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ANCA Negative GPA (Wegener's Disease) Years after Thymectomy

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Abstract

A 78-year-old man with a history of resection of a mediastinal mass in 2008, initially diagnosed as a typical carcinoid tumor, presented 17 years later with a progressively enlarging subcutaneous pectoral mass. Given his oncological history, recurrent malignancy was strongly suspected. However, extensive radiological, histopathological, and immunological investigations demonstrated that the lesion represented antineutrophil cytoplasmic antibody (ANCA)-negative granulomatosis with polyangiitis (GPA). ANCA-negative GPA is an uncommon subtype of this autoimmune small-vessel vasculitis and is often characterized by localized disease, predominantly affecting the respiratory tract without renal involvement. This case highlights a rare presentation of ANCA-negative GPA mimicking recurrent thoracic malignancy many years after mediastinal tumor resection. Awareness of such atypical manifestations is essential to avoid misdiagnosis and ensure timely initiation of appropriate immunosuppressive therapy.

Keywords

Chest Wall Tumor, Carcinoid, Thymoma, Granulomatosis with Polyangiitis, M. Wegener

1. Introduction

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare autoimmune antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis that can occur at any age but is most commonly diagnosed in adults between 40 and 65 years of age. The disease primarily affects the respiratory tract and kidneys, although involvement of the skin, eyes, peripheral nerves, and other organs may occur. Diagnosis is based on a combination of clinical findings,

laboratory investigations including ANCA testing, imaging studies, and, when indicated, histopathological examination. Treatment typically involves immunosuppressive therapy aimed at inducing remission, preventing disease progression, and limiting organ damage. The pathogenesis of GPA involves a complex interplay of genetic susceptibility, environmental factors, and immune dysregulation, resulting in necrotizing granulomatous inflammation and vasculitis of small- and medium-sized blood vessels [1]-[3]. Although most patients are ANCA-positive, a subset presents with ANCA-negative disease. Despite the absence of detectable ANCA serology, these patients may exhibit clinical and histopathological features characteristic of GPA. In ANCA-positive GPA, ANCAs are thought to activate neutrophils, leading to endothelial injury and vascular inflammation. In contrast, the pathophysiology of ANCA-negative GPA remains incompletely understood. Alternative mechanisms, including T-cell-mediated granulomatous inflammation and localized immune responses, may play a more prominent role than circulating autoantibodies. Histopathological findings frequently demonstrate necrotizing granulomatous inflammation similar to that observed in ANCA-positive disease, suggesting that ANCA-independent pathways can contribute to disease development and progression. Clinically, ANCA-negative GPA is often associated with more localized disease, particularly involving the upper respiratory tract and lungs, whereas severe renal involvement appears less common than in ANCA-positive patients. Management generally follows established treatment strategies for ANCA-associated vasculitis, including immunosuppressive therapy tailored to disease severity and organ involvement [4]-[6].

We report a unique case of ANCA-negative granulomatosis with polyangiitis occurring many years after thymectomy and manifesting as a chest wall protrusion, an uncommon presentation that broadens the recognized clinical spectrum of this disease. Written informed consent for publication was obtained from the patient. In accordance with local regulations, ethical approval was not required due to the retrospective nature of this case report.

2. Case Presentation

In September 2025, a vital retired male security officer of 78 years, was referred to the pulmonologist with a subcutaneous non-tender thoracic mass at the right sternal border, which had been gradually growing over the past few years. Apart from the cosmetic inconvenience patient had no complaints, no ear, nose or throat issues and his physical examination was otherwise unremarkable (**Figure 1**).

His medical history was notable for a median sternotomy performed in January 2008 for resection of a mediastinal tumor, which was histopathologically diagnosed as a typical carcinoid, a well-differentiated neuroendocrine neoplasm characterized by indolent growth [7]. The patient had never smoked tobacco. Follow-up evaluation in 2012 revealed no evidence of recurrent disease. However, during routine follow-up in 2021, chest radiography demonstrated two consolidative lesions in the right lung. Subsequent computed tomography (CT) of the chest iden-

tified multiple tumors in the right lung, raising suspicion for recurrent carcinoid disease [8] (**Figure 2**, arrows (A), (B) and (C)).

