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Pneumologia

Severe restrictive syndrome in context of bilateral diaphragmatic paralysis - case series

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Abstract

English:

The authors present a miniseries of two clinical cases of bilateral diaphragmatic paralysis, of different etiologies, which provide the opportunity to discuss the particularities of the clinical manifestations, the paraclinical investigations necessary to establish a positive diagnosis, the elements of differential diagnosis, the general therapeutic principles addressed to respiratory failure secondary to diaphragmatic paralysis.

Keywords

diaphragmatic paralysis • Parsonage-Turner syndrome • amyotrophic lateral sclerosis

Sindrom restrictiv sever în contextul paraliziei diafragmatice bilaterale - serie de cazuri

Rezumat

Romanian:

Autorii prezintă o miniserie formată din două cazuri clinice de paralizie diafragmatică bilaterală, cu etiologii diferite, oferind oportunitatea discutării particularităților manifestărilor clinice, investigațiilor paraclinice necesare pentru stabilirea unui diagnostic pozitiv, elementelor de diagnostic diferențial și principiilor terapeutice generale adresate insuficienței respiratorii secundare paraliziei diafragmatice.

Cuvinte-cheie

paralizie diafragmatică • sindromul Parsonage-Turner • scleroză laterală amiotrofică

1. Introduction

Bilateral diaphragmatic paralysis is a rare entity, with varied etiologies, most of the time being of other specialties competence. The impact on respiratory mechanism and gas exchange is usually very important, which is why patients affected by this pathology are addressed to pulmonologists for severe restrictive syndrome and respiratory failure.

2. Clinical Problems

The authors present cases of two patients, admitted to the National Institute of Pneumophthisiology „Marius Nasta" in 2022 (patient 1), respectively in 2017 (patient 2).

The first patient is a 28-year-old male, ex-smoker 12 packages-year, with professional cement powders exposure of 14 years, unvaccinated COVID-19, without chronic diseases, which initially presented with progressive exertional dyspnea

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and the association of orthopnea that started 36 hours before the presentation to another health facility, preceded by pain at the level of the scapular with professional cement powders exposure of 14 years, belt, for 2 weeks, for which he receives pain reliever treatment at home, without relieving the symptoms, considered as a depressive-anxious disorder. Subsequently, he presented at a distance of 3 months to the our emergency department following effort dyspnea with orthopnoea, right hemithoracic pain, hemoptysis.

Clinical data: at admission, out patient was tachypneic, with a respiratory rate of 30-40/min, with SpO2 = 97% while on oxygen via nasal cannula (2L / min), normal conformed thorax, 1/2 inferior dulness bilaterally, without rales lung sounds. The patient tolerates lateral decubitus, but does not tolerate dorsal decubitus. Without clinical phenomena of hypoxemia with hypercapnia (cyanosis, disorientation, flapping tremor, etc.) No signs of peripheral neuromuscular disease.

The second patient was a 63-year-old male, ex-smoker 90 packages-year. He had a history of dyslipidemia, high blood pressure, coronary artery disease, chronic heart failure NYHA Class II, who presented to the our emergency department following acute onset of breathlessness and secondary algic syndrome after a chest household trauma.

Clinical data: SpO2 = 88% while on a oronasal mask (3L / min), thoracic kyphosis1/2 inferior matity bilaterally, without rales lung sounds. Paradoxical thoraco-abdominal movements. Impossibility of adopting the supine position. The patient shows clinical phenomena of hypoxemia with hypercapnia. No signs of peripheral neuromuscular disease.

Differential Diagnosis

Disorders of the motor neuron (Multiple Sclerosis, SLA - Charcot / Lou-Gehrig's disease, the anterior horn of the spinal cord syndrome, Post-polio syndrome, the high medullary traumatisms), myopathies, muscle dystrophies, Guillain-Barré syndrome, the Parsonage-Turner syndrome (brachial plexus neuropathy), Pompe disease (alpha-glucosidase acid deficiency or acid-maltase deficiency), postinfectious (herpes zoster, HIV/AIDS, septic condition / critical patients, etc.) , polineuropathies of other causes, after cardiac surgery or lung transplantation, neurosarcoidosis, mediastinal humors, Charcot-Marie-Tooth syndrome, POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, M protein growth, skin changes), idiopathic [1].

