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ABSTRACT

Objectives: Gluten sensitivity can engender neurologic dysfunction, one of the two commonest presentations being peripheral neuropathy. The commonest type of neuropathy seen in the context of gluten sensitivity is sensorimotor axonal. We report 17 patients with sensory ganglionopathy associated with gluten sensitivity.

Methods: This is a retrospective observational case series of 17 patients with sensory ganglionopathy and gluten sensitivity. All patients had been followed up for a number of years in dedicated gluten sensitivity/neurology and neuropathy clinics.

Results: Out of a total of 409 patients with different types of peripheral neuropathies, 53 (13%) had clinical and neurophysiologic evidence of sensory ganglionopathy. Out of these 53 patients, 17 (32%) had serologic evidence of gluten sensitivity. The mean age of those with gluten sensitivity was 67 years and the mean age at onset was 58 years. Seven of those with serologic evidence of gluten sensitivity had enteropathy on biopsy. Fifteen patients went on a gluten-free diet, resulting in stabilization of the neuropathy in 11. The remaining 4 had poor adherence to the diet and progressed, as did the 2 patients who did not opt for dietary treatment. Autopsy tissue from 3 patients demonstrated inflammation in the dorsal root ganglia with degeneration of the posterior columns of the spinal cord.

Conclusions: Sensory ganglionopathy can be a manifestation of gluten sensitivity and may respond to a strict gluten-free diet. *Neurology*® 2010;75:1003–1008

GLOSSARY

PNS = paraneoplastic neurologic syndrome; **SG** = sensory ganglionopathy.

Sensory ganglionopathy (SG), also known as sensory neuronopathy or sensory neuron disease, is an asymmetric exclusively sensory neuropathy with some well-recognized pathologic associations. SG can be a manifestation of Sjögren syndrome.[1] It is also one of the classic paraneoplastic neurologic syndromes (PNS).[2] The association of SG with PNS and Sjögren syndrome implies an immune-mediated pathogenesis at least in the context of these diseases. However, it can also be a feature of some genetic diseases such as Friedreich ataxia as well as hereditary sensory and autonomic neuropathy.[3]

Gluten sensitivity is a systemic autoimmune disease triggered by the ingestion of gluten in genetically susceptible individuals. Neurologic dysfunction can be the presenting feature even in the absence of an enteropathy.[4]

The 2 commonest neurologic manifestations are ataxia (gluten ataxia) and neuropathy (gluten neuropathy).[4] The commonest type of neuropathy seen in the context of gluten sensitivity is symmetric sensorimotor axonal peripheral neuropathy.[5] Mononeuropathy multiplex,[6,7] small fiber neuropathy,[8] autonomic neuropathy,[9] and pure motor neuropathy[7,10] have also been described in the context of gluten sensitivity but are much less common. We have previously reported the clinical, neurophysiologic, and pathologic characteristics of patients with gluten sensitivity and peripheral neuropathy.[5] We have also reported the effect of gluten-free diet in

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patients with symmetric sensorimotor axonal neuropathy.[11] In this report, we describe our experience with the prevalence, clinical characteristics, and treatment of SG in patients with gluten sensitivity.

METHODS **Standard protocol approvals, registrations, and patient consents.** This is a retrospective observational case series of patients regularly attending the gluten sensitivity/neurology and peripheral neuropathy clinics run by 2 of the authors. The South Yorkshire Research Ethics Committee has confirmed that no ethical approval is indicated given that a gluten-free diet is a recognized treatment and that all investigations/interventions were clinically indicated and did not form part of a research study.

Patients. Patients with chronic neuropathies attend a dedicated neurology clinic based at the Department of Neurology, Royal Hallamshire Hospital, Sheffield, UK, established for 35 years. All patients are routinely screened for gluten sensitivity using estimation of circulating antigliadin (immunoglobulin G and immunoglobulin A), tissue transglutaminase, and endomysium antibodies. Those patients who have significant titers (as defined by the kit manufacturer) of one or more of these antibodies undergo duodenal biopsy to look for any evidence of enteropathy and are followed up in the dedicated gluten sensitivity/neurology clinic. It is our normal clinical practice to advise patients with peripheral neuropathy and serologic evidence of gluten sensitivity, in the absence of any other etiologic cause for the neuropathy, to adhere to a gluten-free diet irrespective of the presence or not of an enteropathy. This practice is based on previous studies that have demonstrated an increased prevalence of gluten sensitivity in patients with neuropathies, and improvement of the neuropathy with strict adherence to a gluten-free diet in patients with sensorimotor axonal neuropathy,[11] gluten ataxia,[12] and gluten myopathy.[13] All patients are followed up on a 6 monthly basis. Repeat neurophysiologic assessments were obtained if clinically indicated.

