

## RECUPERATION IN A CASE WITH CERVICAL EPENDYMOMA AND A ATYPICAL ONSET- CASE REPORT

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### Abstract

**Purpose.** This case presentation was meant to describe the atypical onset of the symptomatology, patient complaining in the beginning of walking troubles, progressively aggravated, the paresthesias and weakness in upper limbs being noticeable after a few months. The neurological examination correlated with the imagistic findings determined the surgery, with total surgical resection of the tumor, without postoperative complications with a good recuperation after initiated the kinetherapeutic programme and relieved symptomatology.

**Methods.** This case study relates the relationship between imagists and neurologists and recuperation team can have difficulties in establishing an accurate diagnosis and the same importance is the early initiation of a recuperation programme and that shows once again the benefits of team work.

**Results.** The particularity of presented case release in the pseudotumoral imagistic aspect, even though the symptoms, neurological signs and evolution of symptomatology were more suggestive for an ischemic stroke. We underline the importance of complete imagistic examination, in our case the absence of contrast substance made it impossible to perform a contrast MRI, examination necessary for a more accurate diagnosis and a correct design of passive and active recuperation has a good outcome.

**Conclusions.** This case presentation was meant to underline the importance of clinic diagnosis and correlation of symptomatology with imagistic findings, without minimize the role of imagistic examination, so necessary for an accurate diagnosis. Another important aspect is that sometimes even experienced imagists and neurologists can have difficulties in establishing an accurate diagnosis, and that shows once again the benefits of team work.

**Key words:** cervical ependymoma, imagistic examination, kinetic programme.

### Introduction

To underline the importance of clinic diagnosis and correlation of symptomatology with imagistic findings, without minimize the role of imagistic examination, so necessary for an accurate diagnosis. Another important aspect is that sometimes even experienced imagists and neurologists can have difficulties in establishing an accurate diagnosis, and that shows once again the benefits of team work.

It is a case study realized in hospital of Constanta, department of neurology in cooperation with Faculty of Physical Education.

### Results.

Ependymomas are the most common spinal cord tumors in adult patients, representing ~60% of all intramedullary tumors. Although they may occur at any age, they are noted to occur most frequently in middle-aged patients, and they have no particular gender predilection. There is an association between intramedullary ependymomas and NF-2 (K. Al moutaery, 1996; P. Celli, 1993; L. Cervoni, 1994). Likewise, most sporadic ependymomas also show mutations in NF-2 gene. The full spectrum of ependymoma is encountered in the spinal cord-cellular, tanycytic, malignant, mixed and myxopapillary ependymoma, as well as subependymoma. Most often, the myxopapillary ependymoma is found in the filum terminale or

cauda equine and therefore is considered an extramedullary tumor (16). Most spinal ependymomas are histologically benign, rarely show infiltrative growth, and do not form tumor capsules; however, the interface between the tumor mass and surrounding normal cord tissue is relatively well defined (F.J. Epstein, 1993; M.J. Fine, 1995; M. Graf, 1999).

In general symptoms are not specific, develop over several years, and are attributed to chronic cord compression causing myelopathy. The differential diagnosis includes other intramedullary tumors such as astrocytomas and hemangioblastomas as well as nonneoplastic processes, such as demyelinating diseases and cervical spondylotic myelopathy (F.Y. Donmez, 2008; D.R. Lefton, 1998; P.C. McCormick, 1990).

Dysesthesias caused by spinothalamic tract compression are the most common initial symptoms in numerous series. The MRI characteristics of intramedullary ependymoma, although on T1-weighted images, high signal on T2-weighted ones, marked but heterogeneous contrast enhancement, and a well-demarcated tumor margin. A tumor-related syrinx occurs in approximately 50% of intramedullary spinal cord tumors. The most instances, the tumor pathology is low grade, and therefore treatment objective is to achieve a gross-total resection without inflicting additional neurological morbidity

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(R.A. Morantz, 1979; F.G. Moser, 1992; A.R. Rezai, 1996).

