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Atypical neuropilar cerebellar immunochemistry staining in rapidly progressive cerebellar syndrome in woman with breast cancer

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Introduction: Rapidly progressive cerebellar syndrome (RPCS) is frequently paraneoplastic in postmenopausal women with breast and ovarian cancer, associated with so-called high-risk for cancer antibodies [1]. This syndrome is a treatable condition but often with no full recovery [2].

Case presentation: A 67-years old woman was admitted to our Neurology Department for a ten days onset of nausea, vomit, sickness, weight loss (almost five kilograms in two weeks), marked gait instability, dysarthria, hypophonia, diplopia, dysphagia and limb and trunk ataxia, Modified Rankin Scale (mRS 4).

Results: The patient underwent several brain MRI scans, with nonspecific findings. Lumbar puncture was carried out showing elevated cerebrospinal fluid (CSF) protein concentration (48 mg/dl) and mild elevation of cell counts (27 cells/mm3), with glucose level in normal range (71 mg/dl). Cultural, virological exams and most common anti-cerebellar antibodies were all negative or nonspecific. PET total body scan and mammography detected a mammary lesion (BI-RADS 6) with axillary lymphadenopathy.
Biopsy with immunochemistry and biological profile found out estrogen receptor (ER) positive breast cancer. Using indirect immunofluorescent tissue based assay exploiting lightly fixed rat brain tissue, we detected on CSF an uncharacterized neuropilar staining involving the molecular layer of the cerebellum.
In the suspicion of paraneoplastic RPCS, the patient underwent five days of intravenous immunoglobulins treatment (twenty grams/day), according to patient's weight, then replaced by steroid with slow tapering. Surgery was excluded by the Breast Unit team, and nonsteroidal aromatase inhibitors (letrozole) treatment was started.
At 6 months follow-up, there was a little neurological improvement with persistence of dysarthria, and ataxic features, mRS 3.

Discussion: The features of immunochemistry staining resemble anti-Tr/Delta/Notch-like Epidermal growth factor-related Receptor (DNER) antibodies pattern. Our patient presented several clinical features in common with RPCS associated with these antibodies, usually lymphoma and solid tumors related [3], instead no breast cancer cases were still reported.

References:

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