

A Rare Case of Ewing's Sarcoma of the Ovary Treated With Proton Therapy – A Case Report

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Abstract

Background: Primary Ewing's sarcoma of the ovary is an exceptionally rare malignant small round cell tumor, typically affecting young women. It belongs to the Ewing sarcoma family of tumors, characterised histologically by small round blue cells, CD99 positivity, and the EWSR1-FLI1 gene fusion. Diagnosis relies on histopathology, immunohistochemistry, and molecular testing. Management parallels skeletal Ewing's sarcoma, combining surgery, systemic chemotherapy and selective radiotherapy. Prognosis is guarded, though long-term survival has been reported with multimodal therapy.

Case Presentation: A 24-year-old woman presented in May 2019 with abdominal pain, nausea and stage I acute kidney injury. Imaging revealed a large right ovarian mass without metastases. She underwent fertility-sparing laparotomy including right salpingo-oophorectomy, infracolic omentectomy, ascitic drainage and lymph node sampling. Histopathology demonstrated monodermal teratoma with malignant transformation to peripheral primitive neuroectodermal tumor (Ewing's sarcoma). A single para-aortic lymph node contained a 1 mm metastatic focus.

Management and Outcome: She received postoperative VDC/IE chemotherapy as per EuroEwing protocol, followed by proton radiotherapy to pelvic and para-aortic nodal regions [45 Gy/25 fractions (RBE)]. Proton therapy was well tolerated, with only mild (grade 1) gastrointestinal toxicity and no significant late adverse effects. She remains in remission as of April 2025, with a disease-free survival of 65 months.

Keywords

Ewing's sarcoma, ovary, proton therapy, peripheral primitive neuroectodermal tumor, case report.