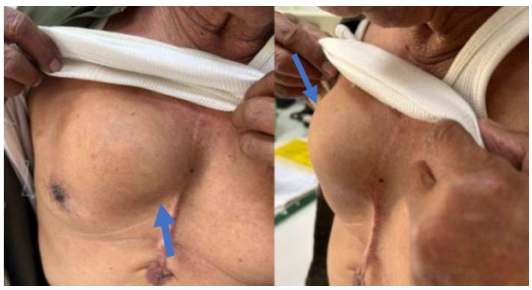

Figure 1. Protruding subcutaneous mass (arrow) on the right pectoral area adjacent to sternotomy scar.

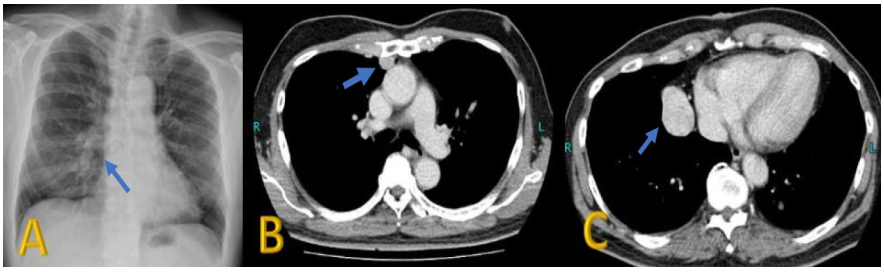

Figure 2. (A): chest x ray showing lesions in the right lung and chest CT scan showing a substernal tumor in (B) and a pulmonary tumor in the right lung (C) (arrows).

Primary lung carcinoma was considered less likely because of the multiplicity of the pulmonary lesions, the absence of pleural or pericardial effusion, and the lack of cervical, axillary, hilar, or mediastinal lymphadenopathy. Although ultrasound-guided transthoracic biopsy of the pulmonary tumor was considered feasible for obtaining a histological diagnosis, a somatostatin receptor imaging study was preferred to support the presumptive diagnosis of recurrent carcinoid tumor and to evaluate for metastatic disease elsewhere in the body [9]. Detection of metastatic disease would have important therapeutic implications, including consideration of treatment with somatostatin analogues such as octreotide [10] [11]. Because neither somatostatin receptor imaging nor the corresponding therapeutic options were available in Suriname at that time, the patient was referred abroad. Owing to financial constraints, however, he was unable to pursue further evaluation and was subsequently lost to follow-up until his referral by a general surgeon in September 2025 for assessment of a growing subcutaneous thoracic mass. A chest x ray was done showing growth of the pulmonary tumors detected in 2021. Ultrasonography of the subcutaneous thoracic mass demonstrated a highly vascularized subcutaneous tumor measuring 9 cm by 5 cm. Laboratory investigations demonstrated: hemoglobin 8.5 mmol/L, leukocyte count 7.4×10^9/L, platelet count 180×10^9/L, C-reactive protein (CRP) 0.7 mg/dL, urea 5.6 mmol/L, creatinine 98 μmol/L, sodium 139 mmol/L, and potassium 4.2 mmol/L. Urinalysis, including urine sediment examination, was unremarkable, with no evidence of he-

maturia, proteinuria or red blood cell casts. CT scanning of the chest, performed in November 2025, confirmed the presence of a subcutaneous thoracic tumor measuring 78 × 52 × 91 mm, with both solid and cystic components. In addition, multiple pulmonary tumors of varying size were identified in the right lung, the largest measuring 67 × 41 mm (Figure 3). A smaller tumor was also present in the left lower lobe. Right hilar lymphadenopathy was noted, whereas pleural and pericardial effusions were absent. CT imaging of the abdomen revealed no abnormalities.