3. Results of clinical findings

Patient 1.

Laboratory: WBC 14.35 10^3/mcL (3.91-10.90), NEU 89.40% (41-70.7), LYM 5.10% (19.1-47.9), EO 0.00% (0.03-0.59), D-dimer 86 ng/mL (0-243), APTT 43.50 sec (25.1-36.5), Fibrinogen 479 mg/dL (238-498), VSH 13 mm/h (2-15), Ferritin 369.2 ng/mL (25-350), PCT 1.28 ng/mL (0-0.5), negative ANA test, negative HIV test, negative HCV-antibody test, negative HBs-antigen test, negative RT-PCR tests (SARS-COV-2, Flu A, Flu B, RSV).

EKG and Echocardiography: Sinus tachycardia; mild pulmonary arterial hypertension (PAH)

EAB: pH 7.40, pCO_2 40.8 mmHg, pO_2 75.2 mmHg, sO_2 95.2%

Lung volumes by plethysmography testing: severe mixed lung disease (FVC 55.2%, FEV_1 51.2%, FEV_1 % VC MAX 78.59%, MEF50 42.2%)- with bronchodilator reversibility, moderate decreased DLCO 43.4% – treatment recommendation with ICS/LABA 1x2puff / day, SABA if necessary. Subsequently, one month away, spirometry was performed in orthostatism, respectively in clinostatism, with the result: -21% FVC (1030 ml) between the two examinations.

Pleural ultrasound: Diaphragm mobility on bilateral sides were reduced during deep breathing. There was significant difference in the mobility of the bilateral diaphragm on the right side compared to the left during deep breath and decreased diaphragmatic excursion during deep exhale (1-1.5cm); without liquid overflow into the pleural cavities.

Chest CT-scan (fig.1, fig. 2): important ascent of diaphragmatic domes, mainly on the right; basal condensation with alveolar filling and presence of internal air bronchogram in the middle lobe, with pneumonic substrate.

Bronchoscopy: bilateral condensation (aerobic culture result from the bronchoalveolar aspirate from the middle lobe: *H. Parainfluenzae*) - recommendation of antibiotherapy according to antibiogram.

Polysomnography: average SpO_2=92%; minimum SpO_2=82%; decrease in SpO_2>3%=5.2 sleep events/h;

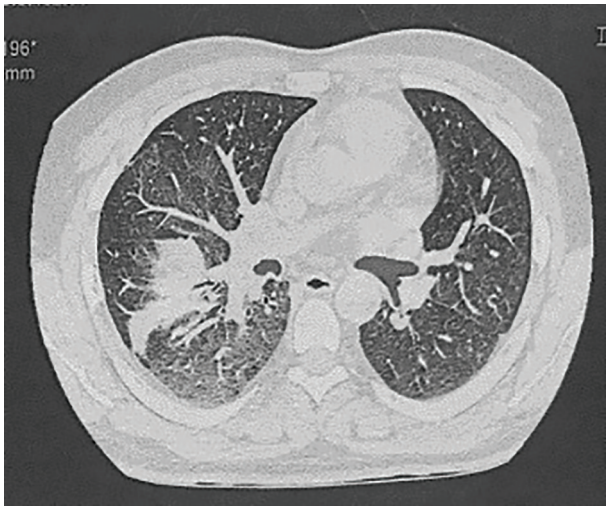

Figure 1.

