

Neuro-muscular rehabilitation approach with favorable results in a case of a tetraplegia after a cervical ganglioneuroma – case presentation

POPESCU Cristina ^{1,2}, BAILA Mihai ¹, DAIA Cristina ^{1,2}, ANDONE Ioana ^{1,2}, SERBAN Daniel ¹,
RADUCANU Cristian ¹, MANESCU Mihaela ¹, ONOSE Gelu ^{1,2}

Corresponding author: BAILA Mihai, e-mail: baila_mihai@yahoo.com

1.The Teaching Emergency Hospital “Bagdasar-Arseni” (TEHBA), Bucharest, Romania
2.The University of Medicine and Pharmacy “Carol Davila” (UMPCD), Bucharest, Romania

Abstract

This is the case of a 52-year-old male patient with tetraplegia and muscle respiratory failure, due to an extradural compressive cervical ganglioneuroma (C1-C2), with a history of neurological symptoms for 3 years. The tumor was completely surgically removed and the patient followed a neuro-muscular rehabilitation program (NMRP) for 4 months, consisting of rehabilitation nursing, physical and occupational therapy. At the end of NMRP the patient’s achievements were: unilateral supported walking, partial autonomous daily living functioning, the improvement of quality of life and facilitation of social and professional participation.

Keywords: *ganglioneuroma, spinal cord tumor, neuro-muscular rehabilitation program, non-traumatic spinal cord injury, spinal cord compression,*

Introduction

It is known that peripheral neuroblastic tumors develop from neural crest cells. These are multipotent stem cells arising from the ectodermal layer of the embryonic disc, that migrate to the developing tissues and organs in order to differentiate into various cellular lines: glial cells, sympathetic-adrenal cells, melanocytes, chondroblasts, osteoblasts (1,2,3). Ganglioneuroma (GN) is a type of tumor emerging from the neural crest cells with histological and clinical features of benignity: well differentiated cellularity of sympathetic-adrenal cells, organized stroma, rich vascularization(4). It grows slowly and can attain large dimensions(5,6). The ganglioneuroma most frequently develops in the paravertebral sympathetic ganglia and in the chromaffin cells of the adrenal glands and is mostly localized in the posterior mediastinum, retroperitoneum, and adrenal gland(1). The localization in the paraspinal cervical region is rare (7). These tumors occur in children but can also develop in adults, where they are mostly localized in the adrenal glands and affect males and females with ages between 40-50 years (1,7). Paraspinal ganglioneuroma can be diagnosed accidentally, by observing and touching a tumor mass or during an imagistic examination(7), but it can produce symptoms by compressing neighboring vascular and neurological structures. When the tumor extends towards the spinal cavity the patient can develop motor, sensory or autonomous deficiencies specific for the spinal cord compression syndrome(5,6). The treatment modalities of paraspinal GN depend on the oncological risk and the clinical and biological status of the patient (8,9). The elective treatment of the symptomatic paraspinal ganglioneuroma is the complete surgical tumoral excision (8,9). Despite being a benign tumor, the patient needs periodic neurosurgical follow-up

for discovering possible elements of disease progression (10).

The neuro-muscular rehabilitation program (NMPR) performed in specialized medical units comprises specific rehabilitation procedures like treatment of functional neuro-motor and autonomous disorders (genital-urinary, digestive), management of pain, psychological and educational support, and also other multidisciplinary medical services (11). The intensity and duration of the medullary compression, as well as the particularities of the tumor disease and patient’s comorbidities are elements that can predict the rehabilitation prognostic(11). The early recognition of neurological symptoms that may be caused by a non-traumatic spinal cord injury and the initiation of the specific medical, surgical and rehabilitation treatment could lead to a better functional autonomy of these patients.

Case presentation

This paper presents the case of a 52-year-old male, diagnosed in the Neurosurgery Department II of the Teaching Emergency Hospital Bagdasar-Arseni on March 2019 with spastic tetraplegia and muscle respiratory failure, due to a tumor located in the left vertebral arches C1-C2 and which extended intracranial towards the left vertebral foramina C1-C2 and C2-C3 (figure A, B, C). The injury was considered a neurosurgical emergency and completely removed, leading to spinal decompression.