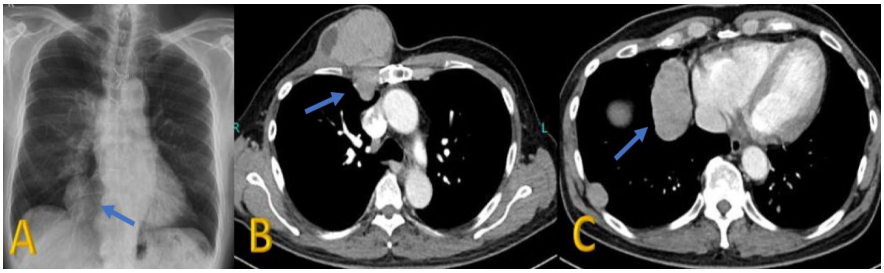

Figure 3. Compared to 2021: chest x ray (A) shows growth of the tumor in the right lung. (B) shows growth of the subpleural tumor with protrusion through the chest wall and (C) also shows growth of the thoracic tumor compared to 2021.

To establish a definitive diagnosis, an ultrasound-guided percutaneous biopsy of the thoracic mass was performed. Histopathological examination demonstrated a predominantly inflammatory infiltrate composed of epithelioid histiocytes, lymphocytes, eosinophils, and neutrophils, with focal granuloma formation and areas of necrosis. Features of vasculitis were also observed (Figure 4).

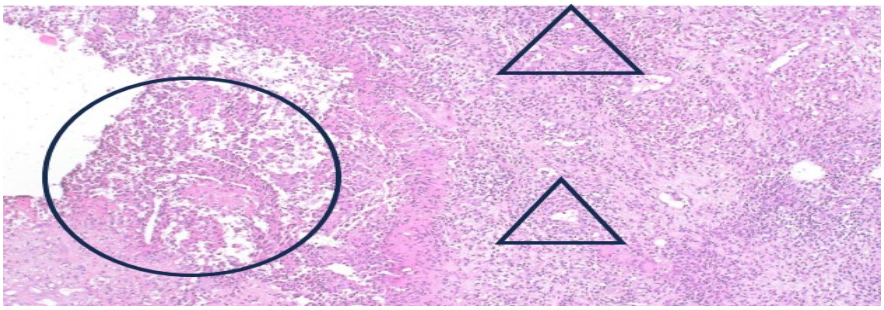

Figure 4. The left side of the image reveals a focal area of necrosis (ellipse), surrounded by an inflammatory infiltrate predominantly composed of histiocytes. The right side demonstrates multiple vessels exhibiting inflammatory involvement of the vessel walls (triangles) (hematoxylin and eosin stain, 100× magnification).

Ziehl-Neelsen staining was negative for acid-fast bacilli, while immunohistochemical studies excluded both carcinoid tumor recurrence and carcinoma. Collectively, the histopathological findings were suggestive of granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis. Following the biopsy findings the antineutrophil cytoplasmic antibody (ANCA) test was performed to further support the diagnosis of GPA. The PR3-ANCA and MPO-

ANCA test results turned out to be negative. The diagnosis was discussed with the patient, and immunosuppressive therapy was proposed. However, the patient declined treatment because he was asymptomatic and, considering his age of 78 years, opted against immunosuppressive therapy. At present, he remains asymptomatic, and a watchful waiting strategy has been adopted.

