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A Case Report: Autoimmune Encephalitis Secondary to Paraneoplastic Syndrome in a Sigmoid Adenocarcinoma Patient

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ABSTRACT

Paraneoplastic syndrome is a syndrome caused by the remote and non-metastatic effects of cancer and can present with neurological signs and symptoms. This is a case report of a sixty-five-year-old patient who presented with agitation, confusion and slurred speech, found to have positive anti-neuronal nuclear antibodies (ANNA2) consistent with Paraneoplastic syndrome and was

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later diagnosed with sigmoid adenocarcinoma with para-aortic lymph node involvement. This presentation of colon cancer is a rare occurrence that the author felt should be looked at in detail.

Key Words: Paraneoplastic neurological syndromes, autoimmune encephalitis, colon cancer, sigmoid cancer

Abbreviations: ANNA2 - Anti-neuronal nuclear antibodies

1. INTRODUCTION

By definition, paraneoplastic syndromes are known as the remote effects of cancer that result from a disturbance other than that of the tumor and its metastasis, infection, ischemia or metabolic disease. paraneoplastic neurological syndromes are rare, with a prevalence of less than 1/10,000 cases of all cancers. As they are caused by an autoimmune response directed towards the cancer and the nervous system, diagnosis was made possible by detecting anti-onconeural antibodies which can be 90% specific, although not always present(1). In the case discussed here, the patient presented with agitation, confusion and slurred speech. Anti neuronal nuclear antibodies (ANNA2) were detected which is consistent with paraneoplastic syndrome.

2. CASE REPORT

2.1. History

The patient is a 65 year old gentleman who initially presented with a fall one for which he was admitted to a community hospital. Prior to this his family stated he was developing some memory loss over the past five years with difficulty sleeping. The impression during this admission was that the fall was secondary to a transient ischemic attack (TIA). After being discharged, the family noticed the patient was becoming more confused, unable to speak properly, searching for words with episodes of agitation and difficulty walking. One month after his first admission, the patient became very agitated and his speech was incoherent. The patient denied any history of headache, visual disturbance or swallowing problems. The patient has a past surgical history of a left nephrectomy for renal cell cancer. His past medical history was significant for a TIA seven years ago for which he was on clopidogrel, hypertension and some renal dysfunction. He was also noted to be anemic and was scheduled for a colonoscopy. The patient had a past history of smoking and no significant history of alcohol consumption.

2.2. Examination

On examination the patient was vitally stable. The patient had difficulty following commands and required extensive prompting to persist with tasks. A neurological exam showed intact sensation and motor function, with normal tone. The patient had evidence of dysdiadokinesis and cerebellar signs. Patients speech output was minimal with some echolalia. The patient demonstrated signs of receptive and expressive dysphasia when trying to obtain a history with word finding difficulties and difficulty speaking.

2.3. Work up and management

A CT scan of the head revealed some white matter microvascular ischemic changes and cortical atrophy. An MRI of the brain revealed some evidence of chronic small vessel ischemia with disproportionate atrophy of the frontal and parietal lobes. A Doppler of the carotid vessels showed them to be widely patent. A lumbar puncture was performed, and analysis of the CSF was positive for ANNA 2. The patient was diagnosed with autoimmune encephalitis, and started on steroids and intravenous immunoglobulin but failed to show any signs of improvement. A Colonoscopy was performed and it showed a sigmoid cancer. A CT scan of the abdomen was suggestive of localized disease with no lung or liver metastasis. The patient then had an open sigmoid resection with diverting ileostomy performed as an inpatient. This was uneventful except for a high output stoma which resolved spontaneously. After the surgery the patient showed some evidence of mild improvement from a neurological standpoint. The patient was kept in hospital for rehabilitation before being discharged to a nursing home. The pathology of the tumour revealed a T3N2aM0 sigmoid adenocarcinoma. He was discussed at the multidisciplinary tumour board and the decision was made to give the patient chemotherapy. The patient continued to follow up with us and showed some continued mild neurological improvement especially in his speech, but nevertheless, still had a significant deficit.

Aljifri and Farsi,
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3. DISCUSSION

Neurological presentation of paraneoplastic syndrome caused by Sigmoid colon cancer is a very rare disease. Nonetheless, the study of paraneoplastic neurological syndrome is interesting and thought provoking. The syndrome is believed to be a result of the activation of an autoimmune response triggered by the cancer and directed towards antigens common to cancer and the nervous system. There had been a few scattered case reports on breast, prostate, and small cell lung cancer presenting with paraneoplastic neurological syndromes (3). One report described a case of sigmoid colon cancer presenting with polymyositis secondary to paraneoplastic disease (4), and another described a case of colorectal cancer presenting with paraneoplastic syndrome mimicking adult still's disease (5). Cases similar to the one described in this manuscript have been reported. One of them is a case of a 53-year-old patient with colonic adenocarcinoma presenting with confusion and slurred speech among other central nervous system manifestations that markedly improved after surgery like the case discussed here. Unlike this case, however, they did not detect any onco-neuronal antibodies, which confirm that their presence is not required to make a diagnosis of paraneoplastic syndrome (6). Another report from Ohio described a case of rectal carcinoma with dementia in 2011 which fulfilled four criteria of diagnosis (7). Correct and early diagnosis of paraneoplastic neurological syndromes can allow early detection of an occult tumor and the treatment of the tumor may stabilize a disorder that otherwise would be difficult to treat (2). This case offers a good teaching point as well as providing insight to possible differential diagnoses to patients presenting with paraneoplastic neurological symptoms especially older patients.

4. CONCLUSION

Paraneoplastic neurologic syndromes in the setting of colorectal cancer are very rare. When they do occur they can sometimes improve with treatment of the primary cancer, so every effort should be made to offer patients surgery as soon as possible both in order to remove the primary tumour and to relieve these distressing symptoms.

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