

2. Kim D, Song I, Keum S, Lee T, Jeong MJ, Kim SS <ET-AL>. Lack of the burst firing of thalamocortical relay neurons and resistance to absence seizures in mice lacking alpha(1G) T-type Ca(2+) channels. *Neuron*. 2001;31:35–45.
3. Rosenfeld WE, Benbadis S, Edrich P, Tassinari CA, Hirsch E. Levetiracetam as add-on therapy for idiopathic generalized epilepsy syndromes with onset during adolescence: analysis of two randomized, double-blind, placebo-controlled studies. *Epilepsy Res*. 2009;85:72–80.
4. Blumenfeld H, Klein JP, Schridde U, Vestal M, Rice T, Khera DS, et al. Early treatment suppresses the development of spike-wave epilepsy in a rat model. *Epilepsia*. 2008;49:400–9.
5. Suzuki S, Kawakami K, Nishimura S, Watanabe Y, Yagi K, Seino M <ET-AL>. Zonisamide blocks T-type calcium channel in cultured neurons of rat cerebral cortex. *Epilepsy Res*. 1992;12:21–7.
6. Hoy SM. Zonisamide: a review of its use in the management of adults with partial seizures. *Drugs*. 2013;73:1321–38.
7. Wilfong A, Schultz R. Zonisamide for absence seizures. *Epilepsy Res*. 2005;64:31–4.
8. Hurst R, Chiota-McCollum N, Tatum W. Adult absence semiology misinterpreted as mesial temporal lobe epilepsy. *Epileptic Disord*. 2014;16:471–6.

J.L. Camacho Velásquez\*, E. Rivero Sanz, A.A. Sanabria Sanchinel, S. Santos Lasaosa, J.A. Mauri Llerda

*Servicio de Neurología, Hospital Clínico Universitario Lozano Blesa, Zaragoza, Spain*

\* Corresponding author.

E-mail address: [jlcv2002@hotmail.com](mailto:jlcv2002@hotmail.com)

(J.L. Camacho Velásquez).

2173-5808/

© 2016 Sociedad Española de Neurología. Published by Elsevier España, S.L.U. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## Thymic cyst associated with myasthenia gravis<sup>☆</sup>



### Quiste tímico asociado a miastenia gravis

Dear Editor,

Thymic cysts account for 1%–5% of anterior mediastinal masses,<sup>1</sup> and may be congenital or acquired. Acquired thymic cysts have been associated with such systemic diseases as lupus erythematosus, Sjögren syndrome, rheumatoid arthritis, Hashimoto thyroiditis, HIV infection, and in very rare cases, with myasthenia gravis (MG).<sup>2</sup>

We present an extremely rare case of MG associated with thymic cyst.

The patient was a 34-year-old former smoker with no history of disease. He attended our department due to a 2-month history of fatigue of the facial muscles, with no additional symptoms. The physical examination revealed mild left ptosis, which did not increase with maintained vertical gaze, as well as weakness of the tongue and the left orbicularis oculi and oris muscles.

We made a diagnosis of MG; this was confirmed by the results of an anti-acetylcholine receptor (anti-AChR) antibody analysis (22.1 nmol/L) and a single-fibre EMG in the left orbicularis oculi muscle (semi-pathological jitter; 47% blocking). These findings are compatible with a postsynaptic alteration in neuromuscular transmission. A mediastinal computerised tomography (CT) scan revealed a cystic lesion in the prevascular space, measuring 78 × 79 × 77 mm, with well defined contours and thin walls, of liquid density, showing no contrast uptake and no evidence of lymph node involvement (Fig. 1).

The mediastinal lesion was surgically resected 4 months after diagnosis. The anatomical pathology study identified it as a unilocular thymic cyst with follicular hyperplasia. A cytology study of the pleural fluid was negative for malignant cells, suggesting a cyst of lymphoid content.