The use of adjuvant therapy in the treatment of intramedullary ependymoma is controversial. There is a general consensus that radiation is not required if gross-total resection is achieved. However, after subtotal resection, the recurrence rate for these tumors is unacceptably high (Y. Nemoto, 1992; J.S. Schweitzer, 1992).

About the myxopapillary ependymoma who arise from the terminal filum, the most common symptoms are back and radicular pain.

We present the case of a 50 years old, male patient, who was admitted in our clinic for walking troubles and pain in the lumbar region and right lower limb, symptoms with progressive onset of approximately 2 months.

The first neurological examination rise the suspicion of a right L5 paretic lumbosciatic, and decided to perform a lumbar IRM examination that proved the existence of several disc hernia of multiple levels L3-L4, L4-L5, L5-S1. After consulting the neurosurgeon and having patient improvement, the disc hernia it was surgically removed with good postoperative evolution, without pain and walking troubles.

After few months patient returns in our clinic complaining of aggravating walking troubles, paresthesia and weakness in upper limbs, symptoms progressively installed in the last 3 months. The neurological examination showed Frankel D incomplete tetraplegia, with C4 level, predominant brachial; equal exaggerated tendon reflexes, superficial and profound loss of sensibility with C4 level. We decided to perform cervical and brain MRI (with and without gadolinium enhancement), that showed the presence of intramedullary mass extending from C3 to C4, with associated edema localized in the cervical and thoracic cord substance and brainstem (Fig. 1,2,3).

The clinical history and imagistic investigations were positively for a cervical intramedullary mass and imposed surgery, with total tumor resection and posterior medullary decompression. Biopsy of the lesion indicated a grade II intramedullary ependymoma.

After surgical procedure we begun an early recuperation.

Specific literature data show that approximative 75% of all ependymomas have intramedullary localization, being common in patients aged 15-45 years, and by histologic appearance they are myxopapillary subtype.

MRI of the spine with and without gadolinium enhancement is the study of choice. MRI permits evaluation of the cord substance itself for masses and associated findings such as edema,

hemorrhage, cyst, syringomyelia, and cord atrophy. Complete surgical resection is the treatment for intraspinal ependymomas. Total resection is generally curative, without postoperative irradiation. In addition to its role in identifying the tumor, preoperative imaging is essential in planning care.

This case study is particular by the atypical onset of the symptomatology, patient complaining in the beginning of walking troubles, progressively aggravated, the paresthesias and weakness in upper limbs being noticeable after a few months. The neurological examination correlated with the imagistic findings determined the surgery, with total surgical resection of the tumor, without postoperative complications and relieved symptomatology.

We initiate passive and active kinetics therapy program:

I. The initial phase, immediately after the surgery:

- in the early stages of the disease, the total or partial paralysis of the limbs is lax;
- the objective of the rehabilitation is the maintenance of the articular mobility in complete amplitudes and the prevention of muscular contractions;
- the correct posture of the limbs in functioning position;

The functional recovery of deficit of upper limbs

The rehabilitation of the upper limb started early, in our case – on the 5<sup>th</sup> day since the debut of the neuro-muscular deficit.

In the initial phase, when the proximal extremity of the upper limb is taken care of, there must be voluntary control of the shoulder and elbow, if possible, in different plans; all movements should be as far away as possible from the sinkinetic schemes. In the beginning, there is recommended to accentuate the spasticity of the hand to any movement of the upper limb root. Therefore, during the active mobilization of the proximal extremity, the hand shall be kept in an inhibition position, that is: total extension of the fingers and of the fist with the thumb in abduction.

-the passive mobilization of all of the joints of the affected limb is done gently, but it must be insisted in order to carry on the full amplitude of the movement. Every joint should be separately mobilized, holding at the extremities of the mobilized segments (a joint is not passively mobilized through another joint);

-the training of the body symmetry is made through bilateral activities, then alternative unilateral ones, and finally through reciprocal activities;

-when the overall condition allowed it, (on the 30<sup>th</sup> day since the debut), the Kabat technique is

applied, the diagonals for the upper limb; once the spasticity is installed, the new conditions of the neuro-muscular deficit forced the adjustment of the therapeutic tactics;

-it is important to know that with some hemiplegics there may reside a lack of usage of the hand, although motility is recovered. This is explained by the profound sensory disorders due to the involvement of the upward sensory paths which are very close on the pyramidal path, at the level of the cortex and the inner capsule.

