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

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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;
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 55×58×44 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 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.
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 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 low-grade 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, 400x). Immunohistochemical analysis showed (B) strong expression for pankeratin (400x) and (C) PAX8 (400x). (D) Few immature lymphocytes resulted positive for TdT (400x). PAX8, paired-box gene 8; TdT, terminal deoxynucleotidyl transferase.

Timeline and duration of the treatment:

Computed tomography: well encapsulated round mass of the anterior mediastinum, with focal calcifications
CT-guided biopsy: type A thymoma
Interdisciplinary tumor board: as the patient refused surgery, a radiologic follow-up was established
Symptoms: dyspnea, wheezing
Fiberoptic bronchoscopy: tracheal stenosis caused by a vegetating endoluminal tumor
Rigid bronchoscopy with mechanical resection
Discharge in II POD
1-yr Follow-up: no sign of recurrence

Figure 4 Timeline for clinical presentation, diagnosis of thymoma invading the trachea, surgery and follow-up.
thymoma in very elderly patients may be acceptable but careful follow-up, aimed to rule out a possible invasive growth, should be carried out.

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Footnote

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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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