Case Report

Paraneoplastic cerebellar ataxia as the initial manifestation of occult fallopian tube carcinoma : a case report

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- **54 year old male with subacute progressive gait difficulty,**
- **slurring of speech**
- **weight loss**

History of Patient Illness :

A 54-year-old post-menopausal lady, from rural Bengal, presented in our neuromedicine outdoor with a history of subacute onset, progressive unsteadiness of gait, incoordination of limbs, slurring of speech and significant weight loss over the past 5-6 months. Her past history was unremarkable except for a vaginal hysterectomy surgery (oophorectomy status unknown) which was done ten years ago for uterine prolapse.She did not give any history of breast or gynecological malignancies in her family members.

General Examination :

She was alert and oriented poor nutrition and multiple, bilateral hard, non-tender, discrete supraclavicular lymph nodes.

Neurological Examination :

Revealed bilateral gaze-evoked nystagmus, scanning type of dysarthria, dysmetria, dysdiadochokinesia, and severe gait ataxia.

Other Systemic Examinations (including breast) :

Were unremarkable. A thorough search for occult malignancy was planned, and gynecological referral was made.But no adenexal pathology could be detected clinically on vaginal examination.

Investigation :

Her routine blood investigationswere all within normal limit.Magnetic resonance imaging (MRI) of brain revealed diffuse cerebellar atrophy. Cerebrospinal fluid study showed mild pleocytosis. Fine needle aspiration cytology (FNAC) from the left supraclavicular lymph node suggested metastatic deposit from carcinoma. A screening for primary was carried out, including contrast enhanced computed tomography (CT) scan of thorax, abdomen and pelvis which reported the presence of multiple enlarged, necrotic paratracheal, mesenteric and retroperitoneal lymph nodes without any evidence of primary. Therefore, a whole body 18F-Flurodeoxyglucose (FDG) Positron emissions tomography (PET)-CT scan was obtained whichshowed a large solid-cystic left adenexal space occupying lesion (SOL) measuring 3.93 cm X 3.48 cm X 5.76 cm with multiple loculation and increased FDG uptake in the solid part with SUVmax (Standardized uptake value) of 12.91, probably ovarian in origin with non-visualization of uterus and with multiple, distant lymph nodes metastasis (Fig 1).Serum CA-125 level was markedly elevated (1866 units/ml) and paraneoplastic serum antibody panel was positivefor anti-Yo antibody in high titers.

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Provisional Diagnosis :

Occult ovarian malignancy causing PCD.

Gynecological consultation was made again and she was posted for staging laparotomy under general anesthesia.

The abdomen was opened through midline, low-vertical incision, left ovary found to be normal; left fallopian tube found fibrosed at proximal and distal end, and is adhered with a mass measuring 4 cm X 3 cm, omentum and transverse colon. Excision of the mass was done along with infracolic omentectomy, left salpingectomy and left oophorectomy. Right ovary was also found normal but adhered to gut; right fallopian tube fibrosed at proximal and distal end and was adhered to pelvic peritoneum. Right salpingectomy with oophorectomy, and appendectomy was done and peritoneal washing was taken. On hystopathological examination the tumor showed serous papillary carcinoma with micropapillary pattern (Fig 2). The carcinoma has invaded the muscle coat of the fallopian tube and has extended through the serosa into the adjacent fibroadipose tussue. Lymphovascular invasion was found. No perineural invasion was noted. The carcinoma has not invaded the adjacent ovary. The other ovary and fallopian tube were unremarkable. No evidence of metastasis was found in the resectedomental tissue.

Therapy :

She received six cycles of platinum-based chemotherapy with some improvement in her general health condition but neurological condition remained more or less same.

Follow-up :

Imaging, performed 6 months post-surgery, did not reveal any residual disease or relapse.

