

Type A thymoma invading the trachea: a case report

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Abstract: Thymomas generally grow applying pressure to the adjacent organs but they may infiltrate the capsule and extend directly to them. Invasion of the airway is extremely rare and generally related to high-grade thymomas. Few cases of thymoma with endobronchial polypoid growth and only one patient with tracheal invasion have been reported. We present a typical type A thymoma with endoluminal growth almost completely obstructing the trachea. A 93-year-old man was referred to our hospital with severe dyspnea and wheezing. He already presented a diagnosis of type A thymoma, discovered occasionally 8-year before and followed up with a yearly computed tomography (CT) scan. The CT scan of the thorax at admission, revealed a bulky anterior mediastinal tumor with an endotracheal growth, almost obstructing the airway. The fiberoptic bronchoscopy confirmed severe tracheal stenosis caused by a vegetating endoluminal tumor. Due to the severity of symptoms, we performed an emergency rigid bronchoscopy aimed to relieve the upper airway. The endotracheal tumor had a relatively small base of implant and was totally removed with mechanical resection. Pathological examination revealed a proliferation of oval or spindle neoplastic cells with bland nuclei and inconspicuous nucleoli and very few immature lymphocytes. Tracheal invasion by type A thymoma was finally diagnosed. No clinical evidence of endotracheal recurrence was recorded after a 15-month follow-up.

Keywords: Thymoma; airway invasion; rigid bronchoscopy; case report

Received: 24 September 2020; Accepted: 20 April 2021; Published: 25 December 2021. doi: 10.21037/med-20-63 **View this article at:** http://dx.doi.org/10.21037/med-20-63

Introduction

Thymomas are the most common primary tumors of the anterior mediastinum (1). Generally they are indolent tumors but occasionally may show malignant behavior, by invading adjacent organs (2). According to the WHO classification thymomas are divided in five subtypes (A, AB, B1, B2, B3) based on the morphology of epithelial cells and the amount of lymphocytic component (3). Type A thymomas are diagnosed at an early stage in over 90% of cases and are characterized by a 10-year survival rate close to 100% (4).

Few cases of thymoma with endobronchial growth have been described and the only reported patient with tracheal invasion had a B3 thymoma (5). We present a typical type A thymoma with polypoid endotracheal growth, almost completely obstructing the airway.

We present the following article in accordance with the CARE reporting checklist (available at http://dx.doi. org/10.21037/med-20-63).

Case presentation

In April 2019 a 93-year-old man was referred to our hospital with severe dyspnea and wheezing from at least 5 weeks. Comorbidities: previous *Legionella* pneumonia;

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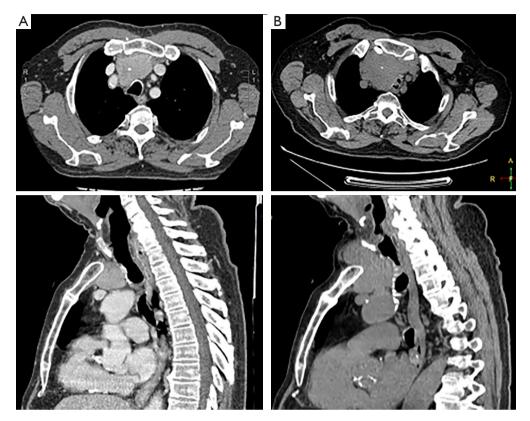


Figure 1 Radiological images of type A thymoma. (A) CT scan performed in 2011 demonstrated a well encapsulated round mass measuring 55×58×44 mm with a calcification in the anterior-superior mediastinum; (B) CT scan performed in 2019 showed the thymic tumor invading the tracheal wall and growing inside the tracheal lumen. CT, computed tomography.

several episodes of colonic diverticulitis; benign prostatic hyperplasia. The patient was non-smoker, with an 8-year history of type A thymoma, discovered occasionally, and diagnosed by means of percutaneous core-needle biopsy. At the time of diagnosis, the computed tomography (CT) scan revealed a well encapsulated round mass of the anterior mediastinum, measuring $55 \times 58 \times 44$ mm with focal calcifications (*Figure 1A*). As the patient refused surgery and radiation therapy, a follow-up with a yearly CT scan was scheduled, showing slow-growing disease until the last exam of February 2018, which revealed tracheal compression without any sign of invasion.

At the physical examination inspiratory stridor and tachypnea were recorded. Blood exams were in the normal range. The CT scan of the thorax revealed a bulky anterior mediastinal tumor with an endotracheal polypoid growth, almost obstructing the airway (*Figure 1B*). At the fiberoptic bronchoscopy we observed a severe tracheal stenosis caused by a vegetating endoluminal

tumor with a mild extrinsic compression (Figure 2). An emergency rigid bronchoscopy, aimed to relieve the upper airway obstruction, was performed. Because of the critical airway obstruction, in order to facilitate the intraoperative conduct, we used an original alternative method for airway management with combined tracheal intubation and rigid bronchoscope (6). The endotracheal tumor had a relatively small base of implant and was totally removed with mechanical resection, using an 8.5 mm Storz ventilating rigid bronchoscope. Tissue fragments were entirely composed by a proliferation of oval or spindle neoplastic cells with bland nuclei and inconspicuous nucleoli, arranged in a fascicular pattern. Very few immature lymphocytes were present throughout the tumor. Immunohistochemical analysis showed a positive expression in neoplastic cells for pankeratin, paired-box gene 8 (PAX8), cytokeratin 19, cytokeratin 7, p63 and p40 and negativity for CD5, CD117 and CD20. Immature lymphocytes resulted



Figure 2 Bronchoscopic examination revealing a pink polypoid tumor in the mid-trachea causing a severe tracheal stenosis with 20% residual lumen.

positive for terminal deoxynucleotidyl transferase (TdT) (*Figure 3*).

