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Case Report

Granulomatosis with polyangiitis with lacrimal gland enlargement and pancreatic swelling: A case report and a literature review

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ABSTRACT

A 62-year-old man had bilateral eyelid swelling for 4 months. Two months before admission, he developed fatigue and lost 5 kg of body weight. Further examination revealed elevated serum C-reactive protein, normal angiotensin-converting enzyme, elevated proteinase-3 antineutrophil cytoplasmic antibody (PR3-ANCA), and normal immunoglobulin (Ig)G4 concentration. Chest X-ray and computed tomography showed no enlarged hilar lymph nodes, but positron emission tomography-computed tomography showed fluorodeoxyglucose accumulation in both lacrimal glands, in lung nodules, and in the pancreas. Tissue biopsies of the lacrimal glands and pulmonary nodules showed granuloma with giant cells, but no IgG4-positive cells or fibrosis. Pancreatic tissue showed no findings of autoimmune pancreatitis. In the 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for granulomatosis with polyangiitis, the total score was 10 points. Final comprehensive diagnosis was granulomatosis with polyangiitis, based on the negative results of differential diseases, such as IgG4-related diseases and sarcoidosis. Prednisolone 60 mg/day was started on Day 8, and rituximab 500 mg/body/week on Day 12. After beginning treatment, general malaise and lacrimal gland enlargement were resolved, PR3-ANCA and C-reactive protein became negative, and the nodular shadow in the lungs disappeared. This is the first report of granulomatosis with polyangiitis presenting both lacrimal gland and pancreatic lesions.

KEYWORDS: Granulomatosis with polyangiitis; pancreatic enlargement; lacrimal gland enlargement

Introduction

Granulomatosis with polyangiitis (GPA) is an antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV). It is an autoimmune disease characterised by inflammation of small blood vessels leading to multiple organ damage. GPA typically results in necrotising granulomatosis in the ear, nose, upper and lower respiratory tracts, necrotising glomerulonephritis, and necrotising vasculitis of small- and medium-sized blood vessels. Pancreatic and lacrimal gland lesions are uncommon in GPA [1]. However, there have been cases of AAV with either pancreatic or lacrimal gland involvement [2–32]. Of the reported cases of AAV diseases with pancreatic lesions, GPA was the most common, followed by microscopic polyangiitis (MPA), and there have also been reports of eosinophilic granulomatosis with polyangiitis (EGPA) [2–20]. Similarly, GPA was the most common among the reported cases of AAV diseases with lacrimal gland lesions, and there have also been reports of EGPA [21–32]. Here, to the best of our knowledge, we present the first known case of AAV presenting with both pancreatic and lacrimal gland lesions at the same time. The similar symptoms between this and IgG4-related diseases (IgG4-RD) require careful differentiation.

Case presentation

A 62-year-old Japanese man had bilateral eyelid swelling and visited our Department of Plastic Surgery. Biopsies of the lacrimal glands were performed, and a granuloma was found. Two months later, he developed fatigue and his weight decreased from 67 to 62 kg. Further examination revealed elevated serum C-reactive protein (CRP, 3.85 mg/dl), normal eosinophil count (180 cells/μl, 2.2% of white blood cell), elevated erythrocyte sedimentation rate (20 mm/h), normal urinary findings (protein, red blood cells 0–1/HPF, cast 0–1/LPF), normal creatinine (0.99 mg/dl), normal KL-6 (304 U/ml), normal angiotensin-converting enzyme (ACE, 10 U/l), normal immunoglobulin (Ig)G4 (87 mg/dl), and elevated proteinase-3 anti-neutrophil cytoplasmic antibody [PR3-ANCA, 4.5 IU/ml (reference range <2.0)]. Positron emission tomography-computed tomography (PET-CT) showed abnormal accumulation of fluorodeoxyglucose (FDG) in bilateral lacrimal

