Reply

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We thank Drs Galassi and Luppi for their interest in our recent case report.¹ Although they express doubt about the paraneoplastic etiology of the neuropathy in our patient, we believe the strong temporal relationship of symptom improvement, electrophysiologic resolution of the demyelinating neuropathy, and disappearance of the anti-MAG antibody without specific immunotherapy are all strong indicators of a causal relationship with the resected colon cancer. DADS neuropathy is a chronic neuropathy, and clinical and electrophysiological improvement is difficult to achieve with even intensive immunotherapy, while the anti-MAG antibody also tends to persist. That these features all improved promptly following surgical cancer resection makes a strong case for a causal link with tumor. However, we acknowledge that ongoing surveillance for tumor recurrence remains a clinical requirement.

The Galassi and Luppi patient is quite different from ours. In their patient a treatment responsive demyelinating neuropathy consistent with CIDP occurred approximately 4 years prior to diagnosis of colon cancer. Galassi and Luppi do not provide any additional evidence for a causal link with the tumor in their patient apart from the change in responsiveness to immunotherapy, which may well be coincidental. The later discovery of an IgG lambda paraprotein raises the possibility of a paraproteinemic demyelinating neuropathy. However this may also be coincidental, as IgG MGUS may occur in otherwise healthy older individuals, while CIDP features are not different in patients with or without IgG MGUS.

We believe that the case for a paraneoplastic etiology is compelling in our patient. If we apply the criteria discussed by Graus et al,² our case would be classified as "a non-classical syndrome that resolves or significantly improves after cancer treatment without concomitant immunotherapy, provided that the syndrome is not susceptible to spontaneous remission" which the expert panel accepts as "definite paraneoplastic syndrome". The Galassi and Luppi patient in contrast presented as a non-classical syndrome with negative onconeural-antibodies and late development of cancer, considered at best "possible paraneoplastic".

Reference:

1. Galassi G, Luppi G. Distal acquired demyelinating symmetric neuropathy associated with colorectal adenocarcinoma: should it be termed paraneoplastic? Muscle Nerve 2015 (accepted 17 September 2015, in press)
2. Graus F, Delattre JY, Antoine JC, Dalmau J, Giometto B, Grisold W, et al. Recommended diagnostic criteria for paraneoplastic neurological syndromes. J Neurol Neurosurg Psychiatry 2004;75:1135–1140.

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Title:

REPLY: DISTAL ACQUIRED DEMYELINATING SYMMETRIC NEUROPATHY ASSOCIATED WITH COLORECTAL ADENOCARCINOMA: SHOULD IT BE TERMED PARANEOPLASTIC?

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