CASE REPORT

Granulomatosi s with polyangiitis in a patient with polydipsia, facial nerve paralysis, and severe otologic complaints: a case report and review of the literature

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Abstract

Background: Granulomatosis with polyangiitis, formerly known as Wegener granulomatosis, is a necrotizing vasculitis with granulomatous inflammation that belongs to the class of antineutrophil cytoplasmic antibodies-positive diseases. It occurs in a localized and a systemic form and may present with a variety of symptoms. Involvement of the upper respiratory tract is very common, while neurologic, endocrinological, and nephrological dysfunction may occur.

Case presentation: We describe the case of a 29-year-old Central European male patient presenting with severe bilateral sensorineural hearing loss, otorrhea, and one-sided facial nerve paralysis. The patient was unsuccessfully treated with i.v. antibiotics at another hospital in Berlin, and tympanic tubes were inserted. After presentation to our emergency room, he was hospitalized and further diagnostics started. Increased fluid intake and 12 kg weight gain over the last months were reported. The patient was diagnosed with granulomatosis with polyangiitis and diabetes insipidus. The patient’s condition improved after treatment with rituximab.

Discussion: A comprehensive PubMed search of all articles with granulomatosis with polyangiitis and diabetes insipidus was conducted to assess which combination of symptoms occurs simultaneously and whether other parts of the pituitary are commonly involved. The 39 selected articles, describing 61 patients, showed that ear–nose–throat involvement occurred most commonly, in 71% of cases. Of patients, 59% had involvement of the anterior pituitary gland, while true panhypopituitarism occurred in 13% of cases. Only one case report featured the same set of symptoms as described herein.

Conclusion: Granulomatosis with polyangiitis is a highly variable disease, commonly involving the upper airways, but that may present with symptoms solely related to the pituitary gland. Clinicians should have a low threshold to investigate for granulomatosis with polyangiitis in patients with therapy-resistant otorrhea. Patients may present with a complex set of symptoms, and integrating different specialists when additional symptoms occur may lead to faster diagnosis.

Keywords: Granulomatosis with polyangiitis (D014890), Hearing loss, sensorineural (D006319), Hypopituitarism (D007018), Diabetes insipidus (D003919), Facial paralysis (D005158)
inflammation. It involves primarily small and medium-sized vessels with a predilection for the upper and lower airways. Necrotizing glomerulonephritis is common [1]. The current definition was formed in 2012 by the Chapel Hill Conference for Consensus Criteria [2].

GPA belongs to a group of anti-neutrophil cytoplasmic antibody (ANCA)-positive diseases called ANCA-associated vasculitis, although ANCA testing may be negative in 10–20% of cases [3].

Research led to the identification of different subsets of ANCA-positive diseases. Anti-proteinase 3 activity (PR3-ANCA) is most common in GPA, while patients with microscopic polyangiitis and Churg–Strauss syndrome are most often anti-myeloperoxidase ANCA (MPO-ANCA) positive [4].

GPA occurs in a systemic and a localized form. ENT involvement occurs in up to 90% of individuals. The sinonasal area is most commonly affected [5–7]. Otolologic involvement is the second most common presentation [8, 9]. Localized forms of GPA with involvement of the ear and/or the upper respiratory tract have been described, especially in younger patients [10]. Dysfunction of the central nervous system is common at some point of the disease; the result can be dysfunction of cranial nerves II, VI, and VII, but also endocrine involvement of the pituitary gland [11–14]. Severe headache is the most common nonendocrine symptom of hypophysitis. Chiasmal syndrome and ocular paresis may occur due to the increased sellar mass. Isolated DI centralis may occur as an endocrine symptom and is characterized by deficient secretion of arginine vasopressin (AVP), leading to clinical symptoms of polyuria and polydipsia. More frequently, combined hormonal insufficiency of the anterior and posterior pituitary occurs. Hypogonadotrophic hypogonadism, secondary hypothyroidism, and potentially life-threatening secondary adrenal insufficiency are the most frequent endocrine disorders of the anterior pituitary, at least in patients with primary hypophysitis, in contrast to the finding of vulnerable gonadal and growth hormone axis and more robust adrenal and thyroid axes in patients with pituitary adenoma [15]. However, data on hypophysial function of patients with secondary hypophysitis due to GPA are scarce.