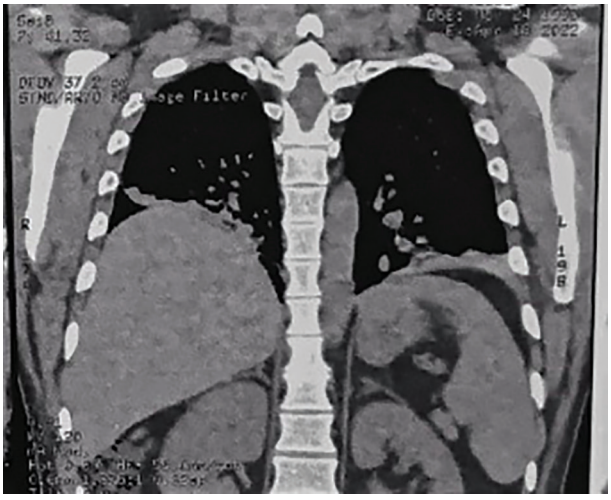

Figure 2.

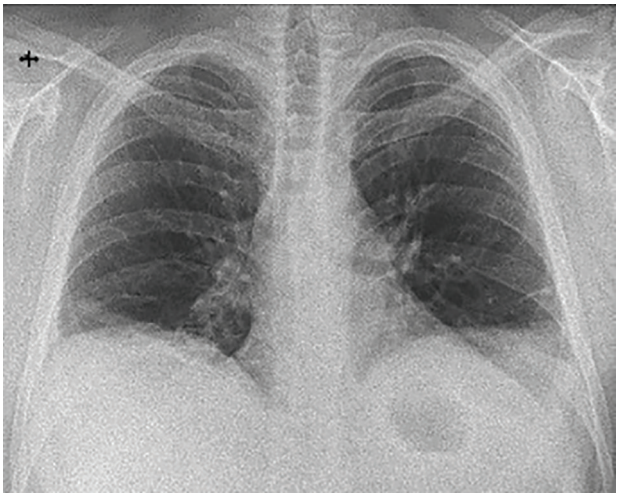

Figure 3.

AHI=2.7 sleep events/h with hypopneal predominance - recommendation of APAP/BiPAP(IPAP/EPAP 7/4cmH_2O)

Neurological evaluation:

EMG: lack of response of diaphragmatic muscle to phrenic nerve stimulation, normal studies on the nerves and bilateral deltoid muscles, with excluding polyneuropathy or damage to the neuromuscular junction; no signs of motor neuron damage. Therfore, the suspicion of Parsonage-Turner syndrome limited to the phrenic nerve is raised. Under these conditions, treatment with oral Methylprednisolone was initiated, with the weekly progressive decrease of the doses. Diagnostic test with Miostin intravenous injection was negative.

The patient was discharged from the hospital and was advised to continue NIV at home according to the recommended parameters; respiratory physiotherapy; will continue oral antibiotic treatment with Amoxicillin 875mg/Clavulanate 125mg and Levofloxacin 500mg for another 7 days, will continue inhaled treatment with ICS/LABA 1x2puff / day, SABA if necessary; treatment with Methylprednisolone 16mg with progressive decrease in dosages; spine MRI; pulmonological and neurological evaluation after 3 months.

Currently, the patient continued the NIV with the same parameters, the clinical situation and the imaging aspect have improved **(fig. 3)**, EAB: normoxemia, normocapnia, lung volumes by plethysmography testing described FVC = 50.4%, FEV_1 = 46.2%, MEF50 = 35.1% DLCO is moderately decreased and Kco within normal limits. The patient is periodically reevaluated pulmonologically and neurologically.

Patient 2.

EAB: pH 7.28, pCO2 91,1 mmHg, pO2 70,4 mmHg, sO2 92,6%

Lung volumes by plethysmography testing: -12% FVC (450ml) compared to a previous test 6 months ago (FVC

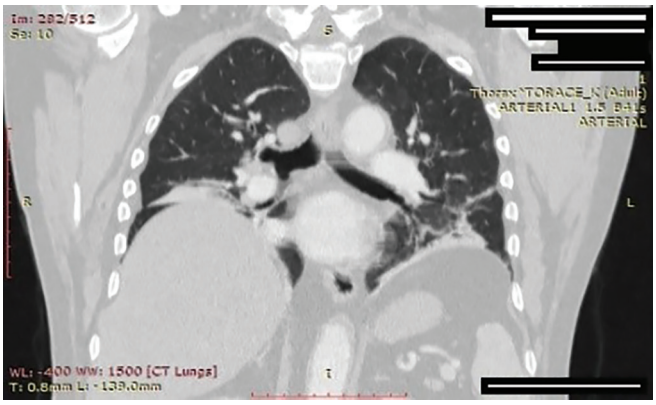

Figure 4.

