Tracheobronchial stenosis in patients with granulomatosis with polyangiitis: histopathological findings and long-term outcome

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Abstract

Objective

Tracheobronchial stenosis (TBS) occurs in a minority of patients with granulomatosis with polyangiitis (GPA). We aimed to i) review the literature on histopathological findings in patients with GPA and TBS, and ii) describe histopathological findings and long-term outcomes in a local cohort.

Methods

PubMed was searched for articles including biopsy findings in patients with GPA and TBS until May 2023. Our local cohort included patients diagnosed with GPA and TBS during 1991–2022. Demographic and clinical characteristics, number of biopsies, histopathological findings (GPA-characteristic vs. non-specific) and number of endoscopic dilations during follow-up were recorded.

Results

In the reviewed literature, 203 patients were identified; 128 (63%) had a total of 296 biopsies; 64 (22%) with GPA-characteristic findings. The local cohort comprised 27 patients, the median (range) age at GPA diagnosis was 31 (14–76) years, the follow-up time was 17 (0–31) years. TBS occurred 2 (0–21) years after the GPA diagnosis. In the local cohort, 67 biopsies from 26 patients were available. GPA-characteristic findings were seen in 15 (22%) of these. Twenty-three patients underwent an endoscopic dilation during follow-up, and the number of dilations was 5 (1–100). Eighteen patients had ≥2 dilations; the interval between dilations was 23 (2–103) months.

Conclusion

The majority of patients with GPA and TBS experience recurring airway stenoses. GPA-characteristic findings are seen in approximately one in five tracheobronchial biopsies obtained from GPA patients with TBS. A biopsy could be of clinical value in the diagnostic process or to exclude other causes of stenosis.

Key words

granulomatosis with polyangiitis, tracheobronchial stenosis, subglottic stenosis, histopathology, endoscopic dilation

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Introduction

Granulomatosis with polyangiitis (GPA) is a systemic autoimmune inflammatory disease, commonly involving the upper and lower airways and kidneys (1-4). Tracheobronchial stenosis (TBS) is a manifestation occurring in a minority (8–23%) of patients with GPA; it is often seen independently of other features of disease activity (5-9). Stenoses are most often located in the subglottic area, but endobronchial stenoses may also be seen (9-11). Endoscopic dilation with concomitant perioperative therapies such as steroid injections or laser excision have been applied to relieve symptoms, but the long-term effect is generally poor, and many patients require recurrent procedures (12). Similarly, no standard systemic treatment regimen is agreed upon, since the evidence for efficacy of specific immunosuppressive therapies is based on small retrospective cohorts (9, 11, 13-15).

Studies investigating histopathological findings in patients with TBS are heterogeneous (7, 11, 16-18). In a study assessing 140 serial biopsies performed in 26 patients with TBS, non-specific acute and chronic inflammation was the most common finding, whereas characteristic granulomatous inflammation or vasculitis was rare, regardless of the gross appearance of the trachea (7). In another study including 27 patients, 26 serial biopsies only showed granulomatous inflammatory changes consistent with GPA in 15% of the specimens (17). However, biopsies were only available in 16 (59.3%) of the reported patients, and clinical disease characteristics were not thoroughly addressed (17). Thus, the clinical value of histological findings in patients with TBS may be limited. Moreover, studies addressing TBS including both histopathological findings and long-term outcomes are few (7, 11).

With this study, we aimed 1) to review the literature on histopathological findings in TBS, and 2) to describe histopathological findings and long-term outcomes in a local cohort of 27 patients with GPA and TBS followed at our tertiary care centre during 1991-2022.

Methods

Literature review

PubMed was searched for articles published up until May 2023 using combinations of the following search terms: "Granulomatosis with polyangiitis", "GPA", "Wegener's granulomatosis", "WG", "tracheobronchial stenosis", "subglottic stenosis", "histological findings", "pathological findings" and "biopsies". We included English language articles describing adult patients with GPA. The studies should include at least one biopsy with description of the histopathological findings. Titles, abstracts and if necessary, the original articles were screened for eligible studies. The reference lists of the eligible studies were subsequently screened for additional eligible studies. The histological findings of biopsy specimens obtained from the patients were recorded.

