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Myasthenia-like paraneoplastic syndrome secondary to suspected ovarian cancer.

Ornjira Angkanawatana.
Medical Student, Bristol University

BACKGROUND

Paraneoplastic neurological syndromes (PNSs) are defined as "the pathological involvement of the nervous system in the course of malignancy"[1]. Literature acknowledges myasthenia gravis (MG) as a PNS most commonly associated with thymoma[5]. However, there is no recognised association of MG with ovarian cancer.

CASE PRESENTATION

A 67-year-old woman presented with gradual onset non-fatigable slurred speech with dysphagia, diplopia, bilateral ptosis, and a right-sided facial droop and weakness particularly in her right arm. The dysphagia had been present intermittently for ten days prior to hospital admission. There was no remarkable past medical history of disease. On examination, the patient's abdomen was grossly distended with a large palpable pelvic mass that was deviated to the right.

INVESTIGATIONS

The patient underwent head CT/MRI scans and a lumbar puncture to exclude primary neurological causes for her symptoms. Results for all three tests were unremarkable.

Nerve conduction studies revealed deficits of neuromuscular junction transmission and large sensory fibres. Thus, the patient was tested for acetylcholine receptor (AChR) antibodies, demonstrating a result of >20 nmols/L consistent with a diagnosis of myasthenia gravis.

Blood tests for tumour markers CA-125, CA19-9, and carcinoembryonic antigens were found to be elevated at 164 KIU/L, 63 ug/L, and 1300 KIU/L, respectively. Transabdominal ultrasound scan (USS) and CT abdomen identified a huge complex part-solid part-cystic mass of approximately 25 cm (9.84 inches) in length arising from the right adnexa, demonstrating significant intrinsic vascularity.

DIFFERENTIAL DIAGNOSIS

With the tumour's location and radiological characteristics, supported by elevated tumour marker levels, the most likely diagnosis was ovarian carcinoma.

Considering unremarkable findings on head CT and MRI scans, malignant meningitis and Paraneoplastic neurological syndrome (PNS) were considered as possible causes of the patient's neurological symptoms. Malignant meningitis was excluded with normal CSF. Hence, combining the diagnosis of malignancy with the patient's nerve conduction studies and seropositive AChR antibodies results, the working diagnosis was myasthenia-like paraneoplastic syndrome secondary to ovarian cancer.

TREATMENT

The patient was fed via a nasogastric tube and supplemented with fortisip compact to increase nutritional intake whilst her dysphagia improved. This was facilitated through speech and language therapy (SALT), which also aimed to improve her dysarthria.

Myasthenia gravis symptoms were treated with pyridostigmine; with propantheline to decrease cholinergic side effects. The patient's FVC was monitored to safeguard development of myasthenia crisis.

Regarding the ovarian cancer, the patient was scheduled for surgical tumour resection via total abdominal hysterectomy with bilateral salpingoophorectomy, omentectomy, and appendicectomy.

Outcome and follow-up

The patient responded well to pyridostigmine, showing most significant neurological recovery in her right arm. Her dysphagia and slurred speech resolved completely. Her right facial droop remained but was demonstrating good recovery. After her laparotomy, the patient was to be followed-up by the gynaecology oncology team.

DISCUSSION

Ovarian tumours account for around 10% of PNS-associated malignancies, the most common syndrome for ovarian carcinoma being paraneoplastic cerebellar degeneration.[3] PNS often antedates diagnosis of cancer, and in patients with neurological deficit with no known malignancy, detection of onconeural antibodies such as anti-Yo strongly suggests the presence of ovarian tumour.[3]

Presently, paraneoplastic antibodies are not proven to be pathogenic[7] and are therefore only useful as diagnostic markers rather than targets for treatment.

As the primary cause of PNS is malignancy, neurological outcome can improve after tumour excision[8]. However, the question lies in whether a diagnosis of PNS associated with ovarian tumour alone is adequate to constitute an indication for surgical management.[3]

LEARNING POINTS

PNS can precede manifestation of a tumour; therefore, initial diagnosis of PNS should trigger a search for associated tumour. With paraneoplastic myasthenia gravis, specific investigations should be taken to exclude thymic tumour.

As PNS can mimic any neurological syndrome, a high index of clinical suspicion is important for early diagnosis and prompt management. In ovarian tumours, early resection is a significant part of PNS treatment and improves the outcome.

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