3. Discussion

During the retrospective analysis of this case, it became apparent that additional investigations had been performed on the mediastinal tumor specimen resected in 2008. At that time, tissue samples had been referred to an overseas reference laboratory for extended immunohistopathological evaluation. The reference laboratory results of February 2008 demonstrated that the lesion had been misclassified initially and was in fact a type A thymoma rather than a typical carcinoid tumor. This finding was only identified during the preparation of this case report. Although distinguishing between these entities on the basis of cytological features alone can be challenging, both type A thymomas and typical carcinoid tumors are generally associated with a relatively favorable long-term prognosis. Nevertheless, both neoplasms are known to exhibit late recurrence, sometimes occurring many years after the initial surgical resection [7]. A comparison of **Figure 2** and **Figure 3** illustrates the progressive enlargement of the lesions identified in 2021, which at that time were interpreted as recurrent thymic neoplastic disease. Over a four-year interval, substantial growth was observed. Most notably, the substernal lesion shown in **Figure 2(B)** demonstrated progressive extension through the anterior chest wall, ultimately resulting in the clinically apparent right-sided subcutaneous pectoral mass. This pattern of growth appeared highly suggestive of locally invasive recurrent disease originating from the previously resected mediastinal tumor. Given the patient's medical history, together with the radiological findings of an enlarging substernal mass, chest wall involvement, and multiple pulmonary lesions, the initial clinical suspicion was that these abnormalities represented recurrent or metastatic thymoma occurring many years after the original thymectomy [8]. Such a diagnosis appeared plausible in light of the known, albeit uncommon, propensity of thymomas for late recurrence. Unexpectedly, histopathological examination of the biopsy specimen obtained from the subcutaneous pectoral mass demonstrated no evidence of recurrent neoplastic disease. Instead, the lesion showed characteristic inflammatory and granulomatous features consistent with granulomatosis with polyangiitis (GPA; formerly Wegener's granulomatosis). Further serological evaluation revealed the absence of antineutrophil cytoplasmic antibodies (ANCA), establishing the diagnosis of ANCA-negative GPA [4]-[6]. Approximately 10% - 20% of patients with GPA are ANCA negative, although the underlying mechanisms responsible for the absence of detectable autoantibodies remain incompletely understood [4]-[6]. ANCA-negative GPA has been reported to exhibit a clinical phenotype distinct from that of ANCA-positive disease, often characterized by more localized manifestations, less extensive systemic involvement, and a lower frequency of renal disease [4]-[6]. The predominantly localized

disease pattern observed in our patient is consistent with these previously reported clinical characteristics. An additional noteworthy aspect of this case is the potential relationship between prior thymectomy and the subsequent development of autoimmune disease. The thymus plays a central role in T-cell maturation and the establishment of immunological self-tolerance. Thymectomy performed for indications other than myasthenia gravis has been associated with the later development of autoimmune disorders, although such complications remain rare [12]. It has been hypothesized that removal of the thymus may result in long-term alterations in immune regulation, including persistent changes in circulating cytokine profiles and T-cell homeostasis [13]. These immunological disturbances may contribute to the emergence of autoimmune phenomena years or even decades after thymectomy. Although a direct causal relationship cannot be conclusively established in the present case, the occurrence of ANCA-negative GPA seventeen years after thymectomy raises the possibility of a delayed autoimmune consequence of thymic resection and adds to the limited body of literature describing this unusual association.

4. Conclusion

Several years after undergoing thymectomy, our patient developed antineutrophil cytoplasmic antibody (ANCA)-negative granulomatosis with polyangiitis (GPA). The occurrence of GPA following thymectomy is exceptionally uncommon and has only rarely been described in the literature. This observation raises the possibility of a long-term disturbance in immune regulation after removal of the thymus, potentially contributing to the development of autoimmune disease. Although a causal relationship cannot be definitively established, the delayed onset of ANCA-negative GPA in our patient represents a noteworthy and exceedingly rare complication occurring many years after thymectomy.

Conflicts of Interest

We have no conflict of interest to report, nor have we received any funding for the preparation of this case report.

Credit Author Contributions

MSS: data curation, resources, writing, reviewing.

M. C: data curation, resources, reviewing.

IT: data curation, resources, reviewing.

FAG: conceptualization, data curation, original draft preparation, resources, supervision.

