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Case report

Granular cell tumor of the trachea mimicking an infiltrating thyroid cancer. A case report

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ABSTRACT

Introduction and importance: Granular cell tumor (GCT) is a rare neurogenic neoplasm originating from Schwann cells that predominantly affects women and can involve skin and mucousae. In the respiratory system it most frequently involves bronchi and larynx, while it is rare in the trachea.
Case presentation: A 26-year old female smoker was hospitalized for a suspected hypoechoic nodule in the right thyroid lobe closely adherent to the trachea. At preoperative computed tomography tracheal lumen was totally clear. The patient underwent a total thyroidectomy with lymph node dissection and tracheal shaving. The postoperative course was complicated by an extensive subcutaneous bilatreral emphysema associated with respiratory distress appeared on the fifth day.
Bronchoscopy revealed a right anterolateral subcentimeter lesion near the second tracheal ring. Histologically, the diagnosis was consistent with a tracheal GCT developing into the thyroid parenchyma. The patient was discharged on the twentieth postoperative day. At the follow-up bronchoscopy the lesion was completely healed and at the last 12 month follow-up the patient is doing well.
Clinical discussion: Tracheal tumors are uncommon neoplasms accounting for about 2% of the total respiratory tree tumors. In literature we found <50 papers concerning tracheal GCT and in almost all of the cases patients complained about respiratory symptoms.
Conclusion: We report here a rare case of benign GCT of the trachea with extraluminal development, in a young patient who did not complain about preoperative respiratory symptoms, presented on ultrasound as a thyroid nodule with suspected cytology.

1. Introduction

Granular cell tumor (GCT) is an infrequent neurogenic neoplasm originating from Schwann cells and described for the first time by Abrikossoff (in 1926) as "*granular cell myoblastoma*" [1,2].

GCT arises in the skin and mucosae, localizing itself in the cutaneous-subcutaneous tissues and in various locations of the digestive (esophagus), respiratory (bronchi and larynx) and genital (vulva and breast) systems. It is rare in the trachea. It mainly affects females, can be solitary or multiple, and usually exhibits benign biological behavior although some rare cases of malignancy have been described [3].

The histological characteristics (i.e. perineural and vascular involvement, growth pattern, nuclear pleomorphism, areas of necrosis, spindle cells with high mitotic index, pustule-ovoid bodies of Milian and positivity for S100 and CD68 on immunostaining) allow a classification according to the criterion of Fanburg-Smith, that distinguishes them as benign, atypical and malignant tumors, and the more recent one by Nasser, who subclassifies them only as benign and with uncertain malignant potential ones [4,5].

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2. Case presentation

A 26-year old woman was admitted to the Endocrine Surgery Unit of the University Hospital of Messina with a nodule in the upper third of the right lobe of thyroid, which was highly suspicious of malignancy. This hypoechoic nodule, 19 mm in maximum diameter, appeared to be indissociable on ultrasounds from the trachea and cricoid (Fig. 1), it had irregular margins and no vascular signals on the Color-Doppler. US showed no lymphadenomegaly and the trachea appeared aligned. On cytological examination, the report indicated a poorly differentiated carcinoma of the thyroid (category IV, i.e. suspicious for follicular neoplasm, according to Bethesda System for Reporting Thyroid Cytopathology). To better define the suspected involvement of the respiratory tract, a contrast-enhanced computed tomography (CT) scan was performed; it confirmed the presence of hypodense formation of 9 × 10 × 20 mm in the right lobe, characterized by intense and inhomogeneous contrast enhancement, compressing and displacing the trachea to the left, with a remodeled area of the right lateral tracheal wall confirming the infiltration (Fig. 2). No evidence of gross protrusions of heteroplastic tissue in the aerial lumen was detected. Her hormonal panel values (free triiodothyronine, free thyroxine, thyrotropin, thyroglobulin), including also calcitonin and parathormone levels, were within the limit. Preoperative routine laboratory tests, electrocardiogram, chest radiography, and flexible rhino-laryngoscopy were all normal. She did not have any medical comorbidities and had an allergy to penicillins. She smoked 20 cigarettes a day. The patient underwent total thyroidectomy with central compartment lymph node dissection (level VI-VII) and resection of the right strap muscles.