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glands, pulmonary nodules, and the pancreas. Bronchoscopic biopsy of the pulmonary nodules also showed granuloma. The patient was admitted to our Department of Plastic Surgery. On admission, he had general malaise, abdominal pain, and dry eyes and mouth as sicca symptoms, there was no peripheral neuropathy or hearing loss. On physical examination, he had a temperature of 36.9°C, blood pressure of 155/61 mmHg, SpO_2 of 95% (room air), mild hypoxaemia, bilateral lacrimal gland enlargement (Figure 1(a,b)), and there was a slight fine crackle in the right lower lung field. PR-3 ANCA was positive and CRP was mildly elevated. ACE and IgG4 were within reference range. Chest X-ray revealed a cardiothoracic ratio of 46.4% and a nodular shadow at the right costophrenic angle. Electrocardiography and echocardiography were normal. Computed tomography (CT) showed thickened mucosa in the left maxillary sinus (Figure 1(c)), multiple nodules in both lungs, a diffusely enlarged pancreas with patchy areas of poor contrast, and enlarged bilateral lacrimal glands (Figure 1(d)), and PET-CT scan showed abnormal accumulation of FDG in bilateral lacrimal glands, the right lung, and the pancreas, with the maximum standardised uptake value of 4.76 in the lacrimal glands (Figure 1(e)), 7.65 in the right lung (Figure 1(f)), and 8.71 in the pancreas (Figure 1(g)). Tissue biopsies were also performed on the lacrimal glands, the pancreas, and lung nodules. Lacrimal gland biopsy showed scattered granuloma formation with giant cell infiltration, and inflammatory cell infiltration of predominantly plasma cells (Figure 1(h)), while almost no IgG4 positive cells were found (data not shown). Histological findings from the pulmonary nodules showed fibrotic thickening of the alveolar wall, infiltration of inflammatory cells of mainly lymphocytes and neutrophils, and enlarged alveolar epithelium. The alveolar space was filled with foam cells and histiocytes. There was an inflammation with granulomas, but no evidence of vasculitis (Figure 1(i)). Similarly, few IgG4-positive cells were found (data not shown). Pancreatic biopsy by endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) showed mild atrophy of the pancreatic acinus, mild inflammatory cell infiltration of few plasma cells, and no specific findings, such as autoimmune pancreatitis (Figure 1(j)). There were no malignant findings in any of the organs.

The patient met the 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for granulomatosis with polyangiitis (GPA) with a total of 10 points; 5 points for PR-3 ANCA positivity; 2 points for chest imaging showing nodules, masses, and cavities; 2 points for biopsy showing granuloma; and 1 point for imaging showing inflammatory and infiltrative shadows in the paranasal sinuses. Histological findings included both enlarged lacrimal glands and the pancreas containing granulomatous lesions. IgG4-RD, other autoimmune diseases such as sarcoidosis, and infectious diseases such as mycobacteria infections were considered as differential diseases. IgG4-RD was considered to be negative because there was no increase in IgG4-positive cells in serum and tissue. Sarcoidosis was considered to be negative because ACE was normal, and there was no lymphadenopathy including in the hilar region of the lungs on chest X-ray and CT, although it was difficult to completely rule it out based on pathological findings. Mycobacteria infections were considered to be negative because of negative cultures and the interferon-gamma release assays, and there were no caseating granuloma. Based on these results, we diagnosed the present case as GPA. On the 8th day, we started 60 mg/day of daily prednisolone, and rituximab (RTX) 500 mg/body/week was started on the 12th day of admission. RTX was administered three times in total. After the start of treatment, general fatigue and lacrimal gland enlargement disappeared, PR3-ANCA and CRP became negative, nodular shadows in the lung disappeared, and pancreatic enlargement improved (Figure 2(a–f)). In addition, at admission, the Birmingham Vasculitis Activity Score V.3 was 9 points, all new/worse disease: 2 points for weight loss, 4 points for adnexal inflammation, and 3 points for nodules or cavities. After treatment, the score on Day 126 was markedly improved: 0 point. The Vasculitis Damage Index scores at admission and on Day 126 were both 0 point. Over the last 18 months, the patient has continued to improve, except for occasional sinusitis, and the dose of prednisolone has been reduced to 2 mg/day.

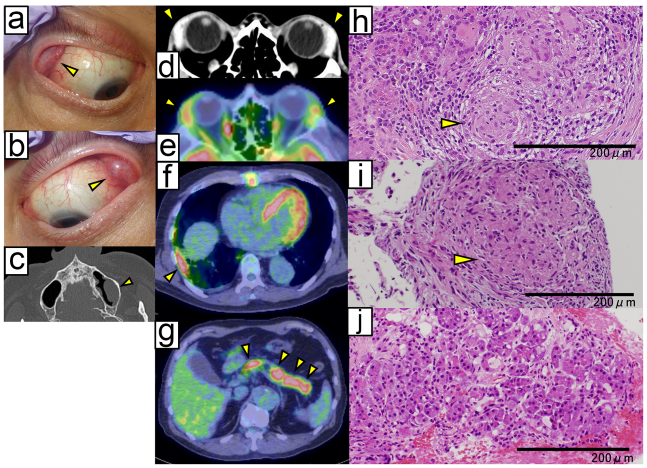

Figure 1. Physical findings, image, and pathological histopathology of this case.

