

Leptomeningeal Carcinomatosis from Primary Mucinous Carcinoma of the Ovary

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Background

Ovarian cancer is the fifth leading cause of cancer death in the United States and at diagnosis, 86% of tumors are advanced FIGO (III-IV). The frequency of brain metastases from primary ovarian cancer is thought to be between 1% and 3%. Leptomeningeal carcinomatosis (LC) is a rare complication of ovarian cancer, characterized by the infiltration of malignant cells into the leptomeninges, the thin layers of tissue that surround the brain and spinal cord. LC is an exceedingly rare manifestation of metastatic ovarian cancer, and essentially a lethal prognosis.

Objective

To present a case of LC from a primary ovarian cancer and provide a brief review of current literature, detailing current modalities of treatment and prognosis following diagnosis of leptomeningeal carcinomatosis.

Case Presentation

A 63 y/o female with a history of platinum resistant recurrent metastatic mucinous adenocarcinoma of the ovary, with peritoneal carcinomatosis, initially underwent standard surgical staging followed by 6 cycles of adjuvant platinum-based chemotherapy. She subsequently recurred one year later and required multiple lines of chemotherapy. While on her 6th line of therapy with single agent paclitaxel, she presented with unsteady gait, vomiting, and worsening daily debilitating headaches not relieved by medication. An MRI of her brain showed areas of enhancement in the subarachnoid spaces concerning for LC, including diffusely along the cerebellar folia (with areas of adjacent edema in the cerebellum) (Figure 1). There were also areas of enhancement in the bilateral internal auditory canals and partially coating the surface of the brainstem and superior visualized cervical spinal cord. The patient began palliative brain radiation and received 3000cGy in 10 fractions but subsequently passed two weeks later.

Images

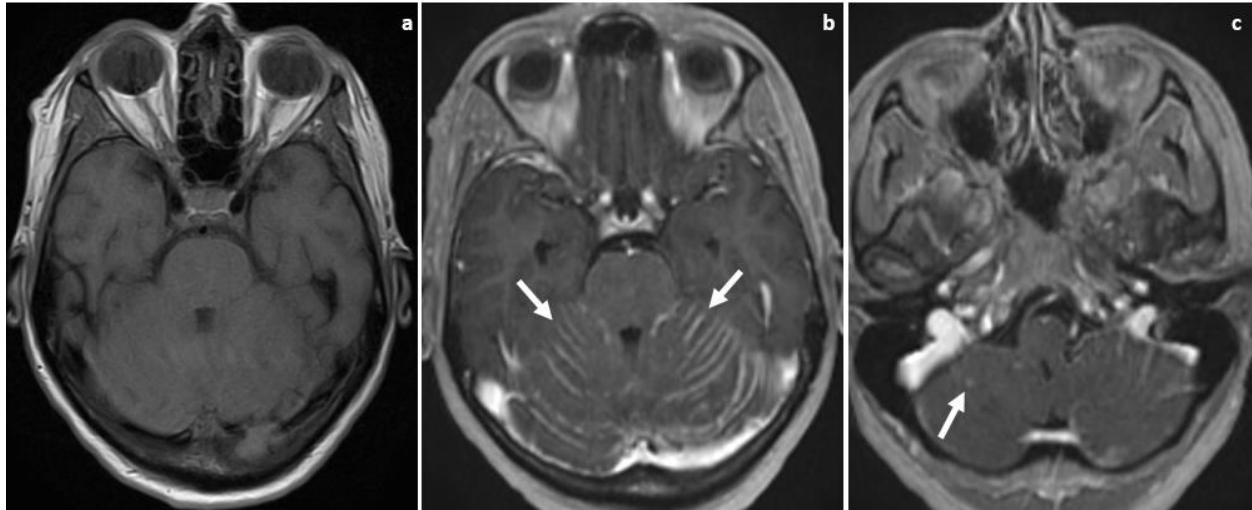


Figure 1: MRI of the brain. Pre-contrast T1-weighted sequence (a) demonstrating no hyperintense signal within the cerebellum. Post-contrast T1-weighted sequence (b) showing leptomeningeal enhancement within the cerebellar folia (white arrows). Post-contrast T1-weighted sequence (c) showing nodular small foci of enhancement (white arrow) concerning for solid intraparenchymal metastasis.

Discussion

The incidence of LC resulting from metastasis of a primary ovarian cancer is less than one percent. After confirmation of leptomeningeal involvement, the median survival is 60 days. More than 50 cases have been described in the literature; however, the majority of these are small case studies, limiting the available data required to develop a formal treatment regimen. In 2020, a single-arm, open-label phase 2 trial of pembrolizumab in patients with LC showed an improvement in three-month survival, however, the sample size was small. Chen et al. examined 19 patients treated at a cancer center in the United States between 1985 and 2002 with ovarian cancer brain metastases. They found that patients who underwent gamma knife radiosurgery (GKRS) in addition to whole brain radiotherapy, resection, or chemotherapy had a median survival of 23 months, compared to 4 months for those who did not receive GKRS. Due to the rarity of LC and the resulting limitation in conducting robust treatment trials, a standardized treatment protocol has not yet been established. The primary approach to treatment continues to involve a combination of surgical resection, radiation, and chemotherapy, encompassing various modalities. In the case of our patient, she began receiving palliative radiation to the brain shortly after the diagnosis of LC. Unfortunately, she passed away just 14 days after the diagnosis.

Conclusion

LC originating from a primary ovarian portends a fatal prognosis and short survival time after being diagnosed. Due to the limited available research and rarity of LC from primary ovarian cancer, further investigation and larger-scale studies are necessary to develop a more comprehensive understanding of the optimal treatment strategies for the patients.