Intraoperatively, the nodule was hard in consistency and firmly adhered to the right lateral tracheal plane for about 1 cm, (approximately to the first 2 tracheal rings); therefore in consideration of macroscopic infiltration an antero-lateral right tracheal shaving, using a 15-blade scalpel, was also carried out, with subsequent verification of the tracheal integrity by means of a hydro-pneumatic test, with no evidence of injury. Nevertheless, a right lateral tracheal wall reinforcement was performed was performed using a muscular flap (right sternocleidomastoid muscle) and then fibrin glue was applied. After the surgical procedure, the patient was transferred to an intensive care unit for the realization of a protected awakening over she remained for 48 h. The patient was on intravenous calcium gluconate and calcitriol for severe hypocalcemia arising on the 2nd postoperative day; subsequently calcium gluconate was gradually replaced with oral calcium carbonate.

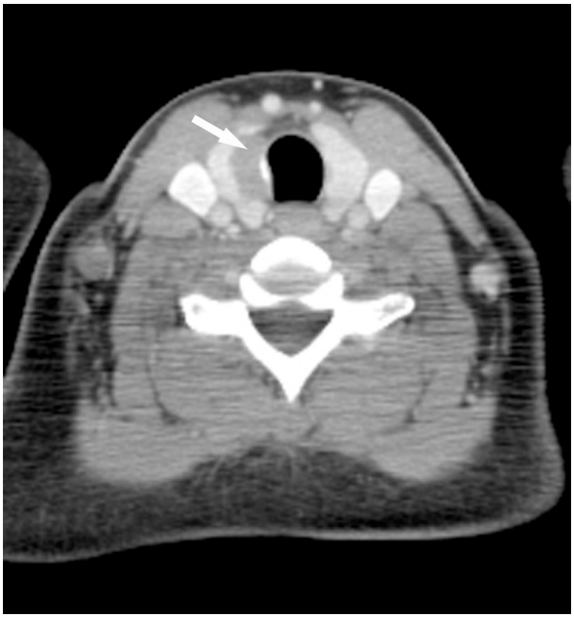

Fig. 2. Preoperative enhanced CT scan showing a remodeled area of the right lateral tracheal wall confirming the infiltration (white arrow).

On the 5th postoperative day, the patient had sudden breathing difficulty after a bout of coughing. The clinical examination revealed a noticeable subcutaneous laterocervical and supraclavicular emphysema, mainly on the left side, and respiratory distress was relieved by the partial reopening of the surgical wound that allowed the escape of an abundant an amount of air. The extensive bilateral subcutaneous emphysema with associated pneumomediastinum was confimed by an urgent CT scan (Fig. 3).

Bronchoscopy revealed a right antero-lateral subcentimeter lesion bulging beyond the edge of the tracheal wall (Video 1), approximately at the level of the second tracheal ring. Therefore, parenteral nutrition therapy associated with intravenous antibiotics (levofloxacin 500 mg) for 5 days was administered as well. Her further hospital stay was uneventful and she was discharged on the 20th postoperative day.

Histologically, the diagnosis was consistent with a tracheal wall GCT developing in the thyroid parenchyma, with Mib-1 < 2% and intense

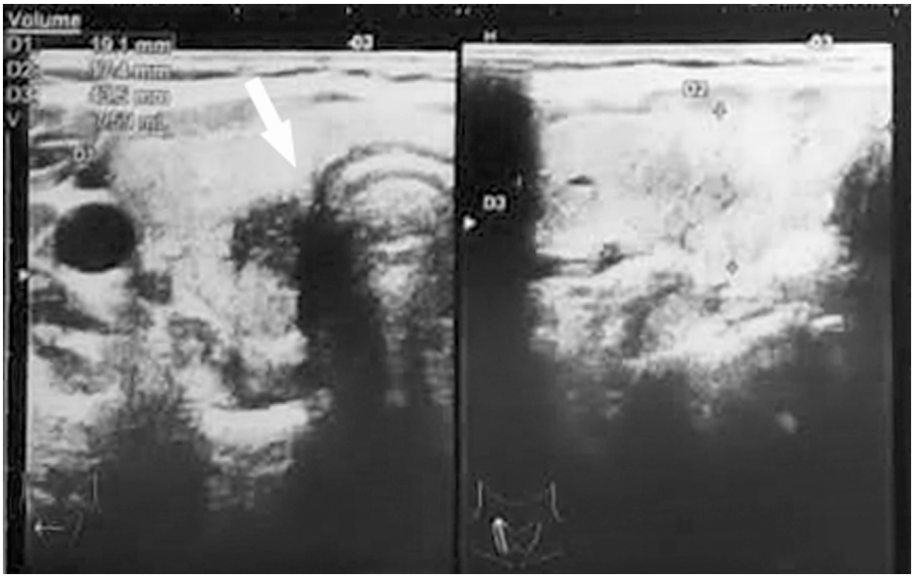

Fig. 1. Ultrasound findings: hypoechoic 19 mm-nodule that seems indissociable from the trachea and cricoid (white arrow).