The neuro-muscular rehabilitation program

The patient was admitted in the Neuromuscular Rehabilitation Department for 4 months. At admission the patient was immobilized in bed and needed permanent care from another person. The general objectives of the neuro-muscular rehabilitation program

(NMRP) were : improvement of gait and daily activities functioning in order to improve the patient’s quality of life and to recover social and professional participation capacities. The parameters which were evaluated before and after the NMRP were: 1. Motor and sensory abilities by using AIS scale; 2. Muscle spasticity – evaluated with modified Asworth scale. 3. Functional abilities – evaluated with Barthel score, functional independence measurement FIM (functional independence measure) and FAC score (functional ambulation categories). 4

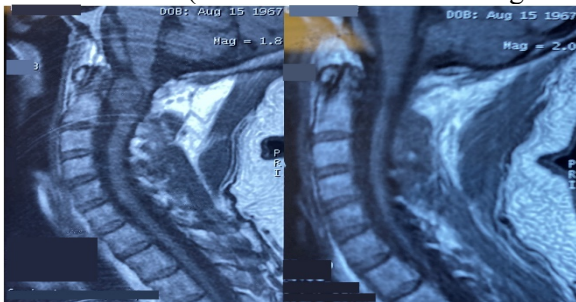


Fig.A. Left image – MRI T1 sagittal sequence showing a well encapsulated hypo-intense lesion, localized at C2 level, that severely compresses the spinal cord. Right image – MRI T1 sequence taken after the complete tumor excision, showing hypo-intense images suggesting edema / inflammation of the spinal cord.

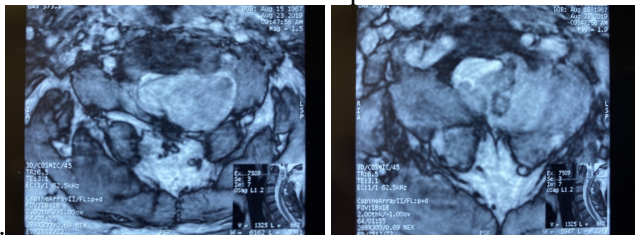


Fig.B MRI T1 transverse sequence - through the middle of the tumor mass (Fig.B) and the inferior region of the tumor mass Fig.C), showing severe spinal cord compression.

The histopathology report concluded that it was a ganglioneuroma. From the patient’s history we describe that he developed 3 years ago left finger paresthesia and the difficulty to button up his shirt with the left hand. In the last year, he began to have left lower limb muscle rigidity and weakness and paroxysmal nocturnal dyspnea. In the last weeks before hospital admission the patient developed a severe motor impairment of the right upper and lower limbs and needed wheelchair assistance for mobilization.

Quality of life, QoL (Flanagan) in order to quantify psychological, physical and social features. NMRP consisted of 5 sessions of physical therapy and occupational therapy per week (30-45 minutes each), for 4 months. At the beginning of motor rehabilitation program the patient was educated to maintain the correct body posture in bed and at the bed side. Rehabilitation nursing care and assistance were performed: bowel and bladder voiding; bladder control was trained by using

intermittent self-catheterization. The patient was mobilized in-bed every 4-6 hours during the entire program. The physical therapy consisted of passive, active-passive and active mobilizations of the upper and lower limbs, orthotic wearing for passive correction of articular stiffness and joint de-posturing of the hands and fingers (predominantly left side), due to muscle spasticity. After the therapeutic indication of complete mobilization, from the neurosurgeon, the patient was progressively trained for bed transfers, wheelchair transfers, and orthostatic posture and to participate to gym exercise sessions. The patient performed multiple exercises with various instruments: wall-bar verticalization , pulley and cable exercises with cuffs for the plegic hands (predominantly left side), motorized cycle ergometer and walking exercises. The occupational therapy was comprised of passive, active-passive, active mobilizations and various gripping exercises. The active mobilizations were performed without or with small resistance – which required orthostatic posturing of the hands (predominantly left side). The medical therapy consisted of administration of low molecular weight heparin (LMWH) until full mobilization and afterwards antiplatelet drugs, non-steroidal anti-inflammatory drugs, analgesics.