Local cohort

Among 338 patients followed for GPA at our tertiary care centre, we retrospectively identified those who had been diagnosed with TBS following a laryngoscopy or bronchoscopy procedure. The following data were collected retrospectively from GPA diagnosis until January 2022 through review of medical charts: dates of GPA diagnosis, TBS diagnosis, first and last endoscopic dilation procedure, and last visit at the clinic. Data on GPA manifestations, biopsy findings, anti-neutrophil cytoplasmic antibody (ANCA) status and specificity, vasculitis damage index (19) at the last visit, history of immunosuppressive therapies and the number of dilation procedures during follow-up.

Biopsy findings

Histopathological findings that included either granulomatous inflammation with or without necrosis, multinucleated (giant) cells, micro-abscesses, or vasculitis were considered characteristic for GPA (18). All other findings were considered non-specific.

Statistics

Descriptive statistics are reported as numbers and proportions for categorical variables and as median values (range) for continuous variables. Data

Table I. Studies retrieved from the literature search.

Study	Year	Design	Number of patients with biopsies / total n of patients (% with biopsy)	Total number of biopsies (n)	Biopsy findings	
					Findings characteristic for GPA*, n (%)	Non-specific findings, n (%)
Catano *et al*. (16)	2021	Retrospective	18/33 (55%)	18	8 (44%), necrotizing granulomatous inflammation	Non-specific inflammation, 10 (56%)
Girard *et al*. (11)	2015	Retrospective	17/26 (65%)	26	12 (47%), granuloma**	
Pajor *et al*. (26)	2015	Case report	1/1 (100%)	1		1 (100%), chronic inflammation, ulcers and granulation tissue
Jakez-Ocampo *et al*. (27)	2010	Case report	2/2 (100%)	2	2 (100%), giant cells, necrotizing ulcers	
Solans-Laqué *et al*. (13)	2008	Case series	6/6 (100%)	6	6 (100%), necrotizing vasculitis and granuloma	
Gluth *et al*. (17)	2003	Retrospective	16/27 (59%)	26	4 (15%)	
Stappaerts *et al*. (28)	2000	Case report	1/1 (100%)	1	None	1 (100%), severe non-specific inflammation
Langford *et al*. (7)	1996	Retrospective	26/43 (60%)	140	7 (5%)	
Herridge *et al*. (29)	1996	Case series	5/5 (100%)	5	1 (20%), granulomatous inflammation and vasculitis	4 (80%), non-specific chronic inflammation
Daum *et al*. (6)	1995	Retrospective	17/17 (100%)	21	12 (57%), presence of either necrosis, giant cells, vasculitis/capillaritis and microabscesses or a combination	
Gaughan *et al*. (30)	1990	Case report	1 (100%)	1		1 (100%), acute and chronic inflammation
Devaney *et al*. (18)	1990	Retrospective	Not stated	17	4 (23%), vasculitis, necrosis, granulomas, giant cells	None
Hoare *et al*. (31)	1989	Case series	4/4 (100 %)	14	None	14 (100%), acute and chronic inflammation
Hellman *et al*. (32)	1987	Case report	1 (100%)	1	1 (100%), granulomas, necrosis, vasculitis	
Waxman and Bose (33)	1986	Case report	2/2 (100%)	3	1 (33%)	2 (67%)
McDonald *et al*. (34)	1982	Case series	3/17 (18%)	3	3 (100%) "tissue consistent with GPA"	
Arauz-Fonseca (35)	1982	Case series	2/10 (20%)	2	1 (50%), granulomatous process with fibrinoid necrosis	1 (50%), chronic inflammation
Lampman *et al*. (36)	1981	Case report	1 (100%)	1	1 (100%), multinucleated cells	
Scully *et al*. (37)	1979	Case report	1 (100%)	1	None	1 (100%), non-specific inflammation
Djalilian *et al*. (38)	1975	Case series	4/5 (80%)	4	None	4 (100%), inflammation and fibrosis
Talerman and Wright (39)	1972	Case report	1 (100%)	3	1 (33%)	2 (67%)
Total			128/203 (63%)	296	64/296 (22%)	

GPA: granulomatosis with polyangiitis.
*Granulomatous inflammation with or without necrosis, multinucleated (giant) cells, micro-abscesses or vasculitis;
**granuloma, vasculitis and necrosis is reported in 47%, 12% and 29% of the biopsies, respectively, however the distribution of each finding is not further described.

were stratified according to sex and ANCA specificity; non-parametric tests are used for group comparisons. Statistical analyses were done using StataNow 18 SE.