Postoperative course was uneventful and the patient was discharged on postoperative day 2, without respiratory symptoms. The fiberoptic bronchoscopy, carried out 3 months after surgery, was negative for endotracheal recurrence. Due to his old age, the patient refused to perform any follow-up radiologic exam. Thus, follow-up was based on clinical evaluation and the patient is alive, without any respiratory symptom, 15 months after rigid bronchoscopy. *Figure 4* displays the timeline for clinical presentation, diagnosis of thymoma invading the trachea, surgery and follow-up.

Written informed consent was obtained from the patient. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and national research committees and with the Helsinki Declaration (as revised in 2013).

Discussion

Thymomas natural history entails a wide variety of biological behaviors. Most patients are asymptomatic or have a parathymic syndrome; symptoms related to invasion of mediastinal structures are sporadically reported, while clinical emergencies caused by thymomas are extremely rare (7). These tumors generally grow applying pressure to the adjacent organs but they may infiltrate the capsule and extend directly to them. The invasion of the airway is very

extend directly to them. The invasion of the airway is very rare (5). In the English Literature 21 cases of endobronchial growth are reported and most of them involved the left bronchial tree (5,8). Prior to the herein reported case, only one patient with endotracheal polypoid growth by an invasive B3 thymoma had been described (5).

Contrast CT of the thorax is the gold standard for the diagnosis of mediastinal masses and bronchoscopy is indicated only when the involvement of the airway is suspected (1). In previous reports, biopsies through a flexible bronchoscope have been attempted, resulting non-diagnostic in most cases (5,8). In the reported case, flexible bronchoscopy was performed in order to define the involvement of the tracheal wall and the residual respiratory space but, since the patient was highly symptomatic and the tracheal obstruction was severe, no biopsies were taken during such procedure. The emergency rigid bronchoscopy allowed to achieve both histology and palliation. Radical resection, stage and histotype are strong prognostic factors for thymoma patients (9). Surgery is the mainstay of treatment for most thymomas, while irradiation and chemotherapy are commonly administered to not-resectable tumors or in adjuvant regimens (1). Furthermore, surgery, as part of multimodality therapy, could prolong survival also for advanced stage thymomas (10). Our patient had been diagnosed with resectable type A thymoma, 8 years earlier, but he refused surgery; because of the low effectiveness of irradiation and chemotherapy on lowgrade thymomas, a strict follow-up had been established. The tumor remained substantially stable for long time and eventually it invaded the tracheal wall growing inside the lumen. The slow growth of the tumor and the small implant base of the endotracheal growth have made the endoscopic palliative treatment performed effective and lasting, so far avoiding the use of stents or laser treatments.

Although type A thymomas are generally considered slow-growing tumors they cannot be defined as benign; some studies indeed have demonstrated invasive growth and distant metastases (4). Furthermore, a new entity of type A thymoma with atypical morphological features invading adjacent organs or spreading outside the chest has been recognized by the WHO classification (3). However, our patient was affected by a type A thymoma without any atypical feature, since no necrosis, atypia and mitotic activity were observed.

In conclusion, conservative treatment of typical type A

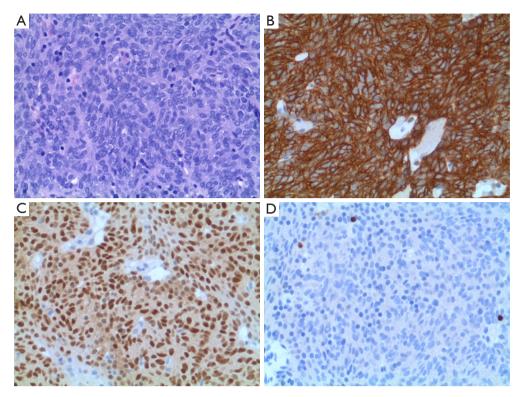


Figure 3 Pathological images of type A thymoma. (A) Tissue fragments were composed by spindle and oval neoplastic cells with bland nuclei and no mitotic activity (hematoxylin-eosin, 400×). Immunohistochemical analysis showed (B) strong expression for pankeratin (400×) and (C) PAX8 (400×). (D) Few immature lymphocytes resulted positive for TdT (400×). PAX8, paired-box gene 8; TdT, terminal deoxynucleotidyl transferase.

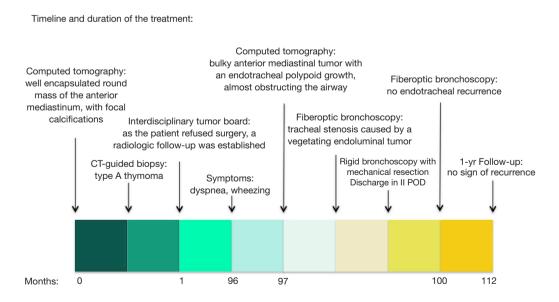


Figure 4 Timeline for clinical presentation, diagnosis of thymoma invading the trachea, surgery and follow-up.

Mediastinum, 2021

thymoma in very elderly patients may be acceptable but careful follow-up, aimed to rule out a possible invasive growth, should be carried out.

Acknowledgments

Funding: None.

Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at http://dx.doi.org/10.21037/med-20-63

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at http://dx.doi. org/10.21037/med-20-63). The authors have no conflicts of interest to declare.

Ethical statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. Written informed consent was obtained from the patient. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and national research committees and with the Helsinki Declaration (as revised in 2013).

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doi: 10.21037/med-20-63

Cite this article as: Tassi V, Puma F, Bellezza G, Ceccarelli S. Type A thymoma invading the trachea: a case report. Mediastinum 2021;5:36.

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