Following diagnosis, the patient was started on treatment with pyridostigmine dosed at 60 mg every 8 hours. This partially improved the ptosis and the strength of the orbicularis oris muscle. One month later, the patient started a slow tapering course of prednisone, beginning at a weight-adjusted dose of 80 mg/48 hours. In the second month



**Figure 1** Chest CT scan. The mass in the anterior mediastinum is compatible with thymic cyst.

<sup>☆</sup> Please cite this article as: Morollón N, Guerrero P, Duarte J. Quiste tímico asociado a miastenia gravis. *Neurología*. 2018;33:405–407.

of treatment, the orolingual weakness showed a marked improvement; the orbicularis oculi muscle weakness persisted until 3 months later, with the patient receiving prednisone at 30 mg/48 hours. The surgical intervention enabled the gradual reduction of the corticosteroid dosage over the following 4 months, reaching 5/2.5 mg on alternating days. Symptoms reappeared at this point, resulting in corticosteroid dosage being increased once more to 10/5 mg on alternating days, and the introduction of azathioprine. The patient remained asymptomatic following the withdrawal of prednisone, although pyridostigmine continues to be necessary.

Thymic cyst is a well defined, water-density lesion of the anterior superior mediastinum. The mass may be congenital or acquired. Congenital cysts are unilocular, with thin, translucent walls and atrophic tissue with no inflammation. Acquired thymic cysts tend to be multilocular, with thicker, potentially fibrous walls and hyperplastic thymic tissue. These lesions are usually asymptomatic, although they occasionally cause dyspnoea, coughing, or chest pain due to compression of adjacent structures.<sup>3</sup>

Thymoma, which accounts for 95% of cases of thymic epithelial tumours, presents as a homogeneous, solid, soft-tissue mass with well-defined borders, occasionally displaying uniform contrast uptake and calcification.<sup>4</sup> Up to 40% of cases of thymoma may involve a cystic component; however, it is very rare for the cyst to account for almost the entire mass.<sup>5</sup>

Radiological differential diagnosis of thymic lesions can be complex. Thymic cysts complicated by bleeding or infection may be mistaken for solid masses due to the increased density in CT images.<sup>6</sup> Thymomas may also develop haemorrhagic, ischaemic, or cystic areas, simulating simple thymic cysts.<sup>7,8</sup> Chest MR images may be of assistance. In MR images, thymoma appears as a homo- or heterogeneous mass. It is hypointense on T1-weighted and slightly hyperintense on T2-weighted sequences, showing restricted diffusion and contrast uptake, whereas thymic cysts are hypointense on T1-weighted and hyperintense on T2-weighted sequences and do not display restricted diffusion or contrast uptake. If the diagnosis remains uncertain, an <sup>18</sup>F-FDG PET scan may be helpful, as this technique provides information regarding metabolism.<sup>9</sup> However, precise classification of a cystic mediastinal lesion requires a histopathology study, particularly if the wall is focally thickened.<sup>10</sup>

Thymic abnormalities affect up to 85% of patients with MG (follicular hyperplasia affects 70% and thymoma 15%)<sup>11</sup>; however, a literature search only identified 6 cases published to date of patients with MG associated with thymic cyst. In 1957, Fongi et al.<sup>12</sup> described a case of myasthenic crisis following excision of a thymic cyst. In 1993, Peacey and Belchetz<sup>10</sup> presented a patient with ocular MG and thymic cyst. In 1995, Okumura et al.<sup>13</sup> reported a case of MG associated with multilocular thymic cysts. In 2012, Mishra et al.<sup>14</sup> described 3 cases of MG with unilocular thymic cysts.

Little is known about the progression of MG following surgery in these cases. In the series reported by Mishra et al.,<sup>14</sup> 2 patients clinically improved and one worsened following resection of the cyst. In other cases, one patient had a myasthenic crisis following surgery,<sup>15</sup> and another

did not improve.<sup>10</sup> Our patient experienced a temporary improvement, lasting less than 6 months, and eventually received long-term immunosuppression.

Thymectomy is included in the treatment protocol for MG and has been demonstrated to achieve clinical improvement, although the benefits of the intervention are not fully understood.<sup>16</sup> The results of the "Thymectomy trial in non-thymomatous myasthenia gravis patients receiving prednisone therapy",<sup>17</sup> currently underway, will provide information on these benefits in non-thymomatous patients.