Neurophysiologic assessments. The diagnosis of SG was made if the patients had clinical signs and symptoms of patchy sensory loss with or without sensory ataxia (sensory ataxia with normal cerebellar imaging) which was supported by electrophysiologic evidence of patchy non-length-dependent sensory fiber involvement with minimal or no motor involvement.[14] Most of the neurophysiologic assessments were performed by one of the authors who is a consultant clinical neurophysiologist (D.G.R.). The same neurophysiologist reviewed all other neurophysiologic assessments performed in all patients included in this report to ensure that these were in keeping with the neurophysiologic features of SG.[14]

RESULTS **Prevalence.** Out of a total of 409 patients with different types of peripheral neuropathies that had been seen and assessed in the dedicated neuropathy clinic, 53 (13%) had clinical and neurophysiological evidence of SG. Out of these 53 patients, 17 (32%) had serologic evidence of gluten sensitivity, 11 (21%) had paraneoplastic syndrome, 10 (19%) had Sjögren syndrome (clinical, serologic, and biopsy evidence), and no etiology despite extensive investigations (idiopathic) was seen in the remaining 15 (28%). Table 1 summarizes the different types of neuropathies. All patients would have had at least 1 neurophysiologic assessment. The prevalence of SG among patients with gluten neuropathy was 8%.[4]

Table 1 Types of chronic neuropathies (excluding patients with a family history and patients with positive genetic testing for familial neuropathies) in 409 adult patients attending a long-established neurology clinic at the Department of Neurology, Sheffield, UK

Types of chronic neuropathies	No. (%)
Symmetric sensorimotor axonal neuropathy	237 (58)
Sensory ganglionopathy	53 (13)
Mononeuropathy multiplex	52 (13)
Small fiber neuropathy	24 (6)
Pure motor axonal neuropathy	24 (6)
Multifocal motor neuropathy with conduction block	8 (2)
Chronic inflammatory demyelinating polyneuropathy	6 (1)
Multifocal acquired demyelinating sensory/motor neuropathy	5 (1)

Clinical characteristics of patients with SG and gluten sensitivity. The mean age of the 17 patients with SG and gluten sensitivity was 67 years (range 47–85). The mean age at onset of the sensory symptoms was 58 years (range 40–80). This is slightly more than the age at onset of sensorimotor axonal neuropathy due to gluten sensitivity, which was 55 years.[4] All 17 patients had serologic evidence of gluten sensitivity. Table 2 summarizes the serologic and histologic findings in these 17 patients. There were 5 male and 12 female patients. Enteropathy on biopsy was seen in 7 of the 17 patients. The severity of the neuronopathy varied from minor sensory symptoms with mild sensory ataxia to marked sensory ataxia causing severe disability necessitating the use of walking aids. None of the patients were wheelchair-bound. The most severely affected patient uses 2 crutches to walk. The disease duration in this patient was 20 years. The majority of these patients[11] had undergone CNS imaging with MRI to exclude cerebellar involvement (common in the context of gluten ataxia). The most severely affected patient also had magnetic resonance spectroscopy of the cerebellum, which had normal results, confirming that the ataxia was peripheral in origin.

Neurophysiologic characteristics of patients with SG and gluten sensitivity. All patients had either absent or attenuated sensory potentials at the time of diagnosis. Ten of the 17 patients showed asymmetric sensory fiber involvement either in a non-length-

Table 2 Serologic characteristics and duodenal biopsy results of 17 patients with SG and gluten sensitivity

Patient	Age at onset, y/sex	Antigliadin IgA/IgG	EMA	tTG	Enteropathy on biopsy	Clinical severity[a]
1	68/M	−/+	−	−	No	Moderate
2	46/M	−/+	−	−	No	Moderate
3	60/F	−/+	N/A	N/A	Yes	Severe
4	48/F	−/+	−	−	No	Mild
5	70/F	+/+	+	>300	Yes	Moderate
6	56/F	−/+	−	−	No	Mild
7	66/F	+/−	−	−	No	Mild
8	56/F	−/+	+	100	Yes	Mild
9	60/M	+/−	−	−	No	Mild
10	60/F	+/+	N/A	N/A	Yes	Mild
11	45/M	+/+	N/A	N/A	Yes	Severe
12	40/F	−/+	−	−	No	Moderate
13	55/M	+/−	−	32	No	Mild
14	40/F	−/+	−	−	No	Mild
15	50/F	+/+	N/A	N/A	Yes	Severe
16	80/F	−/+	−	−	No	Moderate
17	77/F	−/+	+	100	Yes	Moderate

Abbreviations: EMA = endomysium antibodies; Ig = immunoglobulin; SG = sensory ganglionopathy; tTG = transglutaminase antibodies.
[a] Mild = sensory symptoms only; moderate = sensory and significant sensory ataxia; severe = needs walking aids.

dependent fashion or patchy nerve involvement. Three of the 17 patients had unrecordable potentials from nerves in both upper and lower limbs. Four patients showed length-dependent sensory fiber involvement but had no motor fiber involvement, hence supporting the diagnosis of SG.