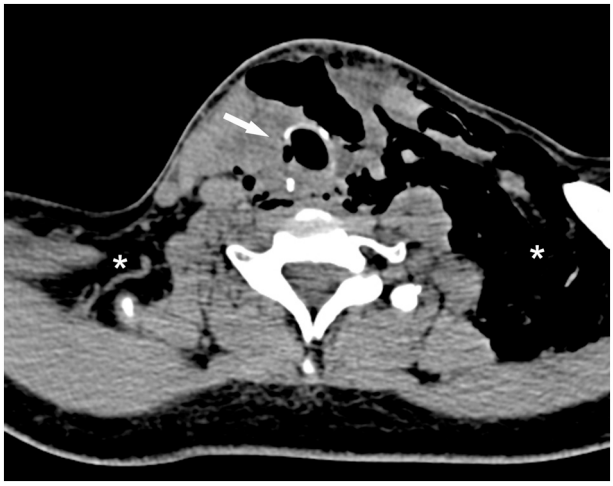

Fig. 3. Postoperative enhanced CT scan revealed a tracheal injury (white arrow) with a noticeable bilateral subcutaneous laterocervical and supraclavicular emphysema, prevalent on the left side (white asterisks).

S100-CD68 positivity (whereas thyroglobulin and cytokeratin-7 were negative), on immunostaining. In the contralateral lobe an incidental papillary microcarcinoma (maximum diameter of 5 mm), capsulated follicular variant, was also found.

At the 3 month follow-up bronchoscopy, the lesion was completely healed (Video 2), as evidenced at control CT, and at the last 12 month follow-up check, the patient is showed well and is regularly followed by endocrinologists.

3. Discussion

Tracheal tumors, both benign and malignant, are uncommon neoplasms accounting for about 2% of the total of the respiratory tree tumors, whose estimated annual incidence of 0.1/100,000 people per year [3,6].

Even today, GCT remains an unclear clinical-pathological entity for the small number of cases reported in literature and for its clinical heterogeneity. In a recent systematic review, which analyzed the studies published in peer-reviewed journals with a number of cases >5, only 2 tracheal locations out of 42 total cases were reported. Overall, in literature reviews carried out using generically the term "*granular cell tumor*", a total of 1499 cases were obtained from 47 studies; among these the respiratory tree was involved in only 36 cases and bronchial localization was the most frequent (27 cases) [2].

In the English literature <50 papers concern tracheal GCT and in almost all of them the patients complained of respiratory symptoms [7].

In the present case, the extraluminal development of the tracheal neoplasm, deepening into the thyroid tissue, simulated an infiltrating thyroid tumor. This occurrence, to our knowledge, has been described in the worldwide literature in <10 similar cases [8]. In all previous cases, tumors were solitary and patients were treated by total thyroidectomy or lobectomy with associated tracheal shaving [9–11]. Our patient, who showed no respiratory symptoms, underwent surgical procedure on suspicion of neoplastic pathology of the thyroid, supported by enhanced CT and fine needle aspiration results.

4. Conclusions

We report, according to SCAR guidelines [12], herein a rare case of benign, undiagnosed preoperatively, GCT of the trachea mimicking a thyroid nodule with suspected cytology.

Tracheal neoplasm extraluminal development, simulating an infiltrating thyroid tumor, represents a rare occurrence. Furthermore, the absence of preoperative respiratory symptoms adds even more peculiarity to the clinical case.

In conclusion, we suggest paying attention to paratracheal nodules with a cytological result characterized by polygonal or spindle elements with abundant eosinophilic granular cytoplasm and nuclear polymorphism, considering the hypothesis of extraluminal development of tracheal neoplasms thus avoiding clinical misdiagnosis and excessively demolitive surgical procedures.

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Patient consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author's contribution statement

- Conceptualization: G.D.
- Data curation: G.D., F.F., A.P. and V.C.
- Patient management: G.D., F.F., A.P. and V.C.
- Writing - original draft: F.F., A.P. and A.I.
- Writing - review & editing: G.D., F.F. and G.F.
- Supervision: G.D. and G.F.

All authors have read and approved the final manuscript and all materials before submission.

Research registration

Not applicable.

Guarantor

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Declaration of competing interest

All authors declare no conflicts of interest.

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