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CLINICAL CASE

Thymoma in childhood. A case report and review of literature☆

Ana Lilia Nolasco-de la Rosa*, Roberto Mosiñoz-Montes, Luis Alberto Nuñez-Trenado, Edgardo Román-Guzmán, Christian Ezequiel Chávez-Villicaña, Guillermo Naranjo-Hernández

Servicio de Cirugía, Centro Médico Ecatepec, Instituto de Seguridad Social del Estado de México (ISSEMyM), Ecatepec, Estado de México, Mexico

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KEYWORDS
Thymoma;
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Abstract
Background: Mediastinal tumours in children are rare. Around 25% of them can be malignant. Thymoma is an uncommon neoplasm, and during adulthood it corresponds to 30% of anterior mediastinum tumours. The peak incidence is between 55 and 65 years.
Clinical case: A case of lymphocytic thymoma case is reported in a 4-year-old patient with no previous or associated symptomatology. There was only a volume increase on the anterior neck region. The neck radiography and neck and chest tomography confirmed an anterior mediastinal mass surrounding the aorta and vena cava, as well as multiple mediastinal lymph nodes.
Conclusions: Early diagnosis and complete resection are the basis for management and prognosis.

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* Corresponding author at: Edificio 14 entrada C departamento 102 Col. Unidad Lindavista Vallejo, Entre poniente 152 y Norte 35, Gustavo A Madero, C.P. 07720, D.F., Mexico. Tel.: +52 55 5567 3524.
E-mail address: anis3791@hotmail.com (A.L. Nolasco-de la Rosa).

PALABRAS CLAVE
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Timoma en edad pediátrica. Reporte de un caso y revisión de la bibliografía

Resumen
Antecedentes: Los tumores de mediastino en niños son poco frecuentes, el 25% de ellos corresponde a tumores malignos. El timoma es una neoplasia poco común, en adultos corresponde al 30% de los tumores de mediastino anterior y con un pico de incidencia es entre los 55-65 años.
Caso clínico: Se reporta el caso de un timoma linfocítico en una paciente de 4 años, sin sintomatología previa ni asociada, solo aumento de volumen en región anterior de cuello. Las radiografías y tomografías de cuello y tórax confirman una masa mediastinal anterior, que rodea a la aorta y la cava, así como múltiples adenomegalias mediastinales.
Conclusiones: El diagnóstico temprano y la resección completa son la base del tratamiento y del pronóstico.

Background

Mediastinal tumours in children are rare. They represent 3% of total surgical chest interventions in children. 45–50% correspond to primary tumours; of these 40–45% are malignant tumours and a third of them present in children under 2 years of age.[1–3] The main types of primary tumours are: thymic, neurogenic, lymphatic, or of germinal or mesenchymal cells; secondary tumours are due to lymphatic dissemination of intrathoracic organs towards the mediastinum.[2] The main location is the posterior mediastinum and the majority are of neurogenic and benign origin.[1]

Since the majority of these tumours are asymptomatic, most are diagnosed as radiographic findings[1] and on occasion present as fever, weight loss, general malaise, severe respiratory failure with dyspnoea, intercostal retractions, atelectasis or more aggressively as superior vena cava syndrome, dysphagia, dysphonia, laryngeal spasm caused by recurrent laryngeal nerve compromise, diaphragmatic paralysis caused by phrenic nerve involvement or Horner syndrome caused by lymph node and sympathetic nerve compromise.[1,2] Once the tumour has been located, clinical study protocol includes: blood biometry, comprehensive metabolic panel, skin and soft tissue infection panel, and tumour markers. A lateral projection chest X-ray is required. The gold standard is simple and contrast CAT. On occasions magnetic resonance imaging, iodine-123 nuclear medicine imaging, iodine-131 testing for ectopic thyroid tissues, technetium-99 testing to examine for gastric mucosa in intestinal duplications or the use of metaiodobenzylguanidine for diagnosing neuroblastoma are carried out. In children the use of cervical and chest ultrasound is usually useful, or even fine needle biopsy. The latter may, however, be limiting for diagnosis and there may be a risk of bleeding.[2,4,5] Standard treatment consists of surgery unless tumours are germinal or lymphatic. A cervical and sternal approach is used, with the performance of mediastinoscopy or video-assisted thoracoscopy.[4,5]

The thymus is a central lymphoid organ, corresponding to the anterior mediastinum, where the stem cells of the bony medulla are divided into mature T lymphocytes, with a tendency to degenerate at around 2 years of age. When this process continues they may grow or become malignant and are functional as both. Hyperplasia (70%) and thymomas (15%) with myasthenia gravis may occur, as the T lymphocytes are intolerant to anti-acetylcholine antibodies.[6] The thymus may be a location for lung, breast or thyroid cancer metastasis. Thymoma is a low frequency tumour of uncertain patterns of behaviour. It corresponds to 26–50% of mediastinal tumours and to 82% of thymus tumours[4,6,7]; diagnosis is generally incidental and occurs through chest X-ray and a CAT scan to determine the location and extension of the tumour.[5,6] With regards to thymoma classification, although no standard classification exists, the one proposed by Masaoka et al.[3] is the most commonly used. This classification is divided into 4 categories in accordance with the stage of tumour infiltration into: *I, capsule intact; II, invasion of the capsule; III, macroscopic invasion into neighbouring organs; IVa and IVb, pleural or pericardial dissemination and lymphogenous or haematogenous metastasis to distant sites respectively.* The WHO classification is: *epitelial A, mixed AB, lymph node B1, spindle-shaped B2 and thymic carcinoma.*[4,6,7] Thymic carcinoma is the neoplasm with the highest malignancy of the thymus. Symptoms tend to present earlier and are aggressive with pain, weight loss, superior vena cava syndrome, dyspnoea caused by pericardial effusion and compression of the airway. It is rare and presentation is atypical of its cells which are not similar in appearance to the original thymus ones. It represents around 5% of mediastinal tumours in adults and accounts for approximately 1–2% of mediastinal tumours in children.[1,2,4,7–9] The treatment of choice for thymus tumour is surgical resection.[4–6,9,10] which may consist of sternotomy, upper cervical sternotomy, thoracotomy, video-assisted thoracoscopy, mediastinoscopy and ideally total surgical resection. If the latter is not performed treatment is complemented with radiotherapy and/or chemotherapy depending on the tumour type, invasion and extension.[4,6–10] Surgical complications may be pneumonia, haemothorax, pneumothorax, sternal dehiscence, mediastinitis, osteomyelitis[10] and less frequently