

DR. SEAN

PITTOCK:

Autoimmune dementia is a recently recognized entity characterized by a condition that's mediated by the immune system, but that's potentially treatable and reversible. The main characteristics that we think of when we suspect a diagnosis of autoimmune dementia are generally a subacute onset of cognitive decline that's often rapidly progressive. Fluctuating course is often seen.

But there are other clues also for a clinician. For example, of personal or family history of autoimmunity, a history of cancer, because sometimes we see all the immune neurologic disorders including autoimmune dementias occurring in the setting of the cancer. So that's very pertinent if the patient has had a history of a recent or even remote cancer. History of heavy smoking it needs to be considered also.

And then the presence of some atypical features for example on the MRI scan or the presence of inflammation of the spinal fluid. Certain neurological associations also can be seen. For example, the presence of myoclonus or tremor or headache. And then importantly, in respect to this talk, we need to think about neural autoantibodies. And that's why this comprehensive evaluation I think will be very helpful for clinicians.

In the cases of dementias, as we have seen patients where they have come with significant cognitive deficit and we've treated them with immunotherapies. And when I say immunotherapies, I mean drugs like steroids or intravenous immunoglobulin or immunosuppressant medications like Cellcept or azathioprine or sometimes rituximab. And we've seen dramatic benefits in some patients, even near-complete reversibility.

I think the main important issue here is the comprehensive nature of this evaluation and the fact that rather than you ordering a test for a paraneoplastic antibody or a test for neural antibodies, you're actually ordering a test for autoimmune dementia, a specific evaluation that we have selected the specific antibodies that we think are relevant to that disease. And it's comprehensive and it is complex.

It includes indirect immunofluorescence assays, ready immuno precipitation assays, cell based assays, Western blot assays, but at the end of the day, you're getting an array of antibodies that are relevant to autoimmune dementias and some of those antibodies would be things like antineuronal antibody type one or [INAUDIBLE] five antibodies. And these antibodies can be associated with autoimmune dementias, but they're generally associated with paraneoplastic dementias. In other words, those antibodies are predictive not only of the presence of a likely autoimmune etiology for the dementia, but also for the likelihood of an underlying malignancy.

And then there are other antibodies like the voltage-gated potassium channel complex antibody and we described this in the setting of rapidly progressive dementias, even disorders that mimic Creutzfeldt-Jakob disease that are very treatable. And those antibodies generally are not associated with malignancies. Those would be more autoimmune dementias. And the whole array of neural antibodies if they're present, I think really just assist the clinician in saying, number one, these antibodies are present and therefore I really need to think about the possibility of an autoimmune dementia. And the presence of these antibodies really supports that.

And number two, does the profile of antibodies indicate the potential for an underlying malignancy? In other words, that it might direct attention not only toward giving an immunotherapy trial, but also toward looking for a malignancy.

The neural autoantibody evaluation and a positive result, it must be interpreted in the clinical setting. And that I think is very important. We don't treat antibodies. We treat patients. And we must interpret the findings in the appropriate clinical context.

We do provide a consultation in the sense that neurology consultant will review the results and will actually report on those results. So you will get a physician created report. But if you have further questions you always have the opportunity to ring us at the Mayo Clinic in the neuroimmunology lab and talk to one of the autoimmune neurologists that works there, and we can certainly guide you in terms of how to interpret the antibody and what tests you might want to think about doing in the setting of an antibody mean positive.