(a, b) Enlargement of both lacrimal grands was found 4 months before admission. (c) Mucous thickening of the left maxillary sinus was found on CT at admission. (d–g) Contrast-enhanced CT and PET-CT were taken 2 months before admission. Both lacrimal glands were swollen (d). FDG was accumulated to the lacrimal grands (e), the lungs (f), and the pancreas (g), and each maximum standardised uptake value was 4.76, 7.65, and 8.71, respectively. (h–j) Pathological findings of the lacrimal gland, the lungs, and the pancreas. There were no malignant findings in any of the tissues. Lacrimal gland (h): HE-staining; formation of granuloma with giant cell infiltration was occasionally observed. Infiltration of inflammatory cells, mainly plasma cells, were observed. No vasculitis was observed. Lung (i): HE-staining; alveolar walls were fibrous and thickened, and inflammatory cells, mainly lymphocytes and neutrophils, infiltrated. The alveolar epithelium was swollen. Foam cells and histiocytes were concentrated in the alveolar space. There was no vasculitis. Inflammatory images with granulomas were observed. Pancreas (j): HE-staining; pancreatic acini were mildly atrophic. Inflammatory cells were mildly infiltrated, but plasma cells were unremarkable. There was no evidence of autoimmune pancreatitis.

Discussion

Our patient had GPA with both pancreatic and lacrimal gland lesions. Pancreatic lesions with GPA are an important organ lesion that may directly relate to life prognosis, since deaths can occur from acute pancreatitis. Conversely, although lacrimal gland lesions are not directly related to life prognosis, we suggest they are important from the view-

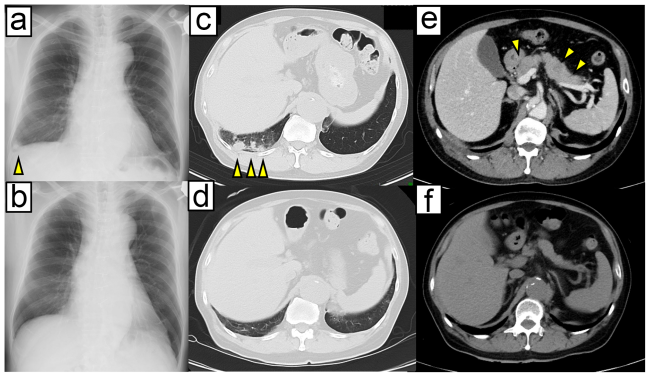

Figure 2. Changes of imaging before and after treatment.

(a, b) Chest x-ray at admission (a) and on Day 126 (b). The nodular shadow was markedly improved. (c, d) CT of the lungs showing nodules on Day 4 (c) and the nodules had disappeared on Day 69 (d). (e, f) CT of the pancreas 2 months before admission (e) and on Day 69 (f). Pancreatitis was improved.

point of early detection of GPA. We searched the literature for reports of other cases of AAV with pancreatic and/or lacrimal gland involvement.

The 20 cases from 19 reports of AAV with pancreatic involvement (12 cases of GPA, 5 cases of MPA, 1 case of EGPA, and 2 cases diagnosed merely as AAV) are shown in Table 1. Of the 19 cases described with imaging findings, 13 cases had masses, 12 cases had pancreatic enlargement, 3 cases had cystic lesions, and 5 cases had obstruction or dilation of the main pancreatic duct or common bile duct (findings were overlapping in some cases) [2–15, 17–20]. Needle biopsy for pancreatic lesions was attempted in seven cases [2, 3, 7, 8, 12, 14, 19]. Vasculitis on needle biopsy was reported in one case [2], five cases did not have specific results [3, 8, 12, 14, 19], and one case was not successfully biopsied [7]. In one case in which specific results were not found by needle biopsy, vasculitis was confirmed after pancreatic resection [12]. On the other hand, vasculitis was found without needle biopsy in four cases after pancreatic resection [4, 11, 15], and in two cases in postmortem autopsies [16, 17]. Therefore, in consideration of accurate diagnosis, needle biopsy is thought to be not so sensitive, and surgical resection is thought to have better sensitivity. Two cases with pancreatectomy were negative for ANCA, but were suspected of pancreatic cancer because of the pancreatic masses, and there were no AAV symptoms outside the pancreas [11, 15]. Of the 20 cases, 16 were treated with immunosuppressive therapy [2–7, 9, 10, 12–16, 18–20]. Of the 12 cases that had description about the effect of immunosuppressive treatment on pancreatic lesions, 8 cases showed reduction or remission of pancreatic lesions [2, 6, 7, 10, 13, 14, 19, 20]. In four cases of acute pancreatitis, there was relapse in one case and three cases became worse, and two of the patients died of sepsis or disseminated intravascular coagulation [3, 9, 16, 18]. In one case, the pancreatic lesion disappeared without treatment [8].