Ethics

This work is based on literature review and retrospective investigation of clinical charts and does not require approval from an ethical committee. The living subjects included in the clinical chart review have provided informed, written consent.

Results

Literature review

An overview of the 21 articles identified in the literature search is seen in Table I. The studies were case reports (9), case series (6) or retrospective studies (6). A total of 203 patients were described in the identified articles. The number of patients per study ranged from 1 to 43.

Local cohort

The local cohort comprised 27 GPA patients, 13 men and 14 women diagnosed with TBS during 1991-2022. Nine (33%) of the 27 patients presented with the stenosis at GPA diagnosis. In the remaining 18 patients, TBS occurred after a median (range) of 6 (1–21) years

following the GPA diagnosis. Twenty-four patients had subglottic stenosis (SGS), 7 patients had endobronchial stenosis (EBS) and 4 patients had both (Fig. 1). EBS developed before SGS in 3 of the 4 patients (time between EBS and SGS: 3–26 years), while in one patient, SGS developed 4 years prior to EBS. Twenty-three patients (85%) were proteinase 3 (PR3) ANCA positive, 3 (11%) were myeloperoxidase (MPO) ANCA positive, while one patient was ANCA negative. The median (range) age at GPA diagnosis was 31 (14-76) years; the follow-up time since diagnosis was 17 (0–31) years. The follow-up time stratified by sex was: 14 years (2–24) for men *versus* 17 years (0–31) for women (p=0.56) and stratified by ANCA specificity: 17 years (1–32) for PR3 positive, 9 years (0–17) for MPO positive and 25 years for the single ANCA negative patient (p=0.25), data not shown. Patient characteristics are shown in Table II.

Biopsy findings

Among the 203 patients identified through literature review, 128 (63%) had at least one available biopsy. In total, 296 biopsies had been performed in this group. The number of biopsies per study ranged from one to 140. The number of biopsies performed in each patient was not systematically reported. Characteristic GPA findings were seen in 64 (22%) of the biopsies (Table I).

In our local cohort, 26 of the 27 patients had at least one and up to nine available biopsies. In total, 67 biopsies were performed. Characteristic GPA findings were seen in 15 (22%) biopsies from 11 patients; these patients were all PR3-ANCA positive. In the 8 patients presenting with stenosis at the time of GPA diagnosis, 1 in 6 biopsies showed characteristic GPA findings. In the 19 patients, who developed a stenosis at a later stage, such findings were seen in 5 of the 10 available biopsies. Biopsy-findings in the remaining biopsies included acute and chronic inflammation, eosinophilocyte infiltration, granulation tissue and fibrosis. The distribution of positive biopsies according to the number of biopsies in each patient is depicted in Figure 2.

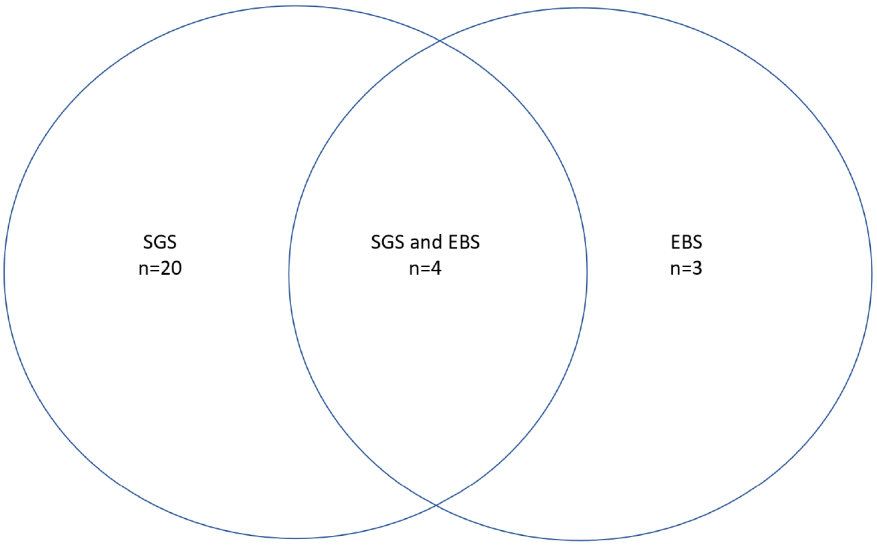

Fig. 1. Distribution of 27 patients with tracheobronchial stenosis.
SGS: subglottic stenosis; EBS: endobronchial stenosis.

