

# Polymyalgia rheumatica in a married couple

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**Abstract:** The case of a married couple developing polymyalgia rheumatica (PMR) consecutively is presented. The 55-year-old wife complained in June 2010 about pain in her neck. Case history, physical examination, and erythrocyte sedimentation rate (ESR) of 80 mm/hour led to the diagnosis of PMR. In May 2011, her 66-year old husband complained about pain in his neck, shoulders, buttocks, and thighs. Considering anamnesis, physical examination, and ESR of 56 mm/hour, the diagnosis of PMR was made. Both wife and husband responded to steroid treatment. When the steroid dose was gradually reduced, both patients relapsed. In order to lower the cumulative dose of glucocorticoid therapy, 10 mg methotrexate per week was added. In the literature, six cases of polymyalgia rheumatica in married couples have been described to date. In four cases, polymyalgia rheumatica occurred first in the wife. The interval of the diagnosis between the spouses ranged from 0 to 89 months. Although in most of the previous case reports a genetic disposition and an infectious agent have been discussed, this hypothesis must be questioned.

**Keywords:** husband, wife, erythrocyte sedimentation rate, C-reactive protein, polymyalgia rheumatica

## Introduction

Polymyalgia rheumatica (PMR) is a relatively frequent disease with a prevalence between 13/100,000 and 133/100,000, with a predominance of people aged > 70 years.<sup>1,2</sup> It has been proposed that environmental factors and infectious agents are plausible in the etiology and pathogenesis of PMR.<sup>3</sup> In this context, six case reports have been published to date in which PMR has been diagnosed in married couples.<sup>4-9</sup> Also, there have been some reports describing several cases of PMR within family members such as siblings.<sup>9-15</sup> Based upon these reports, it has been discussed that a genetic predisposition might exist<sup>9,10,12-16</sup> and an infective agent might be implicated in the pathogenesis of this disease.<sup>9,10,12-15</sup> We present a further case report of PMR in a married couple.

## Case presentation

### Wife

The 55-year-old wife, working as housewife and accordion teacher, complained in June 2010 about pain in her neck. She recognized it when she had problems rotating her head while driving her car. In the following months, the pain extended to the shoulders and the arms. The physical examination of the 156 cm, 68 kg woman showed a limited mobility

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of the upper limbs, with no limitation of the flexibility of the hip and lower limbs. Magnetic resonance tomography showed a disk prolapse at C3/C4. In October 2010, laboratory parameters were determined, and erythrocyte sedimentation rate (ESR) was at 80 mm/h. Rheumatoid factor (RF) and anticitrullinated protein antibody (ACPA) were negative. In her family history, her father had died at the age of 79 years after three apoplectic strokes while her mother was still alive at the age of 92 years. She had two brothers and two sisters; the younger sister had breast cancer. A rheumatologist was consulted, and a diagnosis of PMR was established. Figure 1 shows the course of ESR and C-reactive protein (CRP). Upon diagnosis, therapy with 40 mg of prednisone was started, with reduction by 10 mg every month down to 10 mg prednisone. Unfortunately, she suffered from side effects of prednisone, such as an increase in body mass. In order to lower the cumulative dose of glucocorticoid therapy, 10 mg methotrexate per week was added.<sup>17</sup> Between January 2011 and April 2012, she received 10 mg prednisone daily and 10 mg methotrexate weekly.

Figure 1 Time course of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) for wife (panel A) and husband (panel B).

## Husband

In May 2011, the 66-year-old husband also complained about pain in his neck, shoulders, buttocks, and thighs. In addition, he lost 7 kg of body weight within a period of 6 months. An extensive examination with abdomen sonography, chest radiography, and laboratory analyses was performed in order to exclude cancer. His ESR was 56 mm/hour, and a rheumatologist (different from the one the wife visited) was consulted. Physical examination revealed an unlimited but

painful mobility of the upper limbs. He suffered from pain on palpation on both shoulders, both upper arms, and both thighs. RF and ACPA were negative. Data for his family history were lacking since he grew up in a children's home and was placed with foster parents later. He had four siblings with no rheumatic diseases. Prior to his retirement at the age of 60 years, he had worked as a police officer and postal van driver. Eleven months after the first symptoms of his wife, he also received a diagnosis of PMR. Figure 1 shows the course of ESR and CRP. The husband was afraid to take prednisone due to the side effects of prednisone his wife had experienced. Since his wife could lower prednisone intake due to the simultaneous intake of methotrexate, he finally agreed also to start with prednisone, and ESR started to drop. Upon diagnosis, he received 10 mg prednisone daily and 10 mg methotrexate weekly.