Regarding treatment for pancreatic lesions, intravenous cyclophosphamide was used in two cases, both of which had resolution of pancreatic lesions [7, 13]. Oral cyclophosphamide was used in two cases; pancreatic lesions were resolved in one [10] and there was remission in the other, although there was no clear description about pancreatic lesions [14]. Of the four other cases treated with cyclophosphamide without any description of the route of treatment (orally or intravenously), one showed almost complete remission of pancreatic lesions [19], two cases showed the appearance of pancreatic lesion after cyclophosphamide treatment [3, 18], and one case showed worsening of renal lesions but no treatment effect was described for pancreatic lesions [5]. RTX was used in three cases [3, 5, 6]. One case was treated with prednisolone 1 mg/kg/day and cyclophosphamide, but developed symptoms of pancreatic lesions, and subsequently died of sepsis, despite twice being treated with RTX 1000 mg [3]. One case was treated with prednisolone 250 mg/day, but pulmonary lesions worsened, and cyclophosphamide worsened renal lesions, but RTX decreased PR3-ANCA [5]. One patient was a young female so was treated with RTX instead of cyclophosphamide during steroid tapering, but she developed bilateral otitis media [6].

The 12 cases of AAV with lacrimal gland lesions (10 cases of GPA and 2 cases of EGPA) are shown in Table 2. Lacrimal gland lesions were bilateral in seven cases and unilateral in five cases. In all except one case, lesions other than periorbital lesions were observed [21–32]. Lacrimal gland biopsies were performed for nine patients, eight of which showed AAV-specific findings such as vasculitis or granulomas, and one showed no specific findings [21–23, 25–29, 31]. Immunosuppressive therapy was used in 11 patients, 10 of which showed improvement of lacrimal gland lesions, but 1 patient did not show improvement of lacrimal gland enlargement [21–28, 30–32]. In addition, among the reported cases of AAV with lacrimal gland enlargement, there were also cases in which IgG4-RD was thought to be complicated with GPA or EGPA [33, 34]. Therefore, in order to differentiate whether the lacrimal gland lesion is due to AAV or a complication of IgG4-RD, it is necessary to perform both biopsy and blood examinations.

In our case, the patient's pancreatic lesion was enlarged with low contrast areas, and needle biopsy did not find specific tissue for AAV. After treatment with RTX in addition to prednisolone, the pancreatic enlargement improved (Figure 2). These features were common with previous reported cases of pancreatic lesions in GPA. It is very important to distinguish lacrimal gland lesions from IgG4-RD. On pathological examination of our patient, the peripheral blood IgG4 level was normal, and the number of IgG4-positive cells was negligible, so we could clearly rule out IgG4-RD. There have been no previous reports of AAV with both pancreatic and lacrimal gland lesions, so our case is considered to be very rare.

Completely distinguishing this condition from sarcoidosis is considered to be very difficult. However, sarcoidosis could ultimately be denied because the ACE was normal: there was no lymphadenopathy including in the hilar region on chest X-ray or CT. Further accumulation of cases is needed.

In conclusion, this is the first report of GPA presenting both lacrimal gland and pancreatic lesions. RTX was effective for GPA with both lacrimal gland and pancreatic lesions.

Table 1. Cases of AAV with pancreatic lesions.