We present the case of a patient with localized, severe granulomatosis with polyangiitis with otitis media, facial nerve palsy, and pituitary dysfunction resulting in DI.

Case presentation

The patient is a white German male of 29 years of age who presented with otalgia and serous otorrhea (Figs. 1, 2) to the otolaryngology resident on call in the ER of the Charité University Hospital in Berlin in November 2018. The patient reported having these symptoms for 1 month. He had visited an otolaryngology clinic in October 2018 with facial nerve palsy with House–Brackmann score (HB) < III. At the clinic, treatment was started with i.v. antibiotics and prednisolone as well as
tympanocentesis with insertion of T-tubes. Audiometric analysis revealed combined sensorineural and conductive hearing loss on the right side and conductive hearing loss on the left side (Fig. 3). A computed tomography (CT) scan report on his temporal bone described fluid retention on the mastoid on both sides, possibly with osteolysis. Routine laboratory analysis including human immunodeficiency virus and hepatitis showed slightly elevated C-reactive protein (34.9 mg/l) but no further anomalies. On microbiologic testing, the middle ear fluid revealed biological flora without detection of pathogens. Facial nerve function normalized and otorrhea decreased after receiving treatment. The patient was discharged after 7 days with middle ear tubes in situ.

Upon presentation to the ER, facial nerve function was still abnormal with HB score of II, and extensive serous otorrhea persisted. The middle ear ventilation tubes were in situ. The patient also reported increased fluid intake of 9 L per day and weight gain of 12 kg in 7 weeks.

Laboratory analysis showed positive c-ANCA at 87.0 U/ml with increased anti-PR3 activity in combination testing. Anti-MPO activity was negative. On the basis of this testing, a diagnosis of granulomatosis with polyangiitis was established.

A magnetic resonance imaging (MRI) scan of the brain showed slight widening of the pituitaries infundibulum and unclear inhomogeneity between the anterior and posterior pituitary (Figs. 4, 5).

Clinical and laboratory evaluation of the anterior pituitary function did not indicate insufficiency of the somatotropic, gonadotropic, thyrotropic, or corticotrophic axis. In addition, serum prolactin was within the reference range.

Plasma sodium concentration was 144 mmol/l with plasma osmolality of 300 mosm/kg. In a water deprivation test, urine osmolality increased insufficiently from 131 to 418 mosm/kg. Measurement of hypertonic saline-stimulated plasma copeptin (2.63 pmol/l) confirmed the diagnosis of DI.

Treatment for GPA was successfully started with 1 g rituximab i.v., and ANCA values decreased to 22.2 U/ml at 4 months after discharge and finally to 13.4 U/ml in October 2019 (Additional file 1).

![Fig. 3](image-url)  
Audiometric analysis on 26/11/2018, showing mixed hearing loss on the right side with Fletcher index of 80 dB and conductive hearing loss on the left side with Fletcher index of 40 dB.
Discussion

GPA is a variable disease that may present with a complex set of symptoms [11–14]. The patient in our case presented with otalgia, otorhea, facial nerve dysfunction, hearing loss, and polydipsia. Upon presentation to our department, the posterior pituitary gland function was already affected. While the resulting increase in fluid intake was not seen as a burden by the patient, medical history showed that symptoms of DI had existed before otologic involvement. Referral to specialists in endocrinology and rheumatology was sought after the diagnosis of GPA.

