

Paraneoplastic Cerebellar Degeneration (PCD) in patients with Ovarian cancer, A review of 112 cases

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Background

PCD with Anti-Yo antibodies is a rare but disabling neurodegenerative disease that may point to an occult ovarian cancer. While diagnostic criteria are generally accepted, therapy is largely debatable with a few anecdotal reports of success. Management of the syndrome is often unsatisfying, with severe neurological sequelae. We present an extensive review detailing the epidemiology, clinical behaviour and treatment outcomes of patients with PCD and ovarian cancer.

Methods

PubMed and OVID databases (1985- 2009) were searched using the keywords, PCD and ovarian cancer. Our search yielded 112 reported cases of ovarian cancer in a total of 137 patients with PCD and gynecologic cancers.

Results

Ataxia, dysarthria and vertigo were the most common presenting features. The onset of neurological symptoms preceded the diagnosis of malignancy in 68% cases. Ataxia, dysarthria and vertigo were the most common presenting features. 24% of these patients were diagnosed at Stage 1/2. In four cases, only microscopic carcinoma could be found after surgery. One case of occult ovarian carcinoma was diagnosed with PET-CT when conventional imaging was negative. The median age of these patients was 54 as compared to 56 for patients diagnosed post-cancer. No benefit was seen by surgery in most cases, even with complete eradication of disease. Clinical response was seen in 5 out of 9 patients treated with IVIG with disease stability in 4 and symptomatic improvement in 1. Patients treated within the first month of diagnosis benefited the most. Plasmapheresis was beneficial in 40 % of treated cases with significant neurological improvement in 33% cases. No difference in treatment response was noted in patients with known cancer diagnosis. However, PCD followed a progressive clinical course in more than 70% patients; no patient with PCD & Stage-4 disease derived any benefit from therapy. 28% patients died within 3 years of diagnosis with neurological deterioration being the primary cause of death in more than 60% cases.

Conclusions- Early recognition of PCD & search for occult malignancy can help diagnose ovarian cancer at an early stage in greater than 1/5th patients and in more than 4/5th of patients before widespread metastases, hence increasing the window of opportunity available to oncologists. Therapy is most beneficial if given early, suggesting that PCD should be always be included in the differential diagnosis when considering the etiology of unforeseen neurological symptoms and empiric immunomodulatory therapy should be considered. PCD presents more questions than answers, making it absolutely critical to explore newer therapies and better screening tools.