Case	Author (year)	Year/sex	Pancreatic lesion	Other organ lesions	Pathology of pancreas	Treatment	Effectiveness of treatment	ANCA	Diagnosis
Our case		62/M	Swelling, low-contrast area	Nose, lung, and lacrimal gland	Mild atrophy pancreatic acini, mild inflammatory cell infiltration (FNA)	GC and RTX	+	PR3-ANCA	GPA
1	Yamada *et al.* (2022)	67/M	Low-density mass and swelling	Lung and kidney	Irregularly ruptured smooth muscle of vascular wall in small-size-arteries (FNA)	GC and AZ	+	MPO-ANCA	AAV
2	Alesaeidi *et al.* (2021)	38/F	Mass and acute pancreatitis	Nose	Pancreatitis pattern with no malignant cell (FNA)	GC, CY, and RTX	–	ND	GPA
3	Marvisi *et al.* (2021)	62/M	Pseudocystic lesion	Asthma, nose, and peripheral nerve	Focal destruction of elastin layers within isolated vessels with a mild increase in eosinophils (operation)	CY	ND	MPO-ANCA	EGPA
4	Garbe *et al.* (2021)	49/F	Swelling and pancreatitis	Lung, kidney, peripheral nerve, joint, skin, and parotid gland	ND	GC, CY, RTX, and PE	ND	PR3-ANCA	GPA
5	Sowida (2019)	22/F	Low-density change and fluid-filled cyst	Nose, ear, mouth, and lung	ND	GC, AZ, and RTX	+	PR3/C-ANCA	GPA
6	Suzuki *et at.* (2019)	71/M	Swelling, dilatation of the main pancreatic duct	Lung and kidney	FNA was unsuccessful	GC and IVCY	+	MPO-ANCA	MPA
7	Iida *et at.* (2016)	72/M	Swelling, masses, and poorly enhanced area	Lung, kidney	Nonspecific pancreatitis (FNA)	Not used	ND	MPO-ANCA	MPA
8	Iida *et at.* (2015)	64/F	Swelling and acute pancreatitis	Kidney	ND	GC	–	MPO-ANCA	MPA
9	De Bie *et al.* (2015)	57/M	Mass and stenosis of the distal common bile duct	Nose, kidney, and eye	ND	GC, CY, and PE	+	MPO-ANCA	AAV
10	Kontis *et al.* (2014)	57/M	Mass	None	Necrotic mass with a palisading granuloma and vasculitis (operation)	Not used	ND		GPA
11	Kontis *et al.* (2014)	68/F	Uncinate process, mass, and jaundice	None	Necrotic masses with granulomatous inflammation and vasculitis (operation)	Not used	ND	MPO/P-ANCA	GPA

(continued)

Table 1. (Continued)

Case	Author (year)	Year/sex	Pancreatic lesion	Other organ lesions	Pathology of pancreas	Treatment	Effectiveness of treatment	ANCA	Diagnosis
12	Valerieva *et al.* (2013)	62/F	Swelling and mass	Nose and ear	Detritus without atypical cells (FNA), vasculitis with fibrinoid necrosis, granulomas and giant cells (operation)	GC and AZ	ND	PR3-ANCA	GPA
13	Hamilton *et al.* (2011)	78/F	Mass	Nose, lung, kidney, skin, and joint	ND	GC and IVCY	+	PR3/C-ANCA	GPA
14	Chawla *et al.* (2011)	60/F	Swelling, hypoattenuated lesion	Nose, lung, and kidney	Inflammatory cells with no evidence of malignancy (FNA)	GC, CY, and AZ	+	PR3-ANCA	GPA
15	Tinazzi *et al.* (2007)	48/F	Mass and obstruction of the main pancreatic duct	None	Granulomatous inflammation with necrosis and destruction of the wall of small-medium size blood vessels (operation)	GC, CY, and MTX	ND		GPA
16	Haraguchi *et al.* (2005)	84/F	ND	Lung, kidney, and brain	Fatty necrosis of the pancreatic tissue due to vasculitis (autopsy)	GC	–	MPO-ANCA	MPA
17	Matsubayashi *et al.* (2004)	65/M	Swelling, acute pancreatitis, and low-density lesions	Ear, lung, kidney, and spleen	Diffuse necrotising pancreatitis, vasculitis (autopsy)	Not used	ND	PR3-ANCA	GPA
18	Berney *et al.* (1997)	32/M	First: acute pancreatitis. Second: oedematous pancreatitis and enlargement of pancreatic duct	Kidney	ND	GC and CY	–	C-ANCA	MPA
19	O'Neil *et al.* (1992)	62/M	Swelling, mass, type I DM, and dilated common bile duct	Nose, ear, lung, kidney, thyroid, heart, and peripheral nerve	ND (FNA)	GC and CY	+		GPA
20	Kemp *et al.* (1990)	57/M	Acute pancreatitis and cyst	Nose, ear, pharynx, lung, kidney, and skin	ND	GC and CY	+	ND	GPA

DM, diabetes mellitus; GC, glucocorticoid; CY, cyclophosphamide; IVCY, intravenous cyclophosphamide; AZ, azathioprine; MTX, methotrexate; PE, plasmapheresis; ND, not described.