A comprehensive systematic PubMed search was performed using the search term ‘Wegener granulomatosis AND pituitary dysfunction OR granulomatosis with polyangiitis AND pituitary dysfunction OR Wegener granulomatosis AND diabetes insipidus OR granulomatosis with polyangiitis AND diabetes insipidus.’ The article references were scanned for additional literature, resulting in a total of 65 articles. All case reports with confirmed diagnosis of GPA in addition to diabetes insipidus were included. Articles published in foreign language (other than English or German) and for which no full text was available were excluded. In total, 39 articles were selected for analysis after applying inclusion and exclusion criteria.

The 39 selected articles described 61 different cases for analysis (Table 1). ENT involvement in patients with GPA was very common, with 43 patients reported to have any ENT area affected (72%). Sinonasal disease was very common, with 21 patients affected (35%). Otologic complaints were less common, being reported in nine patients (15%). Two of the selected case series, representing a total of 17 patients, did not specify the involved subarea. This may result in underestimation of the total percentage of patients affected per subarea.

Of the 61 cases, 36 (59%) experienced at least one symptom of anterior pituitary dysfunction. Hyperprolactemia was the most common hormonal dysbalance \( (n = 20, 33\%) \), while hypothyroidism \( (n = 15, 25\%) \) and hypogonadism \( (n = 14, 23\%) \) ranked second and third. Panhypopituitarism was relatively uncommon and occurred in eight (13%) patients.

Most of the patients examined in this study had an established diagnosis of GPA before developing symptoms suggestive of DI \( (n = 33, 54\%) \). The majority of patients \( (n = 35, 57\%) \) developed ENT symptoms suggestive of GPA earlier than symptoms suggestive of DI. In 11 cases of this subgroup \( (31\%) \), the ENT symptoms occurred days to months before DI symptoms. In 18 cases \( (51\%) \), ENT symptoms developed years prior to diagnosis. Six cases \( (17\%) \) did not include a timeframe for the occurrence of the symptoms.

DI was diagnosed before the diagnosis of GPA in 17 cases \( (28\%) \). In this subgroup, the symptoms of DI developed days to month before diagnosis of GPA in three \( (18\%) \) cases and years before diagnosis of GPA in another three \( (18\%) \) cases. The other cases did not offer a timeframe (Table 2).

Among the reviewed articles, only one report matched our patient’s complaints of therapy-resistant otitis media,
facial nerve paralysis, sensorineural hearing loss, and DI [42]. Although GPA can occur as a very limited localized disease, e.g., as antibiotic-resistant otitis media with or without mastoiditis [51–53], we want to highlight that GPA can also occur with a combination of involved localized areas, as described herein. The current case, as well

Table 1 Published articles with patients matching our inclusion criteria, with publication date and site of ENT involvement

