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Scientific Letters

Undiagnosed Myotonic Dystrophy Type 1 in a Patient With Synchronous Thymoma and Thyroid Cancer☆

Timoma y carcinoma tiroideo sincrónicos en paciente con distrofia miotónica de Steinert no conocida

Dear Editor:

Steinert's disease or myotonic dystrophy type 1 (MD1) is a multisystemic autosomal-dominant inherited neuromuscular disorder. Some authors have suggested that it is associated with a greater risk of cancer.[1–4] We report the case of a patient with undiagnosed MD1 who underwent surgery for 2 synchronous tumors.

The patient was a 73-year-old woman, asymptomatic, followed up for multinodular goiter diagnosed of a follicular carcinoma after biopsy. During staging, a mediastinal mass measuring 3.6 cm was revealed (Fig. 1), radiologically consistent with metastasis. The patient had a history of bilateral cataract surgery and basal cell carcinoma. As biopsy of the mediastinal lesion proved impossible, we decided to resect it and to perform a total thyroidectomy. Thus, the patient underwent cervicotomy and partial sternotomy, with successful resection of both lesions. After surgery, extubation was delayed for 48 h, due to ventilatory problems. On day 5 post-surgery, the patient developed respiratory failure due to bronchoaspiration requiring orotracheal intubation, which led to right basal pneumonia. After this was resolved, the patient developed generalized hypoventilation, attributed to muscular weakness which prevented withdrawal of mechanical ventilation.

The pathology report identified a papillary thyroid carcinoma and a mixed B2/B3 thymoma. Myasthenia gravis was suspected, so corticosteroids and anticholinesterase were initiated, and anti-acetylcholine antibody levels and edrophonium tests were requested. However, the patient did not respond to treatment and the studies were negative.

Electromyography revealed a myotonic myopathic pattern typical of MD1, consistent with the patient's clinical characteristics: baldness, drooping eyelids, early cataracts, muscle weakness, and problems swallowing. The diagnosis was confirmed by the results of the genetic study.

In view of the patient's neuromuscular disease, a gastrostomy tube was placed and a tracheostomy was performed to facilitate intermittent disconnection of mechanical ventilation and aspiration of secretions, with a view to discharge with home respiratory support. Despite these measures, the patient died 1 month after surgery due to respiratory failure associated with difficulties in clearing secretions.

MD1 is characterized by progressive weakness and atrophy of the distal and facial muscles, and is associated with repeated failure of the muscles to relax (myotonias). The subclinical forms of this disease may go unnoticed and diagnosis can be late, triggered in many cases by the appearance of an early cataract[5] or after undergoing general anesthesia.

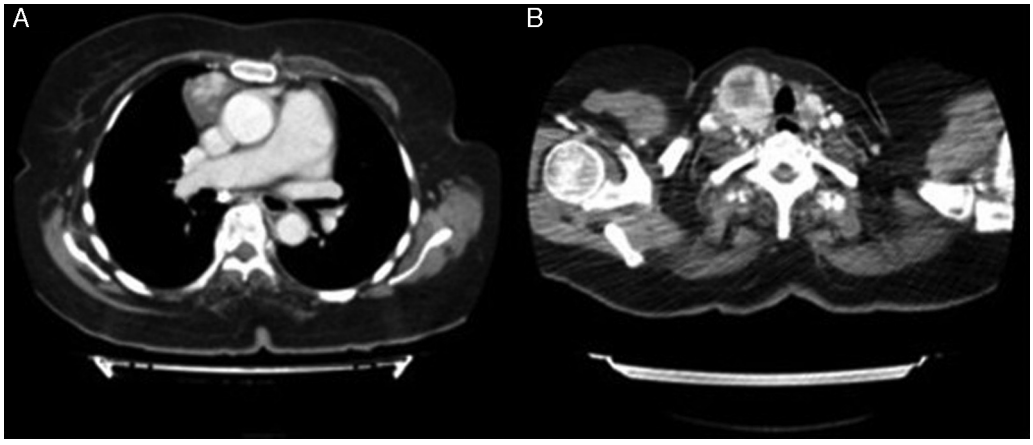

Fig. 1. Computed axial tomography showing a hypervascular mediastinal mass measuring 3.6 cm × 2.7 cm located in the right paracardiac region (A), and multinodular goiter invading the right thyroid lobe (B).