The functional rehabilitation was proximally started, then distally.

Afterwards, the evolution was the following. During hospitalization, the patient went through:

1. Initially, the hand had no voluntary command or can only flex through stereotypic movement.
2. At release, she can actively flex her fingers and thumb, but she cannot extend them except in one position; we explain that it is required to have precision in movement, and not force and execution speed.

The rehabilitator tries the "awakening" of the extensor muscles, with the help of the facilitating techniques, especially those that use the position shifts.

The major objective of the lower limb rehabilitation is thus defined: obtaining a balanced command on different antagonist groups and eliminating the sinkinetics in order to recover a walking as close to normal as possible. Most statistics give percentages between 85 and 95 of recovering walking for hemiplegics.

Methodic indications: in order to prevent the typical flexion stiffness and the external rotation of the hip, knee flexion and equinovarus, we install the patient so as to have the basin flat on the bed, with no flexion of the hip and knee, the lower limb totally coupled so as to avoid its fall in external rotation, the feet is maintained at 90° on the lower leg.

Spasticity is announced through the exaggeration of the ROT and usually begins with the abductors of the thighs and the quadriceps, in our case, on the 30<sup>th</sup> day.

For a good rehabilitation of the walk, it is necessary to make a thorough analysis of the muscular deficit, of the repartition and intensity of the spasticity, of the intensity of the sin-kinetics, to sum up, it is necessary to make a functional evaluation of the patient.

The muscular deficit is mostly recorded (the general scheme of hemiplegia) on the following muscles: psoas, abductors and internal rotators of the hip, the knee flexors, and the leg dorso-flexors. The ischio-tibial muscles are partially respected.

During the evolution, the deficit is modified; the first muscles to recover voluntary contraction capacity are the abductors, the quadriceps and then gluteus maximus.

The muscles that remain most often, deficient, are the common extensor of the fingers, the peroneals, and the middle and small gluteus.

Bearing this in mind, the importance of the correct positioning during the flaccidity period is thoroughly justified.

The rehabilitation therapy develops in two phases:

A. In the initial phase, when the proximal extremity of the upper limb is taken care of, there must be voluntary control of the shoulder and elbow, if possible, in different plans; all movements should be as far away as possible from the sinkinetic schemes. In the beginning, there is recommended to accentuate the spasticity of the hand to any movement of the upper limb root. Therefore, during the active mobilization of the proximal extremity, the hand shall be kept in an inhibition position, that is: total extension of the fingers and of the fist with the thumb in abduction.

-the passive mobilization of all of the joints of the affected limb is done gently, but it must be insisted in order to carry on the full amplitude of the movement. Every joint should be separately mobilized, holding at the extremities of the mobilized segments ( a joint is not passively mobilized through another joint);

-the training of the body symmetry is made through bilateral activities, then alternative unilateral ones, and finally through reciprocal activities;

-when the overall condition allowed it, (on the 30<sup>th</sup> day since the debut), the Kabat technique is applied, the diagonals for the upper limb; once the spasticity is installed, the new conditions of the neuro-muscular deficit forced the adjustment of the therapeutic tactics;

-the traction exercised by the upper fascicles of the trapezius and the sternocleidomastoid flexes the head on the affected side and rotates it on the healthy side.

-the body has a lateral inflexion on the hemiplegic part, with lifting and retrusion of the basin with the descent and retrusion of the humeroscapular belt due to the traction exercised by the broadest muscle of the back.