| Author            | Year of publication | ENT involvement                                                                 |
|-------------------|---------------------|---------------------------------------------------------------------------------|
| Garovic et al. [17]| 2001                | No                                                                              |
| Roberts et al. [18]| 1995                | Case 1: no, Case 2: sinusitis                                                    |
| Katzman et al. [19]| 1999                | Case 1: sinonasal congestion, Case 2: stridor, laryngeal stenosis, septal perforation, saddle-nose deformity |
| Hajji-Ali et al. [20]| 1999             | Sinusitis, epistaxis                                                            |
| Muir et al. [21]   | 2004                | Ear infection (not defined) with cyst                                             |
| Düzgün et al. [22]| 2005                | Dry oral mucosa                                                                 |
| Hurst et al. [23]  | 1983                | Sinusitis                                                                       |
| Haynes et al. [24] | 1978                | No                                                                              |
| Czarnecki et al. [25]| 1995              | Sinusitis, septal perforation with saddle-nose deformity                        |
| Roberts et al. [18]| 1995                | No                                                                              |
| Bertken et al. [26]| 1997                | Sinusitis, epistaxis, external otitis                                            |
| Katzman et al. [19]| 1999                | Sinusitis                                                                       |
| Miesen et al. [27] | 1999                | No                                                                              |
| Goyal et al. [28]  | 2000                | No                                                                              |
| Seror et al. [29]  | 2006                | Case 1: sinusitis, Case 2: gingivitis                                            |
| Špišek et al. [30] | 2005                | Epistaxis                                                                       |
| McIntyre et al. [31]| 2007              | No                                                                              |
| Yong et al. [32]   | 2008                | Sinusitis and epistaxis                                                         |
| Cunnington et al. [33]| 2009             | Case 1: epistaxis, nasal crusting, Case 2: otalgia, otorrhea, hearing loss (not specified) |
| Xue et al. [34]    | 2009                | No                                                                              |
| Barlas et al. [35] | 2011                | Sinusitis                                                                       |
| Santoro et al. [36]| 2011                | No                                                                              |
| Tenorio Jimenez et al. [37]| 2011    | Sinusitis, septal perforation, saddle-nose deformity                            |
| Hughes et al. [38] | 2012                | No                                                                              |
| Pereira et al. [39] | 2013                | No                                                                              |
| Kapoor et al. [40] | 2014                | Eight cases with ENT involvement (unspecified)                                  |
| Peters et al. [41] | 2018                | Epistaxis, nasal congestion, recurrent bilateral otitis media, hearing loss     |
| Ohashi et al. [42] | 2017                | Sensorineural hearing loss, otitis media, facial nerve paralysis                 |
| Esposito et al. [13]| 2017              | Case 1: no, Case 2: serous otitis media, Case 3: no                             |
| Xie et al. [43]    | 2017                | No                                                                              |
| Byrne et al. [14]  | 2016                | Sinusitis                                                                       |
| Eli et al. [44]    | 2016                | No                                                                              |
| Vanderheynst et al. [45]| 2015    | Sinusitis                                                                       |
| De Parisot et al. [11]| 2015             | Six of nine cases with ENT involvement (unspecified)                            |
| Bando et al. [46]  | 2015                | Saddle-nose deformity                                                           |
| Sampei et al. [47] | 2014                | Sinusitis                                                                       |
| Slabu et al. [48]  | 2013                | Sinusitis                                                                       |
| Van Durme et al. [49]| 2011              | Sinusitis, otitis media                                                         |
| Dutta et al. [50]  |                     | Case 1: otorrhea, hearing loss, Case 2: nasal blockage, bilateral ear blockage  |
| Author            | Pituitary function                                                                 | First manifestation and latency                                                                 |
|-------------------|-------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------|
| Garovic et al. [22] | Decreased FSH and LH (on estrogen replacement therapy), decreased prolactin, decreased TSH (on thyroid hormone replacement) | Not mentioned                                                                                    |
| Roberts et al. [23] | Low TSH, low FSH and low LH, low cortisol, DI                                        | Four-month history of deteriorating vision with bitemporal hemianopia, 2 months later DI          |
