Chronic Inflammatory Demyelinating Polyneuropathy

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You may have heard of some new antibodies associated with chronic inflammatory demyelinating polyneuropathy (CIDP). You may be wondering if you need to be tested for those antibodies, and if you had those antibodies what would that mean. I hope to give you a little background to help you think about these antibodies and whether you need to discuss this with your treating neurologist. Please be aware that we are early in learning about these antibodies and there is much that is to be learned yet

If you've met other people with CIDP you are aware that you share many similar symptoms. However it is also likely you have noticed that all people with CIDP are not the same. For a long time it has been clear that there are variations in presentation of chronic acquired demyelinating polyneuropathies. Many of those differences are based on how the disease presents in an individual patient. For example some disorders seem to pick off individual nerves. Examples include diseases like Multifocal Acquired Demyelinating Sensory and Motor Neuropathy (MADSAM), or Multifocal Motor Neuropathy with Conduction Block (MFMN). It is also clear that different disorders respond differently to treatment. For example MFMN does not respond to corticosteroids like prednisone. Lastly we had been aware that some of these disorders have specific associated antibodies like anti Mag, and anti GM1 with Distal Acquired Symmetric neuropathy (DADS) or MFMN.

About ten years ago two new antibodies have been discovered in patients who were diagnosed with CIDP. Since then some other antibodies have also been discovered. These antibodies are directed against proteins that are found in peripheral myelinated neurons. Most interestingly they are all located at the paranode. This is a very important area that helps with conduction of an electrical impulse along a myelinated nerve fibre.

We are still learning about neuropathies associated with these antibodies. The number of identified antibodies is also growing and includes:

Neurofascin 155
Neurofascin 140/186 Contactin 1
Contactin associated protein 1

While some patients may look like they have typical CIDP there may be a difference in how these neuropathies present. For example tremor may be a more common problem in anti-neurofascin associated CIDP. Distal weakness is also commoner in paranodal antibody associated CIDP than in typical CIDP. Most importantly these disorders may respond differently to medical therapies than typical CIDP.

Typical CIDP has a number of possible therapies, but prednisone and IVIG are commonly used first line therapies. For many patients first line treatments result in significant improvement. However at least in some of the paranodal antibody associated variants of CIDP IVIG may be less effective.

Growing evidence suggests alternate therapies may be more effective. Two therapies that appear to be effective in some of the paranodal neuropathies are rituximab and plasma exchange. Given this information it is important we recognized variants early on that may need alternate therapy strategies. This raises the question of who should be tested.

There are no firm guidelines on when to test or who should be tested for these antibodies. This is my opinion. There are two groups that should be tested:

1) Newly diagnosed CIDP patients.
2) CIDP patients that are not well controlled on their current therapies.

Patients with longstanding CIDP who are well controlled on their current therapies without significant side effects need not be tested as at this time it is unlikely to alter therapy.

At your next visit with your neurologist you may want to ask them about these antibodies and whether it is worth being tested. My understanding is that different provinces may have different access to the antibody testing and funding for the tests.

The Bottom Line

1) There are some newer antibodies associated with CIDP
2) They may have slightly different symptom profiles than typical CIDP
3) Some of these variants of CIDP may not respond as well to conventional therapy especially IVIG
4) Consider testing newly diagnosed CIDP patients and poorly controlled patients with CIDP.