Table II. Characteristics of the 27 patients in the local cohort.

Characteristic	value
Women n, (%)	14 (52)
Demography, median (range)	
Age at GPA diagnosis, years	31 (14-76)
Age at TBS diagnosis, years	34 (17-77)
Time from GPA diagnosis to TBS, years	2 (0-21)
Follow-up since GPA diagnosis, years	17 (0-31)
Clinical manifestations, n (%)	
Ear-nose-throat	26 (96)
Eyes	11 (41)
Retro-orbital mass	2 (7)
Skin	11 (41)
Joints	15 (56)
Lungs	12 (44)
Kidney	6 (22)
Peripheral nerve system	3 (11)
Central nervous system	2 (7)
ANCA, n (%)	
PR3 positive	23 (85)
MPO positive	3 (11)
ANCA negative	1 (4)
Immunosuppressants used, n (%)	
Cyclophosphamide	17 (63)
Rituximab (induction)	6 (22)
Rituximab (maintenance)	8 (30)
Mycophenolate mofetil	12 (44)
Methotrexate	23 (85)
Azathioprine	13 (48)
Other	1 (4)

GPA: granulomatosis with polyangiitis; TBS: tracheobronchial stenosis; ANCA: antineutrophil cytoplasmic antibody; PR3: proteinase 3; MPO: myeloperoxidase.

Endoscopic dilation

Twenty-three patients underwent an endoscopic dilation procedure during follow-up, the median (range) number of dilations was 5 (1–100). Stratification by sex showed that the 10 men had 10 (1–100) dilations, and the 13 women had 5 (1–56) dilations, p=0.18. Stratification by ANCA specificity showed that the number of dilations in 19 PR3 positive patients was 10 (1–100) and in 3 MPO positive patients 1

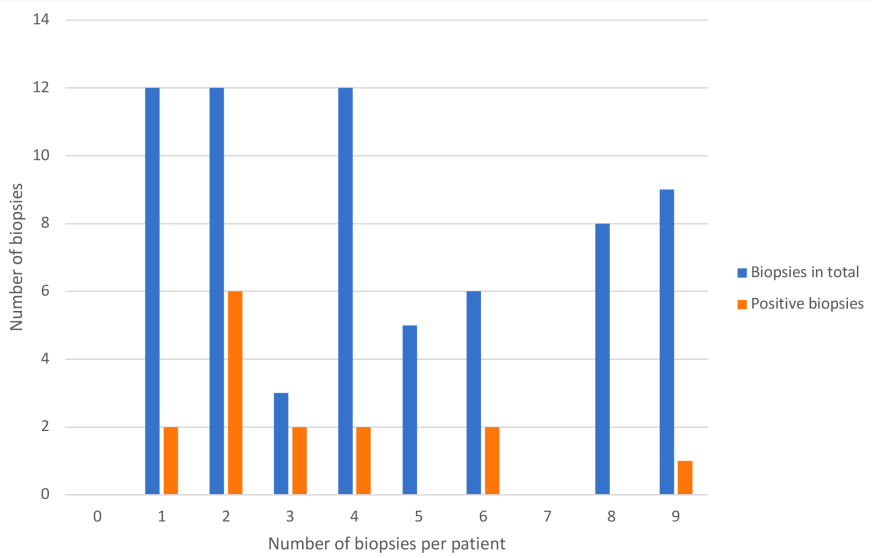

Fig. 2. Distribution of biopsies from the 27 patients in the local cohort.