Although thymic cyst occurs very infrequently, we believe it should be included in the spectrum of MG-associated thymic conditions. Our patient's transient improvement following the excision of the cyst seems to confirm its involvement in the pathophysiology of this autoimmune disease.

Thymectomy would therefore be indicated in these cases in order to rule out the presence of tumours in the cyst walls and/or to delay the start of prolonged immunosuppression.

## References

1. Fuentes J, Fernández F, Fuentes A, Rivera M, Spencer L, González R. Thymic cysts, an unusual disease of the mediastinum. *Revista Anacem*. 2013;VII.
2. Shi X, Nasser F, Berger DM, Nachiappan AC. Large multilocular thymic cyst: a rare finding in an HIV positive adult female. *J Clin Imaging Sci*. 2012;2:55.
3. Won Choi Y, Page MacAdams H, Chol Jeon S, Kyung Hong E, Kim Y-H, Im J-G, et al. Idiopathic multilocular thymic cyst: CT features with clinical and histopathologic correlation. *AJR Am J Roentgenol*. 2001;177:881–5.
4. Baas P, Rhodius R. Thymoma update 2011. *Eur J Cancer*. 2011;47 Suppl 3:S315–6.
5. Takahashi K, Al-Janabi NJ. Computed tomography and magnetic resonance imaging of mediastinal tumors. *J Magn Reson Imaging*. 2010;32:1325–39.
6. Tecce PM, Fishman EK, Kuhlman JE. CT evaluation of the anterior mediastinum: spectrum of disease. *Radiographics*. 1994;14:973–90.
7. Moran CA, Suster S. Thymoma with prominent cystic and hemorrhagic changes and areas of necrosis and infarction: a clinicopathologic study of 25 cases. *Am J Surg Pathol*. 2001;25:1086–90.
8. Suster S, Rosai J. Cystic thymomas: a clinicopathologic study of ten cases. *Cancer Philadelphia*. 1992;69:92–7.
9. Otsuka H. The utility of FDG-PET in the diagnosis of thymic epithelial tumors. *J Med Invest*. 2012;59:225–34.
10. Peacey SR, Belchetz PE. Graves' disease: associated ocular myasthenia gravis and a thymic cyst. *JRSM*. 1993;83:297–8.
11. Raica M, Cimpean AM, Ribatti D. Myasthenia gravis and the thymus gland. A historical review. *Clin Exp Med*. 2008;8:61–4.
12. Fongi EG, Gotlieb D, Vaamonde CA, Buzzi A, Machado E, Perianes I. Myasthenia gravis following excision of a thymic cyst; study of electrolytes during a myasthenic attack. *Prensa Med Argent*. 1957;27:3754–60.
13. Okumura S, Ohta T, Fujioka M, Nakabayashi H. A case of multilocular thymic cyst with myasthenia gravis. *Nihon Kyobu Geka Gakkai Zasshi*. 1995;43:917–21.

14. Mishra AK, Agarwal SK, Pradhan S, Agarwal A. Association of unilocular thymic cyst and myasthenia gravis. *Neurol India*. 2012;60:103–5.
15. Berrih-Aknin S, Le Panse R. Myasthenia gravis: a comprehensive review of immune dysregulation and etiological mechanisms. *J Autoimmun*. 2014;52:90–100.
16. Cea G, Benatar M, Verdugo RJ, Salinas RA. Thymectomy for non-thymomatous myasthenia gravis. *Cochrane Database Syst Rev*. 2013;10:CD008111, <http://dx.doi.org/10.1002/14651858.CD008111.pub2>.
17. Thymectomy Trial in Non-Thymomatous Myasthenia Gravis Patients Receiving Prednisone Therapy. *Clinical Trials gov Identifier: NCT00294658*. Available from: <https://clinicaltrials.gov/ct2/show/NCT00294658> [accessed 10.9.15].

N. Morollón\*, P. Guerrero, J. Duarte

*Sección Neurología, Complejo Asistencial de Segovia, Segovia, Spain*

\*Corresponding author.