42.8%, DLCO was moderately decreased and Kco within normal limits).

Pleural ultrasound: both diaphragmatic domes much raised, with absent respiratory movements; without liquid overflow into the pleural cavities.

Chest CT-scan (fig.4.): important ascent of the diaphragmatic domes, predominantly on the right side.

Polysomnography: TIB=390 min, TST=192 min, sleep efficiency 49,4%, REM sleep cycles 10,5 min; estimated AHI =10/h, of which > 50% central apnea (AC=7/h) - predominantly central sleep apnea syndrome, mild, possibly due to neuromuscular pathology.

Neurological evaluation:

EMG: examination compatible with motor neuropathy, with the presence of signs of active denervation in the muscles of all body segments.

Other tests performed: lumbar puncture, cranial CT-scan, transcranial magnetic stimulation.

After all the investigations carried out, the diagnosis of amyotrophic lateral sclerosis was made - the form with bulbar onset (progressive bulbar paralysis). The patient followed NIV at home (BIPAP S/T, later AVAPS) and home high-flow nasal cannula therapy (HFNC), oral treatment with Riluzol 50mg x2/day, physiotherapy. The pacient died within 1 year after emergency medical admissions.

4. Discussions

Both patients were men, ex-smokers, clinically with a history of progressive exertional dyspnea and the association of orthopnea, diagnosed by imaging with bilateral diaphragmatic ascent, with complex respiratory function tests with severe restrictive ventilatory dysfunction, performed in clino- and ortho-statism. The full set of investigations included fibrobronchoscopy, polysomnography, arterial blood gas (ABG) analysis, immunological blood profile, neurological evaluation with electromyogram (EMG) and lumbar puncture. In the case of the first patient, pneumomediastinal pathologies were excluded by chest CT-scan, as well as by bronchoscopy, neuromuscular junction by EMG, of the type of isolated metabolic or paraneoplastic neuropathies, but a spinal pathology in the cervical region could not be excluded, because the patient does not tolerate decubitus in order to carry out the examination of the spine by MRI. Considering the absence of symptomatology evolution and even discrete improvement under cortocosteroid treatment, it was not considered appropriate to initiate an empirical immunosuppressive treatment, but only to monitor the evolution and possibly complete the investigations, if the patient will allow it.

Other tests that could have been necessary for the diagnosis: the measurement of the transdiaphragmatic pressure and the measurement of the maximum inspiratory pressure (PImax), which were not performed in the presented cases [2,3].

Surgically, robot-assisted thoracoscopic diaphragmatic plication is a safe procedure that can significantly improve dyspnea and is associated with a shorter hospital stay compared with the open approach [4].

5. Conclusions

Bilateral diaphragmatic paralysis must be suspected in all situations of restrictive dysfunction with normal lungs ± signs of peripheral neuromuscular damage. However, the positive diagnosis is laborious and requires a close multidisciplinary collaboration. NIV is often sufficient, diaphragm plication and invasive mechanical ventilation rarely have to be resorted to, and the evolution and prognosis differ depending on the etiology of the bilateral diaphragmatic paralysis.

Authors contributions

1. Lorena-Nicoleta Diaconu
2. Raluca Bobocea † Anca Macri
3. Bianca Valu † Ionela Popa

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Informed Consent Statement

Informed consent was obtained from both patients.

Data Availability Statement

Data used to support the findings of this study are available from the corresponding author upon request.

Conflicts of Interest

The authors declare no conflict of interest.

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