Following the 2012 provisional classification criteria for polymyalgia rheumatica, with a scoring algorithm taking into account morning stiffness >45 minutes (2 points), hip pain/limited range of motion (1 point), absence of RF and/or ACPA (2 points), and absence of peripheral joint pain (1 point), the wife got 4 points and the husband 5 points.<sup>18</sup> According to these provisional classification criteria, patients  $\geq 50$  years old presenting with bilateral shoulder pain that is unexplained by an alternative pathology can be classified as having PMR in the presence of morning stiffness > 45 minutes, elevated CRP, and/or ESR, and new hip pain.<sup>18</sup>

## Discussion

PMR is clinically characterized by severe bilateral muscle pain and aching involving the neck, shoulders, and pelvic girdles associated with morning stiffness.<sup>19</sup> ESR is generally markedly elevated, and both clinical symptoms and elevated

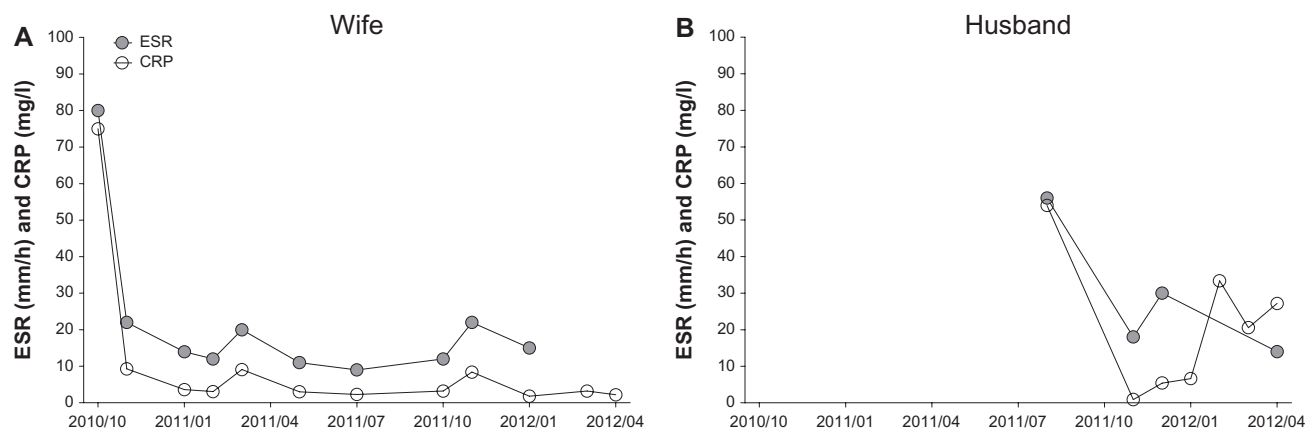


Figure 1 Time course of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) for wife (panel A) and husband (panel B).

ESR decline after treatment with high-dose prednisone.<sup>20</sup> As the syndrome is defined by nonspecific elements and a classification overlap with giant-cell arteritis (GCA), the multiple classification criteria mention that other diseases have to be excluded. There is indeed a wide range of diagnostic procedures looking especially for large-vessel vasculitis, calcium pyrophosphate deposition disease (CPPD), and late-onset rheumatoid arthritis (LORA), and a pragmatic approach by the general practitioner.