References

[1] Manna, R., Cadoni, G., Ferri, E., Verrecchia, E., Giovinale, M., Fonnesu, C., *et al.* (2008) Wegener's Granulomatosis: An Update on Diagnosis and Therapy. *Expert Review of Clinical Immunology*, **4**, 481-495. https://doi.org/10.1586/1744666x.4.4.481

[2] Potentas-Policewicz, M. and Fijolek, J. (2024) Granulomatosis with Polyangiitis: Clinical Characteristics and Updates in Diagnosis. *Frontiers in Medicine*, **11**, Article ID: 1369233. https://doi.org/10.3389/fmed.2024.1369233

[3] Cavoli Gioacchino, L., Rita, P., Luisa, B., Calogera, T., Angelo, F. and Carlo Rotolo Ugo, G. (2013) Clinical and Histological Features in Wegener Granulomatosis. *American Journal of Medical Sciences and Medicine*, **1**, 1-4. https://doi.org/10.12691/ajmsm-1-1-1

[4] Mikami-Saitoh, Y., Kyoyama, H., Hirata, Y., Kikuchi, S., Sakai, K., Mikami, S., *et al.* (2023) Anti-Neutrophil Cytoplasmic Antibody-Negative Granulomatosis with Polyangiitis Manifesting through Pulmonary Nodules and a Genital Lesion. *Respiratory Medicine Case Reports*, **46**, Article ID: 101951. https://doi.org/10.1016/j.rmcr.2023.101951

[5] Andrade, M.M., Fernandes, M., Freire, S. and Cruz, D. (2023) Limited Antineutrophil Cytoplasmic Antibodies (ANCA)-Negative Granulomatosis with Polyangiitis: Successful Response to Rituximab. *Cureus*, **15**, e41826. https://doi.org/10.7759/cureus.41826

[6] McCarthy, E., Mustafa, M. and Watts, M. (2017) ANCA-Negative Granulomatosis with Polyangiitis: A Difficult Diagnosis. *European Journal of Case Reports in Internal Medicine*, **4**. https://doi.org/10.12890/2017_000625

[7] Granberg, D., Juhlin, C.C., Falhammar, H. and Hedayati, E. (2023) Lung Carcinoids: A Comprehensive Review for Clinicians. *Cancers*, **15**, Article No. 5440. https://doi.org/10.3390/cancers15225440

[8] Lou, F., Sarkaria, I., Pietanza, C., Travis, W., Roh, M.S., Sica, G., *et al.* (2013) Recurrence of Pulmonary Carcinoid Tumors after Resection: Implications for Postoperative Surveillance. *The Annals of Thoracic Surgery*, **96**, 1156-1162. https://doi.org/10.1016/j.athoracsur.2013.05.047

[9] Petrella, F., Cara, A., Cassina, E.M., Libretti, L., Pirondini, E., Raveglia, F., *et al.* (2025) Pulmonary Carcinoids: Diagnostic and Therapeutic Approach. *Cancers* (*Basel*), **17**, Article No. 2748.

[10] Costa, F. and Gumz, B. (2014) Octreotide—A Review of Its Use in Treating Neuroendocrine Tumours. *European Endocrinology*, **10**, 70-74. https://doi.org/10.17925/ee.2014.10.01.70

[11] La Salvia, A., Modica, R., Rossi, R.E., Spada, F., Rinzivillo, M., Panzuto, F., *et al.* (2023) Targeting Neuroendocrine Tumors with Octreotide and Lanreotide: Key Points for Clinical Practice from NET Specialists. *Cancer Treatment Reviews*, **117**, Article ID: 102560. https://doi.org/10.1016/j.ctrv.2023.102560

[12] Tsirkin, I., Khateb, M., Aran, D., Kaz, A. and Shelly, S. (2025) Survival and Autoimmune Risks Post-Thymectomy. *Frontiers in Immunology*, **16**, Article ID: 1504496. https://doi.org/10.3389/fimmu.2025.1504496

[13] Kooshesh, K.A., Foy, B.H., Sykes, D.B., Gustafsson, K. and Scadden, D.T. (2023) Health Consequences of Thymus Removal in Adults. *New England Journal of Medicine*, **389**, 406-417. https://doi.org/10.1056/nejmoa2302892