Table 2. Cases of AAV with lacrimal gland lesion.

Case	Author (year)	Year/sex	Lacrimal gland lesion	Other organ lesions outside of orbit	Pathology of lacrimal gland	Treatment	Effectiveness of treatment	ANCA	Diagnosis
Our case		62/M	Bilateral swelling	Nose, lung, and pancreas	Granuloma formation with giant cell infiltration and inflammatory cell infiltration mainly consisting of plasma cells	GC and RTX	+	PR3-ANCA	GPA
21	Spadaro *et al.* (2022)	14/F	Bilateral swelling and unilateral mass	None	Acute-on-chronic inflammation with destruction of normal glandular structures	GC and MTX	+	MPO/P-ANCA	GPA
22	Morros *et al.* (2017)	56/M	Unilateral swelling	Kidney	Perivascular granulomatous inflammation with necrotic foci	GC and CY	+	PR3-ANCA	GPA
23	Al-Hakami *et al.* (2016)	57/F	Unilateral soft tissue thickening of orbit	Ear and joint	Angiocentric necrotising granulomatous vasculitis with extensive eosinophilic infiltrates	GC and RTX	+	PR3/C-ANCA	GPA
24	Hanioka *et al.* (2012)	72/M	Bilateral swelling	Asthma, nose, peripheral nerve, skin, and joint	ND	GC and IG	+	MPO-ANCA	EGPA
25	Khanna *et al.* (2011)	27/M	Bilateral swelling	Nose and lung	Fibro-areolar tissue with necrotising granulomatous vasculitis	GC and CY	+	PR3/C-ANCA	GPA
26	Khandwala *et al.* (2010)	27/F	Unilateral swelling	Asthma, nose, and skin	Eosinophilic granulomatous inflammation with giant cells	GC	+	P-ANCA	EGPA
27	Hello *et al.* (2010)	62/F	Bilateral palpebral xanthelasma with infiltrated lacrimal glands	Nose, kidney, peripheral nerve, skin, and joint	Necrotising and granulomatous angiitis	GC, CY, and AZ	+	PR3-ANCA	GPA
28	Martínez-Gutiérrez *et al.* (2008)	7/F	Bilateral infiltration and swelling	Nose, larynx, lung, and kidney	Lymphoplasmacytoid infiltrate with perivascular granulomas	GC, CY, TNF-α, and IFX	+		GPA

(continued)

Table 2. (Continued)

Case	Author (year)	Year/sex	Lacrimal gland lesion	Other organ lesions outside of orbit	Pathology of lacrimal gland	Treatment	Effectiveness of treatment	ANCA	Diagnosis
29	Kiratli *et al.* (2008)	80/M	Unilateral swelling	Lung and kidney	Vasculitis characterised by fibrinoid necrosis and inflammatory infiltrate in the wall of a small artery, granulomatous inflammation destroying the lacrimal gland containing prominent giant cells	ND	ND	ND	GPA
30	Valmaggia *et al.* (2001)	26/F	Unilateral homogeneous tumour	Lung and kidney	ND	GC and CY	+	PR3/C-ANCA	GPA
31	Lanza *et al.* (1995)	30/M	Bilateral swelling	Nose and lung	Necrotising granulomatosis inflammation with focal inflammation of small vessels within glandular tissue	GC and CY	–		GPA
32	Boukes *et al.* (1985)	23/M	Bilateral swelling	Nose and kidney	ND	GC and CY	+	ND	GPA

GC, glucocorticoid; CY, cyclophosphamide; AZ, azathioprine; MTX, methotrexate; IFX, infliximab; TNF-α, tumour necrosis factor alpha; ND, not described.

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Conflict of interest

None declared.

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None declared.

Patient consent

Written informed consent for the publication of this report was obtained from the patient by the corresponding author.

Ethical approval

Ethical approval is not applicable.

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