| Katzman et al. [24] | DI, no ant. pituitary deficiency                                                     | Cheek and temporal pain, left-sided hearing loss, sinusitis; several days later DI                 |
| Haji-Ali et al. [25] | Not mentioned                                                                      | Latency unclear                                                                                  |
| Muir et al. [26]   | Not mentioned                                                                       | Nasal symptoms before pituitary gland symptoms, latency unclear                                  |
| Düzgün et al. [27] | Ant. pituitary hormones normal                                                       | Two-month sinusitis; later DI                                                                       |
| Hurst et al. [28]   | Not tested                                                                           | Ear infection and DI at same time                                                                  |
| Haynes et al. [29]  | Not tested                                                                           | Otitis media 2 months before diabetes symptoms                                                     |
| Czarnecki et al. [30] | Hyperprolactemia (not biochemical proven)                                            | Polyarthritic, 3 months later serous otitis media, 6 months later sinusitis                       |
| Roberts et al. [23] | Not tested                                                                           | No ENT involvement                                                                               |
| Bertken et al. [31] | Luteinizing hormone response to gonadotropin-releasing hormone and the thyroid-stimulating hormone response to thyrotropin-releasing hormone were blunted | Hyperprolactemia and DI 3 years after general ENT symptoms                                       |
| Miesen et al. [32]  | Hyperprolactemia, low serum testosterone                                             | Polyuria and polydipsia as presenting symptoms                                                   |
| Goyal et al. [33]   | Hypothyromis                                                                         | Several-year history of GPA, polyuria, polydipsia, leathargy, and headaches                       |
| Seror et al. [34]   | Pituitary hormones normal                                                             | 1987 bloody-crusty rhinitis, septum necrosis, saddle-nose deformity, arthralgia; 2002 for polydipsia and polyuria, diagnosis DI established |
| Špišek et al. [35]  | Panhypopituitarism, low adrenocorticotropin-releasing hormone (ACTH), thyroid-stimulating hormone (TSH), follicle-stimulating hormone (FSH), and luteinizing hormone (LH) and deficit of peripheral hormones. Insulin test further revealed growth hormone deficiency, low insulin-like growth factor (IGF-I) concentration → anterior and posterior pituitary insufficiency | First diagnosis of GPA in 1995 with crusty rhinitis, nasal septum necrosis, mouth ulcers. In 2000, presentation with galactorrhea |
| McIntyre et al. [36] | Ant. pituitary insufficiency (low TSH, low prolactin, low LH, low FSH, low estradiol, low cortisol | 2002 skin abscesses, weight loss (15 kg in 3 months), collapses, and erectile dysfunction. Later epistaxis and headaches (no timeframe mentioned) |
| Yong et al. [37]    | DI; further pituitary function tests normal but secondary adrenal insufficiency and secondary hypogonadism | Polydipsia and polyuria as presenting sign with central DI established, otitis media, and thickening of sphenoid and maxillary mucosa found |
| Author                  | Pituitary function                                      | First manifestation and latency                                                                                                                                                                                                 |
|------------------------|---------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Cunnington et al. [38] | No ant. pituitary dysfunction                           | July 2000, epistaxis, haemoptysis, nasal crusting, vasculitic skin rash, and bilateral episcleritis; no specific diagnosis yet. May 2005, polyuria and polydypsia; diagnosis of DI and GPA                                                                         |
|                        | No pituitary dysfunction                                | 1995, GPA with malaise, nose bleeds, and sinusitis; 2004, diagnosis of DI                                                                                                                                                       |
|                        | Prolactin and thyroid function normal, other values tampered by prednisolone and oral anticoagulation    | 2002, otalgia, otorrhoea, and hearing loss; treatment for GPA started. In 2003, polyuria, polydypsia, and frontal headaches; diagnosis of DI                                                                                           |
| Xue et al. [39]        | LH, FSH, GH, and TSH normal                            | Intermittent fever and polydypsia, insensibility of her lower extremities, pitted edema on face and lower extremities since 0.5 year. Five months later, polydypsia; diagnosis of DI and GPA                                                                |
| Barlas et al. [40]     | High prolactin                                         | Polydypsia, polyuria, sinusitis at presentation                                                                                                                                                                                  |
| Santoro et al. [41]    | Low LH and FSH, slightly elevated TSH                  | Previous diagnosis of GPA. At presentation, arthralgia and skin ulcer; 3 months later, fever, cough, and sinusitis; 2 months later, diagnosis of DI                                                                                     |
