SERVIZIO SANITARIO REGIONALE
EMILIA-ROMAGNA
Azienda Ospedaliera di Reggio Emilia
Arcispedale S. Maria Nuova
Istituto in tecnologie avanzate e modelli assistenziali in oncologia
Istituto di Ricovero e Cura a Carattere Scientifico

50 1965 2015 SANTAMARIANUOVA
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Life-Threatening Laryngeal Involvement in Isaacs Syndrome

Codeluppi L, Canali E, Rizzi R, Sabadini R, Marcello N, Motti L

UOC Neurologia, Aricspedale Santa Maria Nuova, Reggio Emilia

Introduction. *Isaacs Syndrome [1] is an extremely rare disease characterized by diffuse peripheral nerve hyperexcitability provoking continuous involuntary muscle activity, cramps, fasciculations and myokimia. In most cases Isaacs syndrome ascribetd to an autoimmune process targeting potassium channels in the proximal nerve membrane and in about 50% of the patient serum antibodies against potassium channels may ben found. Autoimmune Isaacs syndrome may be either primary or secondary, the latter usually related to a neoplasm, constituting thus a paraneoplastic syndrome. Issacs symptoms hyperexcitability symptoms are in most instances only annoying for the patient without constituiting an hazardous condition, moreover the clinical picture often exhibits a good response to immunomodulatory and antiepileptic treatment. We present the case of a patient with a progressive and life-threatening laryngel spasm related to an unusual form of aggressive Isaacs refractory to medical treatment.*

Case Description. *A 65-years-old man presented continuous unexplained diffuse sweating followed after some months by diffuse and continuous muscle twitching. Patient's relatives also repoerted to have noticed in the last year the presence of whistling and wheezing during normal breating in certain position or during prolonged talking. Neurological examination at the admission disclosed diffuse myokimia, unilateral orizzontal gaze paralysis and slight weekness localized only to the deltoid muscles, without any further neurological sign. Electromyography confirmed the diagnosis of Isaacs syndrome, without any evidence of further peripheral neurological diseases. Since the presence of a pacemaker for vasovagal syncopes, MRI scans couldn't be performed. Cerebrospinal fluid analysis was unremarkable and chest-abdomen CT scan discolsed only pleural deposits ascribed by our pneumologist to the resuts of previous professional expositions. Anti-potassium channels antibodies were negative. Laryngoscopy disclosed an adduction spasm of both vocal cords with a subtotal occlusion of the laryngel space. Plasmapheresis was performed with improvement of all symptoms, including the laryngeal ones since whistling and wheezing totally disappeared. However after one moth all the symptoms relapesed and exhibited progressive worsening; a second trial of plasmapheresis was ineffective so steroid therapy (up to 1 mg/kg/die of prednisone) and carbamazepine (up to 800 mg/die) was initiated as suggested by the literature [1]. The patient experienced a strong improvent but after a further month all the symptoms rapidly worsened again with the appearance of subacute severe respiratory insufficency so it was necessaryo to practice an urgent laser cordotomy. In the following months also a diffuse axonal sensorymotor neuropathy appeared. Screening for heavy metals intoxication (mercury and manganese) disclosed only a slight increase of manganese level with normal mercury level. Follow-up is still ongoing.*

Discussion. *We described the case of a patient with Isaacs syndrome presenting an unusual resistance to all the therapies and with a life-threating evolution which is extremely rare for this disease. So far indeed only one case of laryngeal involvement in Isaacs syndrome has been reported [2]. Even if our patient had negativity for anti-potassium channels antibodies, the autoimmune etiology is suggested by the response, even if temporary and scarce, to all the immunomodulating treatments which have bene practised. Mercury intoxication have recently been claimed as mimiking autoimmune Issacs syndrome [3], but in our case mercury levels were within the normal limits. Even if the final diagnosis is still partially open, we think that the atypical features of the patients, associated with the appearance of a sensorimotor unexplained axonal polyneuropathy, suggests the paraneoplastic etiology probably related to an occult cancer. Neurological and oncological follow-up are still ongoing.*

Conclusions. *Even if Isaacs syndorme is usually considered a bening disaese, our case suggests to clinicians that this disease can be also life-threatening when laryngeal muscles are involved. We highlight that our patient, despite an almost complete laryngeal occlusion, was almost asymptomatic in the early course of the disease and that the respiratory insufficency which required urgent cordotomy appeard rapidly with a fast progressive course. We thereofre suggest clinicians to investigate laryngeal functions and motility in patients with Isaacs syndrome at the time of the diagnosis and periodically during the course of the disease.*

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