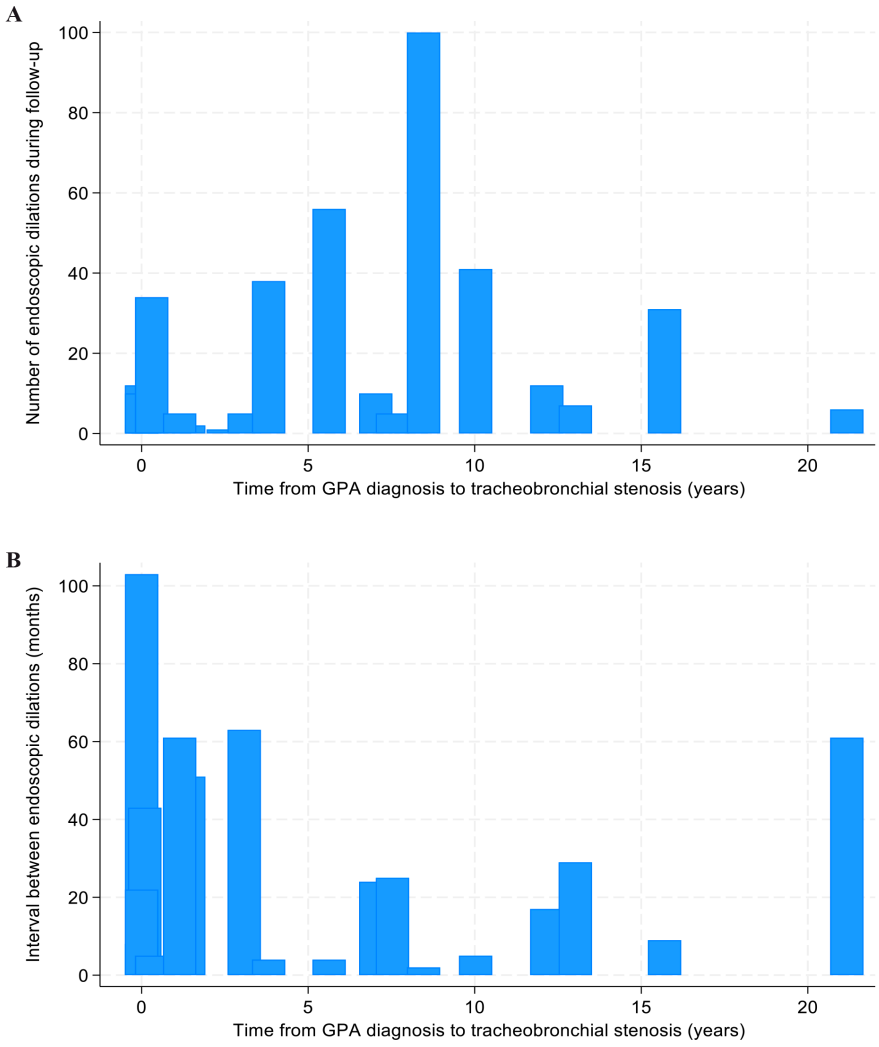

Fig. 3. (**A**) Number of endoscopic dilations according to the time from GPA diagnosis to first occurrence of tracheobronchial stenosis in 23 patients with at least one endoscopic dilation. (**B**) Median interval between endoscopic dilations according to the time from GPA diagnosis to first occurrence of tracheobronchial stenosis in 18 patients with ≥2 endoscopic dilations.

(1-2), p=0.09. One patient was ANCA negative.

In 18 (78%) of the patients, 2 or more endoscopic dilations were performed. Among these patients, the median (range) interval between dilations was 23 (2-103) months. The interval between dilations in the 9 men was 22 (2–43) months and in the 9 women 51 (4–103) months, p=0.29. The interval between dilations was not analysed according to ANCA specificity, since only one MPO positive patient had ≥2 dilations.

The number of dilation procedures and the interval between dilations according to the time from GPA diagnosis to occurrence of TBS are shown in Figure 3a and b, respectively. Two outliers included one patient, who had 100 dilations over a 21-year follow-up period (Fig. 3a), and another patient, who had 2 dilations with an interval of 103 weeks (Fig. 3b). The number and interval between dilations according to age at GPA diagnosis are seen in Supplementary Figures S1 and S2.