Effect of gluten-free diet. Fifteen of the 17 patients with SG and gluten sensitivity adopted a gluten-free diet. This was followed by stabilization in all but 4 patients (mean duration on gluten-free diet for the 15 patients was 7.5 years, range 1–15). The neuronopathy in the other 2 patients who did not opt for the diet progressed: 1 became increasingly disabled and died of pneumonia 10 years after the diagnosis (table 3; patient 2); the other became progressively more ataxic and remains disabled. Four out of the 15 patients on the diet continued to progress despite adherence to the diet. Of these, 2 stabilized after the introduction of immunosuppressive medication (mycophenolate). Of interest was the finding of small bowel inflammation in both of these 2 patients on repeat duodenal biopsy consistent with ongoing exposure to gluten. While at the time of the repeat biopsy one of these patients had no circulating antigliadin immunoglobulin A and immunoglobulin G, endomysium and anti-TG2 transglutaminase antibodies, circulating anti-TG6 antibodies, were present. These antibodies may be more sensitive and specific for the neurologic manifestations of gluten sensitivity.[4] Sensory studies on this patient showed progressive deterioration of action potential amplitude after many years of dietary exposure to gluten (table 3; patient 1). The other 2 nonresponders were found to have persistently circulating antigliadin antibodies implying poor adherence to the gluten-free diet. In one of them repeat endoscopy showed gastric carcinoma. She subsequently died of the disease. It is therefore possible that the SG in this case was paraneoplastic rather than related to gluten sensitivity.

Pathologic findings. Pathologic data were available on 3 patients (2 on gluten-free diet, 1 not) who underwent postmortem examination. All 3 patients died of unrelated causes.

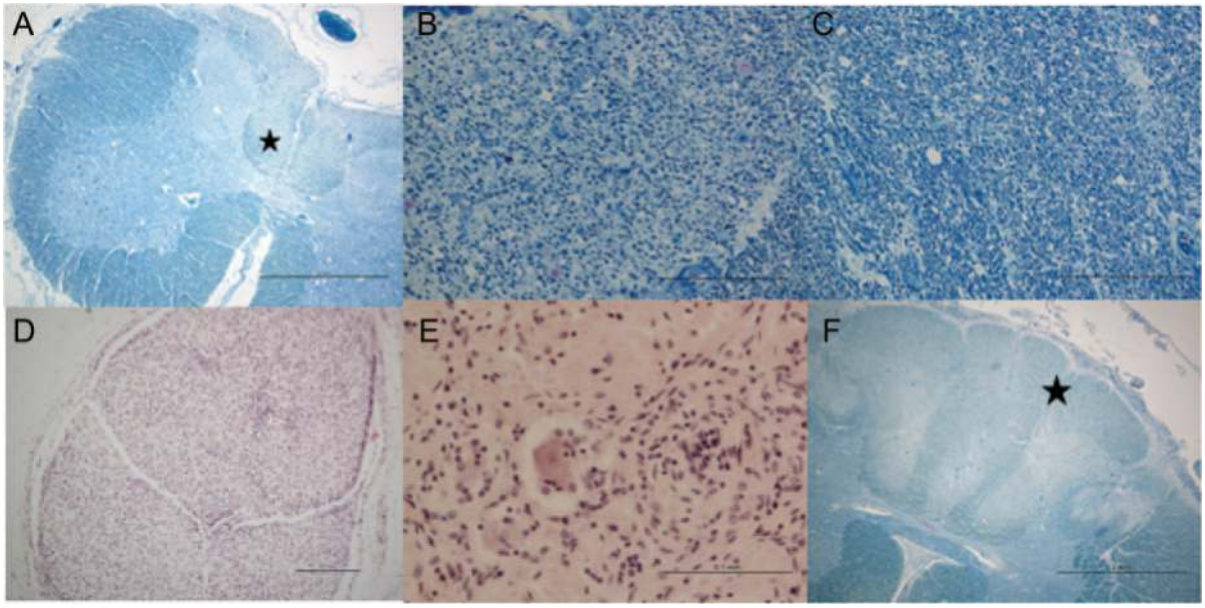
In one of these patients (on gluten-free diet, clinically stable, no enteropathy on biopsy), the spinal cord appeared macroscopically unremarkable. Microscopic examination, however, revealed subtotal loss of myelin from the dorsal columns at all levels of the cord, with preservation of the anterior and lateral columns (figure, A–C). Immunohistochemistry to neurofilament protein showed mild axonal loss in the dorsal columns. No myelin or axonal loss was observed in nerve roots. The neuropathologic findings were those of dorsal column degeneration. Vitamin B12 and copper levels were normal at presentation and imaging of the cord did not reveal any abnormalities. The findings would be consistent with central degeneration resulting from a sensory ganglioneuropathy, although dorsal root ganglia were not available for examination.