| Tenorio Jimenez et al. [42] | Secondary hypothyroidism and hypogonadism             | 15-year history of GPA. At presentation, n. VI palsy, headache, and diplopia. Diagnosis of DI                                                                                                                                     |
| Hughes et al. [43]     | Rathypopituitarism                                      | 2008, initial diagnosis with uveitis and scleritis. 2011, polydypsia, polyuria, head ache, and fatigue; diagnosis of DI                                                                                                           |
| Pereira et al. [44]    | Hyperprolactemia, hypothyroidism, probable DI          | Several-year history of GPA. Hyponatremia at presentation; 4 months later, bitemporal superior quadrantanopia; diagnosis of DI                                                                                                       |
| Kapoor et al. [45]     | 7/8 secondary hypogonadism, 6/8 DI, 4/8 secondary hypothyroidism, 1/8 secondary hypothyroidism, 1/8 hyperprolactin, 1/8 hyperprolactin, 1/8 hypopituitarism | 4/8 with DI as presenting symptom, the rest developed after diagnosis of GPA. Latency not mentioned                                                                                                                             |
| Peters et al. [46]     | Hyperprolactin, hypothyroidism, DI                      | ENT symptoms at presentation. Diagnosis of GPA, 1 year later with cranial nerve palsies                                                                                                                                           |
| Ohashi et al. [47]     | Only DI                                                 | 2011, diagnosis of GPA with ENT symptoms. 2012, polydypsia and polyuria; diagnosis of DI                                                                                                                                        |
| Esposito et al. [20]   | Normal ant. pituitary function at diagnosis; 2 years later, secondary hypogonadism and GH deficiency    | Diagnosis of GPA with ENT symptoms. Four months later, polydypsia and polyuria; diagnosis of DI                                                                                                                                     |
|                        | DI, no ant. pituitary deficiency                        | Polydypsia and polyuria. Four months later, ENT symptoms; diagnosis of DI                                                                                                                                                         |
|                        | DI, no ant. pituitary deficiency                        | Sinusitis, otitis media. Two months later, polydypsia and polyuria; diagnosis DI                                                                                                                                                  |
| Xie et al. [48]        | DI, no test of ant. pituitary function mentioned        | Polydipsia and polyuria as presenting sign                                                                                                                                                                                        |
| Byrne et al. [21]      | DI, no test of ant. pituitary function mentioned        | Presentation with nasal congestion and headache. Four months later, FESS due to sinusitis. One year later, polydypsia, polyuria, and diplopia                                                                                           |
| Eli et al. [49]        | DI, hyperprolactin                                      | Presentation with galactorrhea and amenorrhoea, 1.5 years after fatigue and arthralgias and hemoptysis and shortness of breath; 3.5 years after initial presentation, diagnosis of GPA                                                                 |
| Vanderheynst et al. [50]| DI, ant. pituitary function normal                      | Polydipsia and polyuria and chronic sinusitis at presentation; diagnosis of DI                                                                                                                                                     |
| De Parisot et al. [18] | DI in 7/9, 7/9 hypogonadism, 5/9 TSH deficiency, 4/9 hyperprolactin, 2/9 GH deficiency, 1/9 ACTH deficiency | Pituitary disease diagnosed after GPA in 8/9 patients at median of 585 months, concomitant in one case                                                                                                                              |
| Author          | Pituitary function                          | First manifestation and latency                                                                 |
|-----------------|---------------------------------------------|--------------------------------------------------------------------------------------------------|
| Bando et al. [51] | DI, GH deficiency                            | Chronic sinusitis, COM, auditory disturbance when 34 years of age. With 38 years of age; nasal stiffness, fatigue, appetite loss, saddle-nose deformity. At 43, polydipsia and polyuria |
| Sampei et al. [52] | DI, no test of ant. pituitary function mentioned | DI 4 months before admission, headache and right-sided loss of vision and sinusitis at admission |
| Slabu et al. [53]  | DI, hyperprolactemia, hypothyroidism, low GH  | Longstanding diagnosis of GPA, at diagnosis, polydipsia and polyuria                              |
| Van Durme et al. [54] | DI, hypogonadotrophic hyperprolactemia, primary hyperthyroidism | Nausea and vomiting at presentation, history of sinusitis and otitis media, Polyuric syndrome at presentation, 1 year later fever, rash, and arthralgias; 3 years later, ear discharge, decreased hearing, nasal and oral ulcers |
| Dutta et al. [55]   | DI, no ant. pituitary dysfunction             | Nasal blockage, cough, fever, polyuria, and bilateral ear blockage since 5 months; central DI and GPA established |