Discussion

In this study, we describe the histopathological findings made in biopsy material from tracheobronchial stenoses occurring in the setting of GPA. We also describe the clinical course of GPA patients affected by such stenoses. Based on data reported in earlier publications and on findings made in a local GPA cohort, we see a relatively low frequency of GPA-characteristic histopathological findings in the airway lesions of GPA patients with TBS. Thus, histological findings characteristic of GPA were only observed in approximately one out of five biopsies performed during dilation procedures among GPA patients with TBS described in previous studies as well as in a local cohort. The majority of patients in the local cohort had at least one endoscopic dilation done. No clear association between the number of dilations and demographic or disease-related characteristics was seen.

Characteristic histological findings add 2 out of 5 points needed for the classification of GPA according the 2022 ACR/EULAR classification criteria (20). A biopsy may thus be a supple-

ment to the workup in patients with suspected GPA. Biopsies taken from the respiratory tract often show non-specific inflammation and their value in the diagnostic process is generally considered low (21). However, histopathological studies have often pooled biopsies from several anatomic sites, preventing possible differences in sensitivity and diagnostic yield to stand out (22-24). To our knowledge, this is the first review of studies investigating biopsies from the tracheobronchial area specifically. Twenty-two percent of biopsies showed GPA characteristic findings among the reported cases, out of the 63% with an available biopsy. In our local cohort, 96% (26 out of 27) had a biopsy done, however, the proportion of positive biopsies (22%) was identical. Since our study did not yield a higher proportion of positive biopsies, our observations indicate that approximately one in five positive biopsies from the tracheobronchial area is what can be expected regardless of the setting. A larger prospective study with systematic collection of biopsies at predefined timepoints could potentially add further insights, however, such a study may not be feasible.

Among GPA patients followed at our tertiary care centre, the prevalence of TBS was 8% (27 of 338 patients) which is in the lower end compared with prevalences reported in other cohorts (ranging from 8% to 23%) (7-10, 13, 25). Differences in recruitment strategies (multicentre *vs*. single centre) and classification criteria over time may have contributed to the variation in prevalent cases across studies. The demographic and disease related characteristics of patients in our local cohort are comparable to previously reported data on patients with GPA and TBS (8-11, 14, 25).

The majority of patients in our local cohort had more than one endoscopic dilation done. Although the women and men had a similar follow-up, the women tended to have less dilations and longer intervals between them, but the differences did not reach statistical significance. In a nationwide, retrospective study by Terrier *et al*. including 47 patients with TBS but with a shorter follow-up compared to ours, no such sex-differences were seen (9). Our stratification on ANCA specificity showed that PR3 positive patients tended to have more dilations, however there were very few available MPO positive patients, which hampers any clear conclusions. Similarly, in other studies that have included ANCA specificity, only a minority were MPO positive, and no specific comparisons of outcomes based on ANCA specificity were done (7-9, 11, 13). Terrier *et al*. have reported that a shorter time interval between GPA diagnosis and the need for an endoscopic procedure, was associated with a higher risk of relapsing stenosis (9). Our data did not show any clear association in this direction, but the small numbers of patients under study constitutes a limiting factor in our investigation.

A major strength of this work is the combined inclusion of a large number of patients from the literature and a smaller cohort with a near complete proportion having an available biopsy. Moreover, the follow-up period was long, which is an advantage when evaluating a rare manifestation reoccurring with varying time intervals across the studied patients.

Limitations include the heterogeneity of the reviewed studies, not only in the number of included patients, but also in the reporting of the pathological findings and the timing of the biopsies in relation to the disease course. In the local cohort, we were able to assess some of these aspects in more detail, however, the retrospective chart review lacked detailed information on disease activity and treatment decisions. Our study was thus not suitable for evaluating systemic treatments and comparing them with other reports suggesting promising effects *e.g*. of high dose prednisolone (9) and leflunomide (14).

In conclusion, the majority of patients with GPA and TBS experience recurring airway stenoses. In this study, we did not identify any clear factors associated with recurring TBS. Since one in five tracheobronchial biopsies showed GPA characteristic findings, a biopsy could be of clinical value in the diagnostic process or to exclude other causes of stenosis.

Take home messages

- The majority of patients with GPA and tracheobronchial stenosis experience recurring stenoses.
- GPA-characteristic findings are seen in approximately one in five tracheobronchial biopsies.
- A biopsy could be of clinical value in the diagnostic process or to exclude other causes of stenosis.

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