Table 3 Serial studies in 2 patients with gluten sensitivity and SG showing progressive reduction of sensory action potential over a number of years[a]

	1989	1990	1996	1997	1998	1999	2001	2005	2008
Patient 1, median SAP, μV	6.9	3.2	2.6	0.9	0.3	NR	NR	NR	NR
Patient 2, median SAP, μV			23.2		14.2	12.2	11.5	1.8	

Abbreviations: NR = not recordable; SAP = sensory action potential; SG = sensory ganglionopathy.
[a] The diagnosis of gluten sensitivity in patient 1 was made in 1996. Despite the introduction of gluten-free diet, there was evidence of poor dietary adherence associated with clinical progression. The patient was eventually treated with immunosuppression with clinical evidence of stabilization. Patient 2 never went on a gluten-free diet.

Figure Neuropathologic findings at postmortem from 3 patients with gluten sensitivity and sensory ganglionopathy (SG)



Myelin stain (combined Luxol fast blue/cresyl violet) of spinal cord at L1 showing pallor of the dorsal column (A). Subtotal myelin loss was seen in the dorsal (B) but not lateral (C) columns (A–C from the same patient, on gluten-free diet and clinically stable, patient died of unrelated cause). Nerve root showing lymphocytic infiltration (D). A lighter, diffuse infiltrate is seen in dorsal root ganglion (E) (D and E from the same patient, on gluten-free diet but positive serology, patient died of pneumonia). Low-power view (F) of cervical cord (combined Luxol fast blue/cresyl violet) showing marked pallor of the dorsal column (star), involving both fasciculus gracilis and cuneatus (F from the third patient, clinically stable on gluten-free diet, patient died of unrelated cause). Magnification bars: A and F, 2 mm; B–D, 0.2 mm; E, 0.1 mm.

The general neuropathologic findings of the second patient (on gluten-free diet but positive circulating antigliadin antibodies and enteropathy) have been reported previously.[5] Spinal nerve roots showed patchy loss of myelin staining and inflammatory infiltrates of lymphocytes and macrophages (figure, D). Dorsal root ganglia also showed a less pronounced, diffuse lymphocytic infiltrate with loss of occasional neurons (figure, E). Sensory ganglioneuropathy in this case was therefore inflammatory in nature and in the context of more widespread inflammatory changes affecting central and peripheral nervous system.

In the third patient (on gluten-free diet, clinically stable, enteropathy on biopsy), the spinal cord showed marked degeneration of the dorsal columns at all levels, with preservation of anterolateral white matter. These changes would be consistent with being secondary to her SG.

DISCUSSION Gluten sensitivity is currently best considered as an autoimmune disease with systemic manifestations. It is one of the very few autoimmune diseases where the etiologic agent is known (gluten). Bowel involvement (clinical or pathologic) is not a prerequisite for the diagnosis. The mechanism of neurologic damage has an immunologic basis. Postmortem data show evidence of inflammatory infiltrates with a perivascular distribution in the central (e.g., cerebellum) and peripheral nervous system[5] and in muscle.[4] Emerging evidence suggests that the involvement of different types of tissue transglutaminases is pivotal to the pathogenesis within the particular organ (bowel, skin, or neural tissue) involved in this disease.[4]

This report highlights SG as a presenting feature of gluten sensitivity in the largest cohort of patients (17) so far reported. A case report published in 2007 reported a patient with SG and celiac disease but the patient also had Sjögren syndrome.[15] In a retrospective study of 23 patients with small fiber neuropathy/ganglionopathy from the United States, one patient was reported as having celiac disease.[16] Another retrospective study from the United States reported 20 patients with celiac disease and neuropathic symptoms, some of whom had purely sensory symptoms suggestive of a neuronopathy. Eighteen of these patients, however, had normal neurophysiology, thus suggesting that they had a small fiber neuropathy/ganglionopathy rather than the type of SG seen in the patients reported here.[17]