Discussion

In our patient, cerebellar symptoms preceded the diagnosis of cancer by approximately 5 months, similar to the mean reported by Rojas et al. among patients with anti-Yo positive PCD[1]. There may be a variety reasons for delay in the diagnosis, seen in our patient. Lack of awareness among the treating physicians; past history of hysterectomy with ?oophorectomy; occult nature of the tumor as evidenced by lack of local symptoms, normal per vaginal examination, failed detection even on CT; and lastly, the cost limiting factorof whole body PET-CT scan are some of the major reasons.

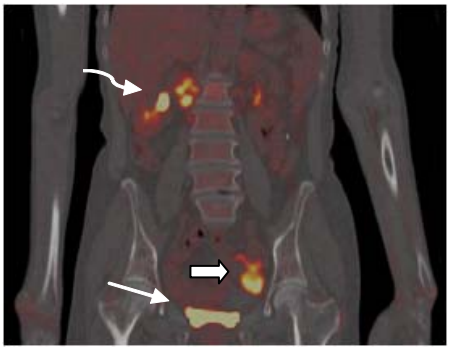

Fig 1 — PET-CT showing hypermetabolic left adenexal SOL (thick arrow), mesenteric and paraaortic lymph nodes with increased FDG uptake (curved arrow), and physiological FDG uptake in bladder (thin arrow)

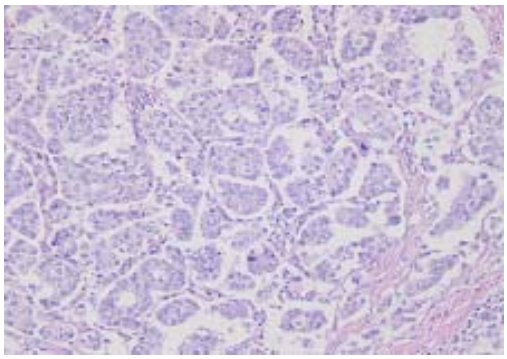

Fig 2 — Histopathological slide of the fallopian tube SOL showing serous papillary carcinoma

Very few case reports are there in the literature showing the association between primary fallopian tube cancer and PCD[2-4]. Therefore, its knowledge is lacking among the neurophysicians. Primary malignancy of fallopian tube itself is very rare, accounting for only 0.1-1.8% of all gynecological malignancies[5]. More than two-third of cases occur in post-menopausal women with a mean age of 55 years[6]. Despite the similar embyogenic origin of fallopian tube and uterus, malignant lesions of fallopian tube behave like ovarian cancers both clinically and histologically.. Most of the primary fallopian tube cancers are papillary serous adenocarcinomas[7]. Due to the close proximity of fallopian tubes, uterus and ovaries, detection of the true primary is challenging, and moreover, it is not routinely suspected. Correct preoperative diagnosis is made only in around 0.3% of the cases and most are diagnosed peri- or post operatively[8]. Ca-125 level in serum is elevated in significant numbers of patients (around 70%) with higher level correlating with the higher clinical staging[9]. The principle of management is same as that of ovarian cancer.

Now regarding the management of neurological symptoms, early tumor detection and it removal offers the best possible choice. Once the cerebellar degeneration is started, it becomes less responsive to the immunotherapies or tumor removal.

Conclusion

This case highlights the importance of suspecting underlying malignancy in case of sub acutely developing cerebellar syndrome even if it is not obvious on routine imaging. Clinical clues include history of anorexia, recent weight loss, cachectic built or presence of lymphadenopathies as in our case. The need for obtaining early whole body PET-CT and paraneoplastic antibody panel, including anti-Yo antibody, cannot be overemphasized. Primary fallopian tube malignancy can cause PCD, and should be suspected in anti-Yo positive patients.

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Editorial Comments :

- **Paraneoplastic neurological syndrome needs special attention.**
- **Every part of nervous system can be affected.**
- **Cancer of lung, breast, overy, endometrium, lymphoma can cause different paraneoplastic neurological syndrome.**

Key words **: Paraneoplastic cerebellar degeneration.**