--the entire hemiplegic part is rotated backwards; in order to inhibit or reduce the spasticity that generates this attitude, as well as to correct the anomalous tonic reflexes, the change of the key points: the neck, the spine, the scapular belt and the pelvic belt, the fingers and toes: these reflex positions should be localized with every patient and corrected every time it is necessary; at the

same time there should be attempted to reduce the spasticity through the methods described in the general part;

-it is important to know that with some hemiplegics there may reside a lack of usage of the hand, although motility is recovered. This is explained by the profound sensory disorders due to the involvement of the upward sensory paths which are very close on the pyramidal path, at the level of the cortex and the inner capsule.

The prognosis of the functional rehabilitation of the hand is linked to many aspects, among which we mention some references to the etiology and topography of the lesion:

-the most serious, from a functional point of view, and, unfortunately, the most frequent, are the cortical or capsular lesions following an ischemia, such as is our case in the ICA territory. If at the debut of the illness, the functional prognosis cannot be determined, two months later it may be known according to: the topography of the lesion, the importance of the sensory and motility disorders.

-the functional prognosis is initially mediocre, and its primary purpose is that of preventing the elbow-shoulder syndrome and learning how to use the arm as a basic helper, as well as the preservation of the future, in case the

rehabilitation should occur (after a year), which is sometimes the case.

The functional rehabilitation was proximally started, then distally.

B. Afterwards, the evolution was the following. During hospitalization, the patient went through:

1. Initially, the hand had no voluntary command or can only flex through stereotype movement.

2. At release, she can actively flex her fingers and thumb, but she cannot extend them except in one position; we explain that it is required to have precision in movement, and not force and execution speed.

#### Conclusions

Given the slow growth and the well-circumscribed quality of these tumors, symptoms generally progress slowly, and the tumors are often present in patients long before diagnosis. It is important to remember that this type of cases can have atypical onset and therefore an accurate anamnesis correlated with the progressive characters of symptomatology could be helpful for diagnosis.

Although it is a rare and slow growing neoplasm, early detection is critical for optimal postoperative functional outcome that is directly related to the preoperative functional status.



Fig. 1: Cervical MRI

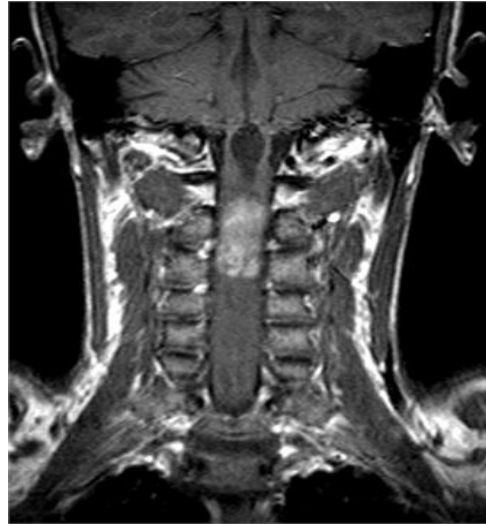


Fig. 2: Cervical MRI

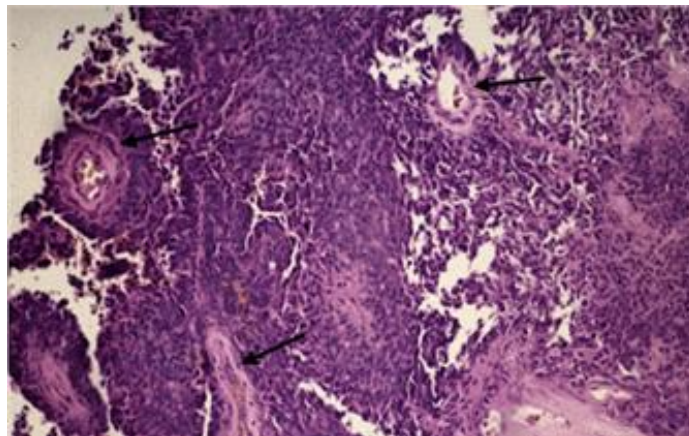


Fig.3: HISTOTAPHOLOGICAL EXAM

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