($F=0.72$). There was no difference in achieving EDSS=6.0 depending on the debut of the disease ($F=0.73$).

Conclusions. Compared with the results of other authors, there are the following peculiarities of the PPMS in the inhabitants of Volyn region: beginning at an age of 30 with the prevalence of polysymptomatic debut. The most typical are disturbances of pyramidal and cerebellar systems with rapid disease progression. Evaluation features PPMS is important for the diagnosis and treatment, planning and providing health and social care.

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XÜLASƏ

VOLIN ƏHALİSİ ARASINDA BİRİNCİLİ PROQREDİENT DAĞINIQ SKLEROZUN GEDİŞİNİN XÜSUSİYYƏTLƏRİ

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Təqdim edilmiş məqalədə Volin rayonunda birincili proqredient dağınıq sklerozun (BPDS) gedışı tədqiq edilmişdir. Təyin edilmişdir ki, BPDS xəstəliyi olanların xüsusi çəkisi aşağı olmuşdur və 7,95% təşkil etmişdir. Bu pasiyentlər koqortunda xəstəliyin 30 yaş dövründə, əsasən piramid və beyincik sisteminin zədələnməsi ilə polisimptom başlanğıcı, əhəmiyyətli dərəcədə əlillik və xəstəliyin sürətlə progressivləşməsi xarakterdir. Əldə edilmiş məlumatlar BPDS xəstələrinə tibbi və sosial yardımın planlaşdırılmasında istifadə edilə bilər.

Açar sözlər: birincili-proqredient dağınıq skleroz, xəstəliyin gedışı.

РЕЗЮМЕ

ОСОБЕННОСТИ ТЕЧЕНИЯ ПЕРВИЧНО-ПРОГРЕДИЕНТНОГО РАССЕЯННОГО СКЛЕРОЗА У ЖИТЕЛЕЙ ВОЛЫНИ

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Изучено течение первично-прогрессирующего рассеянного склероза (ППРС) в Волынской области. Установлено, что доля больных с ППРС сравнительно ниже и составляет 7,95%. Данной когорте пациентов характерно полисимптомное начало заболевания, в 30летнем возрасте, с преимущественным поражением пирамидной и мозжечковой систем, со значительной степенью инвалидизации и быстрым прогрессированием заболевания. Полученные данные могут быть использованы для планирования медицинской и социальной помощи больным с ППРС.

Ключевые слова: первично-прогрессирующий рассеянный склероз, течение заболевания.

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