Metastatic ALK-Positive Inflammatory Myofibroblastic Tumor of the Pelvis Eight Years After Uterine STUMP

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Case Presentation

A 59-year-old woman with a history of uterine smooth muscle tumor of uncertain malignant potential (STUMP), diagnosed in 2017 and treated with ovarian-sparing total abdominal hysterectomy and bilateral salpingectomy, presented eight years later with abdominal and flank pain. Imaging at an outside hospital demonstrated a right pelvic mass causing ureteral obstruction with associated hydronephrosis, prompting transfer to our institution for further evaluation. Computed tomography angiography revealed a right pelvic mass causing distal ureteral obstruction and poor enhancement of the right kidney. Serum tumor markers including CA-125, CEA, CA19-9, lactate dehydrogenase, and inhibin B were within normal limits.

The patient underwent multidisciplinary surgical management including exploratory laparotomy with tumor debulking, ileocecectomy with primary reanastomosis, extensive iliac artery dissection, and right nephroureterectomy. Histopathologic evaluation demonstrated an ALK-positive spindle cell neoplasm involving the ovary, distal ureter, small bowel, pelvic lymph node, and right iliac peritoneal mass. Immunohistochemical staining demonstrated strong diffuse ALK-1 positivity with expression of smooth muscle actin. consistent with inflammatory myofibroblastic tumor (IMT). Despite extensive cytoreductive surgery, the patient experienced short-interval recurrence with progressive locoregional disease involving pelvic structures and the urinary tract. She subsequently underwent repeat multidisciplinary surgery including salpingo-oophorectomy, enterolysis, vascular reconstruction, and ureteral reimplantation. Pathology again confirmed metastatic ALK-positive IMT. Given the metastatic disease burden and ALK positivity, systemic therapy with the *ALK* inhibitor, crizotinib was initiated.

Discussion

IMT is a rare mesenchymal neoplasm with intermediate malignant potential characterized by spindle cell proliferation and inflammatory infiltrates. Approximately 50–60% of cases demonstrate rearrangements of the anaplastic lymphoma kinase (*ALK*) gene, which serve as both diagnostic markers and therapeutic targets. Although many IMTs follow an indolent course, aggressive behavior and recurrence have been reported, particularly in abdominopelvic tumors. The present case illustrates delayed aggressive disease occurring eight years after an initial uterine mesenchymal tumor diagnosis, with multifocal metastatic involvement requiring extensive multidisciplinary surgical management. The identification of *ALK* rearrangements has expanded therapeutic options for patients with recurrent or metastatic disease. Targeted therapy with *ALK* inhibitors such as crizotinib has demonstrated meaningful clinical responses and is increasingly utilized in unresectable or progressive diseases. This case highlights the potential for delayed aggressive behavior in gynecologic IMT and underscores the importance

of prolonged surveillance, multidisciplinary care, and consideration of targeted therapy in patients with recurrent or metastatic disease.