FSH = Follicle-stimulating hormone, LH = Luteinizing hormone, TSH = Thyroid-stimulating hormone, DI = Diabetes Insipidus, ACTH = Adrenocorticotropic-releasing hormone, IGF = Insulin-like growth factor, GH = Growth hormone
as the cases reviewed, highlight the complexity of this disease. This review supports previous research and suggests that more than 70% of patients with GPA and DI have symptoms in the field of otolaryngology [6]. We found that ENT-related symptoms might occur more often before symptoms of DI. These results might explain why early involvement of ENT specialists was found to result in substantially increased survival [54].

In line with our findings, in many of the cases reviewed, patients experienced ENT symptoms at least 1 year before symptoms of DI occurred. Sinonasal disease is the most common manifestation in the head and neck area, but other symptoms occur frequently. The otolaryngologist has to manage acute and chronic symptoms, so knowledge about the different forms of the disease is fundamental.

The specific enlargement of the pituitary infundibulum on MRI is the result of involvement of the stalk and the hypothalamus in patients with DI [55, 56]. These images allow for distinction from other pathologies affecting the gland. While panhypopituitarism occurred in only a few patients, our study suggests that anterior pituitary dysfunction is slightly more common than anticipated before [27, 29]. Even with these numbers, it may be possible that gland dysfunction is underdiagnosed due to the blunted response when treated with corticosteroids, which most patients with GPA receive during the course of their disease. Hyperprolactemia, hypothyroidism, and hypogonadism are the most common abnormalities in our study. Care by the attending physician in regard to possible dysfunction of pituitary gland function is important, especially because several patients developed DI years after diagnosis of GPA. If polydipsia and polyuria are present, tests for anterior pituitary gland function must be performed.

**Conclusion**

This case demonstrates a relatively rare occurrence of DI in a patient with GPA and demonstrates the difficulty of diagnosing the disease. All healthcare professionals involved in the diagnostic process of the disease must have knowledge about its possible variable course. This is especially important since delayed diagnosis can lead to significant morbidity and possibly mortality, while appropriate treatment options exist [57].

**Abbreviations**

GPA: Granulomatosis with polyangiitis; ANCA: Antineutrophil cytoplasmic antibodies; ENT: Ear–nose–throat; PR3-ANCA: Anti-proteinase 3 antineutrophil cytoplasmic antibodies; MPO-ANCA: Anti-myeloperoxidase antineutrophil cytoplasmic antibodies; AVP: Arginine vasopressin; ER: Emergency room; HB: House–Brackmann score; CT: Computed tomography; IGF: Insulin-like growth factor; LH: Luteinizing hormone; FSH: Follicle-stimulating hormone; T3: Triiodothyronine; T4: Thyroxine; ACTH: Adrenocorticotropin; DI: Diabetes insipidus.

**Supplementary Information**

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**Additional file 1.** Flowchart.

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**Author contributions**

Conceptualization: LK, SD; Data curation: LK; Formal analysis: UE, LK; Investigation: SD, LK; Methodology: LK; Project administration: LK; Supervision: SD, HO, SZ; Validation: UE, LK; Visualization: LK; Writing of original draft: LK, UE; Writing, review and editing: SD, HO. All authors read and approved the final manuscript.

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**Availability of data and materials**

All data generated or analyzed during this study are included in this published article and its additional information files.

**Declarations**

**Ethics approval and consent to participate**

Written consent was obtained from the subject mentioned in this case and is available upon request; ethics approval is not applicable to this type of study in Germany.

**Consent for publication**

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

**Competing interests**

The authors declare that they have no competing interests.

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