The current report suggests that therapeutic intervention in the subgroup of patients with SG who also have gluten sensitivity in the form of strict gluten-free diet is associated with stabilization or even improvement of what is generally a progressive disease. This was not a randomized controlled study but an observational case series based on patients with gluten sensitivity and SG that have been followed up for

a long time. Such observational reports have their limitations. However, as a number of patients did not follow our advice for adherence to a strict gluten-free diet (a common problem in patients with celiac disease), this allowed us to observe a progression of the disease when compared to the stabilization seen in patients on gluten-free diet. The stabilization was only seen in patients on strict gluten-free diet if accompanied by elimination of the serologic markers of gluten sensitivity. Indeed in 2 of the patients with poor adherence to the diet (as indicated by the presence of enteropathy and/or persistently positive serologic tests for gluten sensitivity) there was progression of the SG until the introduction of immunosuppressive treatment. In another patient with persistently positive antibodies and no clinical response to the gluten-free diet, the diagnosis of gastric carcinoma was made eventually on repeat biopsy. Untreated celiac disease is associated with a higher incidence of gastrointestinal cancer. If a patient with SG and gluten sensitivity progresses despite strict gluten-free diet, gastrointestinal malignancy should be considered as an alternative/additional cause.

Clinical stabilization and improvement after introduction of strict gluten-free diet and elimination of serologic markers for gluten sensitivity have been described in patients with gluten ataxia,[12] gluten neuropathy (sensorimotor length dependent axonal neuropathy),[11] small fiber neuropathy/neuronopathy,[8] and gluten myopathy.[13] As in previous treatment studies, stabilization or improvement was independent of the presence of enteropathy but was associated with strict gluten-free diet as evident by the elimination of all serologic markers of gluten sensitivity. This is an important point as some case reports that suggest lack of response to the gluten-free diet do not provide evidence of serologic elimination of the antibodies, which is essential for neurologic stabilization/improvement.[4] While we have observed some improvement of the sensory action potentials in a few patients, our experience suggests that these patients did not recover fully and that a degree of damage was permanent. It is unclear if this relates to irreversible degeneration of the posterior columns of the spinal cord as seen in 2 of our patients with SG and gluten sensitivity who underwent postmortem examination. We assume that the cord involvement is a secondary phenomenon following inflammation/degeneration of the dorsal root ganglia. We have not observed any evidence of inflammation of the cord in the limited postmortem data from 3 patients. Given that SG is not fully reversible it is important to investigate the possibility of gluten sensitivity at presentation as therapeutic intervention with a gluten-free diet may prevent chronic disability.

The literature on therapeutic interventions in other causes of SG, such as paraneoplastic, Sjögren, or idiopathic cases, is limited to just case reports or small series. In paraneoplastic SG cases, the disease tends to be rapidly progressive and can only be arrested by early diagnosis and effective treatment of the underlying cancer. This is frequently impossible partly because the cancer is often occult (in most of our cases the diagnosis of paraneoplastic neuronopathy was made using PET scan[18]), but also because the type of cancer frequently associated with paraneoplastic neuronopathy (small cell lung carcinoma) cannot be effectively treated. Improvement of the SG has, however, has been reported in patients who had undergone successful chemotherapy treatment for Hodgkin lymphoma.[19] There are also single case reports suggesting improvement in SG in patients with Sjögren syndrome using IV immunoglobulins and plasma exchange.[20,21] We have observed improvement of SG in a patient with Sjögren syndrome treated with mycophenolate (unpublished observation). Currently there is insufficient evidence to suggest that a specific or indeed any immunomodulatory therapy can be effective in idiopathic SG. In most patients with idiopathic SG and SG seen in the context of Sjögren syndrome and gluten sensitivity the neuronopathy progresses slowly but eventually causes significant disability.

This report suggests that gluten sensitivity is a common cause of SG and that strict gluten-free diet may be an effective therapeutic intervention at least in arresting the progression. Serologic testing for gluten sensitivity should be part of the workup of all patients with SG.

DISCLOSURE

Dr. Hadjivassiliou and Dr. Rao report no disclosures. Dr. Wharton serves as Editor-in-Chief of *Neuropathology and Applied Neurobiology* and on the editorial board of *Histopathology* and receives research support from the Medical Research Council, Alzheimer's Research Trust, and the British Neuropathological Society. Dr. Sanders reports no disclosures. Dr. Grunewald has received honoraria for educational activities from STAC Consultancy LLP. Dr. Davies-Jones reports no disclosures.

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