Results

The results of the evaluated scale

Before the NMRP		After the NMRP	
<i>motor AIS score</i>		<i>motor AIS score</i>	
Right upper limb = 9/25	Left upper limb = 4/25	Right upper limb = 13/25	Left upper limb = 10/25
Right lower limb = 9/25	Left lower limb = 5/25	Right lower limb = 13/25	Left lower limb = 9/25
<i>total sensory AIS score</i>		<i>total sensory AIS score</i>	
Right hemi-body = 23/58	Left hemi-body = 23/58	Right hemi-body = 24/58	Left hemi-body = 24/58
<i>Modified Asworth score</i>		<i>Modified Asworth score</i>	
Right upper limb = 1/5	Left upper limb = 1/5	Right upper limb = 1/5	Left upper limb = 1/5
Right lower limb = 1/5	Left lower limb = 3/5	Right lower limb = 2/5	Left lower limb = 3/5
<i>Barthel Index = 25/100</i>		<i>Barthel Index = 35/100</i>	
<i>motor FIM score = 13/91</i>		<i>motor FIM score = 28/91</i>	
<i>FAC score = 0</i>		<i>FAC score = 3</i>	
<i>QoL modif. after Flanagan = 56</i>		<i>QoL modif. after Flanagan = 60</i>	

The results emphasize the improvement of motor AIS score from 27 to 45 points. Total AIS sensory score slightly improved. Asworth spasticity score remained stationary and slightly increased for the right lower limb. Functional improvements were analyzed with FIM score which increased from 13 to 28, Barthel index that improved with 10 points and FAC score that described the evolution from in-bed immobilization (score=0) to walking capacities on flat ground under the supervision and with minimum support from another person (sc.= 3).

Discussion

At the admission on the Neuromuscular Rehabilitation Department, the patient had a right hemiparesis and a left hemiplegia, global proprioceptive impairments and right superficial hypoesthesia, bladder and bowel dysfunction. Prior to the stage of active neuro-muscular reeducation, the therapeutic means aimed to maintain joint mobility and soften the spastic muscle groups. Muscle spasticity caused joint flexion with mal posture of III-V fingers bilaterally (left > right), and also the tendency of bilateral forearm pronation. These were combated with passive mobilizations and orthotic correction. After therapeutic indication of full mobilization, from the neurosurgeon, the control and coordination of the scapular and pelvic trunk and belts were the primarily trained. Verticalization and gait were performed after the first two week of the NMRP. The patient needed a left ankle-foot orthosis in order to perform steps, for supporting the left drop foot. The patient's initial gait was unstable, mostly on account of the left lower limb, but with the support of the right lower limb and hands, gait exercises were performed. The results were the increased control of the left lower limb proximal muscles, with more efficient trunk balance and gait phases. The diaphragm muscle partial paresis and the usage of cervical and thoracic muscles for ventilation and phonation have predisposed to fatigue. The fine motor skills of the hands improved. The patient gained partial self-care autonomy in activities as eating, facial hygiene, hand hygiene, upper body clothing. Wheelchair self-propulsion was possible after efficiently using both upper arms.

Sensitivity disorders slightly improved. Bladder and bowel control were completely recovered. The functional favorable evolution could continue taking into account that the benign tumor was completely surgical removed, despite the patient's motor instability. The socio-professional reintegration of this active patient, to previous working activities is another long-term objective to be achieved, but the final evaluation needs to be done by the Romanian National Expertise Commission of Work Capacity and Disability.

Conclusion

This paper presents the case of a patient with severe tetraplegia, associated with respiratory symptoms, caused by a rare, benign, spinal tumor, namely cervical

paraspinal ganglioneuroma. The neurosurgical treatment, consisting of tumor excision and spinal decompression was followed by a complex and prolonged neuromuscular rehabilitation program, with favorable short and medium-term results: the improvement of daily activities functioning with partial autonomy (eating, personal hygiene, toilet use, clothing, transferring and walking). An interdisciplinary therapeutic approach is needed to maintain and improve these achievements and develop other long-term goals, especially regarding coordination, walking capacities and performing of daily living activities, which could lay the foundations for the physical reconditioning and socio-professional reintegration of this patient.

Conflict of interest

No conflict of interest declared. This study has approval of the Ethics Commission of the Teaching Emergency Hospital „Bagdasar-Arseni”, nr 3159 of 30.01.2020.”

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