*E-mail address: noemimorollon@gmail.com* (N. Morollón). 2173-5808/

© 2016 Sociedad Española de Neurología. Published by Elsevier España, S.L.U. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## Spontaneous acute epidural haematoma of the cervical spine with an atypical onset resembling ictal symptom<sup>☆</sup>



### Hematoma epidural agudo cervical espontáneo de inicio atípico simulando cuadro ictal

*Dear Editor,*

Spontaneous acute epidural haematoma (SAEH), described by Jackson in 1869, is an infrequent condition with an estimated annual incidence of one case per million population.<sup>1</sup> It accounts for less than 1% of all spinal space-occupying lesions, with peak incidence between the sixth and seventh decades of life.<sup>1–3</sup> SAEH may cause rapid, irreversible neurological impairment and is associated with a mortality rate of 6% to 8%.<sup>3</sup> These lesions are preferentially located in the cervicothoracic region, mainly in the posterior portion of the spinal canal.<sup>1</sup> Cases where no underlying cause or predisposing factor for bleeding (blood dyscrasia, anticoagulant or antiplatelet treatment, tumours, pregnancy, or trauma) can be identified are considered spontaneous; this occurs in 40% to 50% of patients.<sup>2</sup> Most authors suggest a haemorrhagic venous aetiopathogenic mechanism, due to the rupture of valveless epidural venous plexuses in response to a sudden pressure increase at the thoracic or abdominal level.<sup>4</sup> However, other authors suggest bleeding is arterial in origin,<sup>5</sup> secondary to the rupture of epidural radicular arteries, based on the rapid clinical deterioration typically associated with SAEH.<sup>6</sup>

Our patient is a 79-year-old man with no relevant history who presented symptoms of intense headache and neck pain and sudden-onset left hemiparesis of 3 hours' progression.

In view of these symptoms, we activated the code stroke protocol and the patient was transferred to his reference hospital. Motor examination revealed paresis of the left arm (3/5) and left leg paralysis associated with contralateral hypaesthesia without sensory extinction, left extensor plantar reflex, and right flexor plantar reflex. An emergency brain CT scan did not show signs of intra- or extra-axial bleeding or acute brain ischaemia. Suspecting spinal involvement, we performed an emergency cervical CT scan, which showed a hyperdense epidural cervical collection located in the left posterolateral region from C3 to C7 (Fig. 1). After being diagnosed with cervical SAEH, the patient underwent an emergency C3 to C7 posterior cervical laminectomy with microsurgery; the epidural cervical collection was completely evacuated. No underlying vascular anomalies at the C3 to C7 level were identified during the procedure; the anatomical pathology study ruled out the presence of such anomalies in the surgical specimen. A cervical MRI scan performed after the surgery (Fig. 2) revealed the presence of typical postsurgical changes, absence of signs of cervical myelopathy, and small remnants of the epidural collection. The postsurgical study ruled out the presence of any clotting disorders which may have triggered the symptoms. After surgery, the patient displayed good clinical progress, recovering mobility of the left side of the body, with no residual motor or sensory deficits after completing rehabilitation.

SAEH clinically manifests as acute, local, lancinating pain accompanied by motor, sensory, and/or autonomic neurological deficits, normally symmetrical, developing over a period of minutes or days.<sup>7</sup> Unilateral symptoms are infrequent and may lead to misdiagnosis, and diagnostic and therapeutic delay. In these cases, a differential diagnosis of cerebrovascular stroke is essential,<sup>8</sup> given the catastrophic effect of treatment with fibrinolytic agents. Acute neck pain is probably the only baseline clinical finding that may lead us to suspect spinal involvement. The distinctive feature of the case described is that the patient's condition was initially interpreted as a supratentorial ischaemic stroke.

In MRI studies, SAEH in the acute or hyperacute stage appears isointense on T1-weighted sequences and hyperintense on T2-weighted sequences.<sup>9</sup> Although MRI is the diagnostic method of choice, spine CT is a widely available

<sup>☆</sup> Please cite this article as: Arévalo A, Navas M, Pulido P, García de Sola R. Hematoma epidural agudo cervical espontáneo de inicio atípico simulando cuadro ictal. *Neurología*. 2018;33:407–410.