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Several clinical cases of tumors in MD1 patient have been reported. The most commonly described cancers are pilomatrixomas, although they can vary widely. To date, only 3 studies have attempted to clarify this possible association. One of these, based on 1658 patients with MD (types 1 and 2) concluded that these patients had a higher risk of endometrial, ovarian, brain, and colon cancer,[1] and that the risk was higher in women and patients with MD1.[2] Two earlier studies detected an increased risk of thyroid cancer and choroidal melanoma,[3] as well as thymoma, gynecological and lung cancers.[4]

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M. Teresa Gómez Hernández,[a,*] M. Teresa Martín Posadas,[b] M. del Carmen Gónzalez Sánchez[c]

[a] *Departamento de Cirugía Torácica, Hospital Universitario de Salamanca, Salamanca, Spain*
[b] *Unidad de Cuidados Intensivos, Hospital Universitario de Salamanca, Salamanca, Spain*
[c] *Departamento de Cirugía General, Hospital Universitario de Salamanca, Salamanca, Spain*

* Corresponding author.
E-mail address: mteresa.gomez.hernandez@gmail.com (M.T. Gómez Hernández).

Video-Assisted Thoracoscopy Surgery in the Diagnosis and Treatment of Impaled Knives in the Chest☆

Videotoracoscopia para el diagnóstico y tratamiento de los empalamientos por cuchillo en el tórax

To the Editor:

Foreign bodies impaled in the chest are rare and generate a dramatic situation for the patient, the relatives and the trauma team. The standard approach in these cases has always been open thoractomy.[1] Video-assisted thoracoscopy surgery (VATS) is a relatively recent indication in the management of impaled foreign bodies.

We report 2 cases where VATS was used in the management of chest impalement with knives.

In the first case, a 62-year-old man was stabbed in the left thorax with a kitchen knife. The knife entered the thorax at the 4th intercostal space at the level of the anterior axillary line (Fig. 1A). The patient was hemodynamically stable. Chest radiograph showed left hemothorax and the knife could be visualized above the cardiac silhouette (Fig. 1B). He was transferred to the operating room and examined by thoracoscopy, which showed that the knife had penetrated the upper lobe of the lung. The hemothorax was aspirated and the knife was extracted under direct vision. The pulmonary laceration was sutured by thoracoscopically. In the second case, a young man aged 18 years presented with a serrated knife impaled at the level of the 9th thoracic vertebra. His vital signs on arrival at hospital were: blood pressure: 129/57 mmHg, heart rate: 65 bpm, and breathing rate: 20 breaths/min. Neurological examination showed a complete medullary lesion below the wound. No hemothorax or pneumothorax were seen on chest radiograph (Fig. 1C) and computed tomography of the spine showed that the knife had sectioned the spinal cord (Fig. 1D). Orotracheal intubation was achieved by positioning the patient between 2 gurneys in order to place him in a supine position. Thoracoscopic examination revealed a posterior mediastinal hematoma, so the procedure had to be switched to thoracotomy. An injury was revealed in the thoracic aorta that could be repaired with polypropelene suturing. In both cases, the post-operative period was incident-free.

VATS in chest injury was initially indicated for diagnostic purposes and for management of coagulated hemothorax. Abolhoda et al.[2] used it to rule out perforated diaphragm and to successfully treat several cases of post-traumatic retained hemothorax. Lang-Lazdunski et al.[3] extended the indications of thoracoscopy in chest trauma to include persistent hemothorax, intrathoracic foreign body, post-traumatic empyema, and post-traumatic chylothorax. As surgeons gain more experience and technology improves, the treatment of more complex injuries has become possible. Few reports are available on VATS management of foreign bodies impaled in the chest, but we agree with Isenburg et al.[4] in that these patients must be hemodynamically stable before a thoracoscopic approach can be attempted; otherwise, open thoracotomy must be initiated directly.

In brief, the thoracoscopic management of foreign bodies impaled in the chest is a safe and effective option for the hemodynamically stable patient. VATS has specific advantages. It is a minimally invasive diagnostic and therapeutic procedure. In addition to establishing the severity of the lesions, the surgeon can employ it to identify potential complications for removal of the foreign body and to repair the different wounds.

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