In this couple, PMR occurred first in the wife, then in the husband. Regarding the six reported cases in the literature, an interesting finding was that PMR occurred in four of six cases first in the wife (Table 1), as was the case in the present couple. Generally, a ratio of 3:1 of women to men is reported for PMR.<sup>1</sup> In contrast to the existing case reports (Table 1) with PMR in married couples, the present couple was younger but still aged over 50 years, where PMR is frequent.<sup>1</sup> In our two patients treated pragmatically with prednisone, neither GCA, CPPD, nor LORA have been definitely excluded. One may weigh the differential diagnosis by the initial dose

of prednisone to suppress the symptoms with lower doses in case of CPPD-disease or “pure PMR” without GCA. In addition, there may be relapses of the symptoms during the tapering of glucocorticoids in cases of GCA, even without cranial symptoms, as the incidence of occult giant-cell arteritis is clinically underestimated.

Our couple originated from northern Europe. The reported cases of PMR in married couples occurred in Denmark,<sup>8</sup> Sweden,<sup>8</sup> Norway,<sup>9</sup> Scotland,<sup>15</sup> Israel,<sup>9</sup> and the USA.<sup>6,12</sup> It has been reported that the prevalence of PMR is higher in individuals of Scandinavian background.<sup>1</sup> In Southern Europe, the annual incidence of PMR was considerably lower.<sup>1</sup>

Case reports on PMR were published between 1974 and 1992.<sup>4-9</sup> During the last 20 years, no case of PMR in a married couple occurred, or it was not published. Although in most of these cases, a genetic disposition<sup>5,8,9</sup> and an infectious agent<sup>6,7,9</sup> have been discussed, it is difficult to build up this hypothesis on a few case reports. Moss and Soukop concluded that “a powerful argument against an infective etiology remains the long time interval which

**Table 1** Case reports with polymyalgia rheumatica in married couples in the literature

Husband		Wife		Interval between appearance (months)	Details	Reference
Age at diagnosis (years)	ESR at diagnosis (mm)	Age at diagnosis (years)	ESR at diagnosis (mm)			
78	95	76 <sup>#</sup>	98	8	In the wife, treatment with 10 mg prednisone was started and all symptoms disappeared after 48 hours. In the husband, treatment with 60 mg prednisone was ineffective, but after 100 mg prednisone, a marked improvement was noted after 48 hours.	7
71 <sup>#</sup>	49	69	58	0	The wife presented the day after her spouse with clinical symptoms. Both patients were treated with prednisone and symptoms regressed.	6
76	72	78 <sup>#</sup>	64	1	Husband and wife began suffering from the disease within the same month and an infectious etiology seemed plausible. Both were HLA-B8-positive.	10
69	78	69 <sup>#</sup>	57	24	In both patients, low-dose prednisone therapy continued.	9
78	119	74 <sup>#</sup>	92	83	The husband's PMR symptoms started 4 years after his wife's death.	8
68 <sup>#</sup>	60	70	133	89	Five family members with PMR or an illness compatible with PMR were reported. The husband developed the disease 6–7 years earlier than the siblings, highly suggestive that the disease was caused by an infectious agent and genetic disposition is essential for the development of PMR.	11

**Note:** <sup>#</sup>First diagnosis.

**Abbreviations:** ESR, erythrocyte sedimentation rate; PMR, polymyalgia rheumatica; HLA, human leukocyte antigen.

may separate the onset of symptoms in family members or conjugal pairs.”<sup>15</sup> In the present couple, we have no data from the family history of the husband since he grew up in a children’s home and was placed with foster parents later. A genetic relation between husband and wife is unlikely, but therefore cannot be definitely excluded. However, PMR in a married couple argues against genetic background, and this observation proposes an environmental trigger to the disease.

Treatment with 10–20 mg of prednisone daily is suggested for PMR.<sup>21</sup> There are no recommendations in guidelines regarding the exact dose or duration of administration; the drugs and the rate of reduction of prednisone should be adjusted depending on the individual’s response and with consideration of the multiple risks of high-dose and long-term glucocorticoids.<sup>21</sup> A prolonged course of treatment is necessary, and corticosteroids are gradually tapered, guided by regular clinical evaluation and ESR (and/or CRP) measurement. Methotrexate is the best-studied corticosteroid-sparing agent, and may be useful for patients with frequent disease relapses and/or corticosteroid-related toxicity.<sup>22</sup> However, superiority of a combination therapy in reducing the incidence of glucocorticoid-related complications has not been shown yet.<sup>23</sup>

## Disclosure

The authors have no conflict of interest in this work.

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