Case Report Intravenous Leiomyomatosis with Radical Response to Crizotinib

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Summary

Intravenous leiomyomatosis is a rare disease characterized by intraluminal growth of benign smooth muscle into veins and/or lymphatic vessels. Commonly known for its growth within the uterine and pelvic venous system, it can also affect big vessels expreading the inferior vena cava up to the right atrium and pulmonary vessels. The management of unresectable or recurrent disease is not established and literature is scarce. Here we present a case of extensive IV leiomyomatosis with radical response to crizotinib, a selective inhibitor of ALK tyrosine kinase receptor.

Case Report

This is a 65 y/o woman presented with vaginal bleeding and abdominal pain and later diagnosed with pelvic intravenous leiomyomatosis. Despite innumerous surgical procedures, including a TAH-BSO, there was progression of the disease associated with chronic, uncontrolled pain. About three years from time of diagnosis, patient already had extensive disease and was evaluated by surgical oncology for surgical resection that deemed unclear benefit with a very high risk. Patient was seen at oncology clinic and decision was made for next generation sequencing of her tumor. It showed to be to Alk positive on IHC and RNA sequencing detected a fusion protein PPP1R21-ALK on exon 20, what prompted the utilization of crizotinib. Crizotinib was started on a 250mg daily dosage as an oral medication on 3/2020 and on a follow consult in a month from its start patient reported drastic change in her complaints with complete improvement of pain and swollen of lower extremities. Most recently, after being 4 months on therapy, patient reported maintenance of pain control as well as complete resolution of prior chest pain discomfort related to the tumor invasion of the IVC.

Conclusion

Personalized medicine is an evolving field and, as it grows, starts to play an important role in cancer prevention, diagnosis, treatment as well as prognosis. Here we report a successful use of precision medicine, specifically an ALK inhibitor, after the detection of this mutation on NGS of the tumor. This demonstrates a new possibility for this type of tumor, never previously reported, and creates the need for more studies on expression and molecular profiles of this possibly devastating disease.

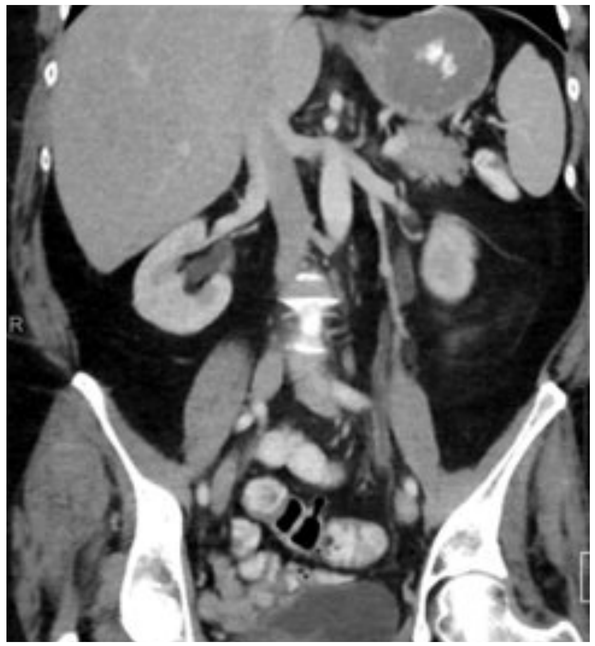

Figure 1: Coronal CT demonstrates a nodular enhancing mass in the right pelvis, abutting the urinary bladder and right pelvic sidewall. Note the normal appearance of the left gonadal vein and valve.

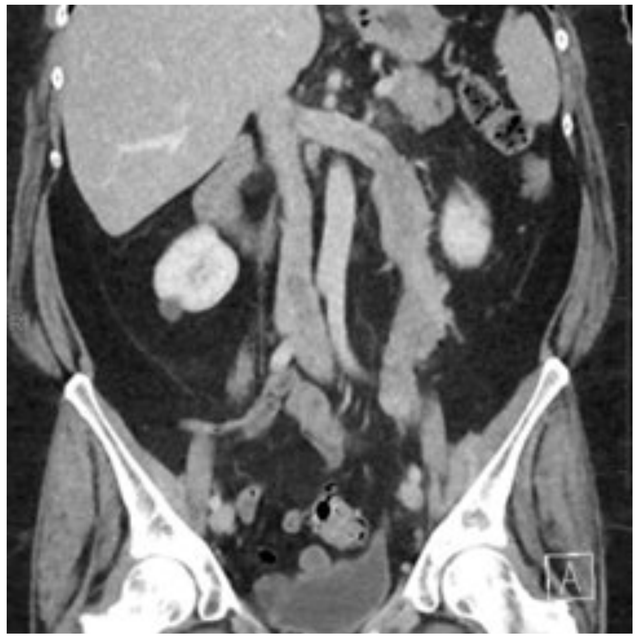

Figure 2: Coronal CT approximately 2 years after diagnosis shows marked expansion of the left gonadal vein with relatively hypodense intravascular tumor, and additional intravascular tumor involving the left common iliac vein at the confluence with the inferior vena cava .

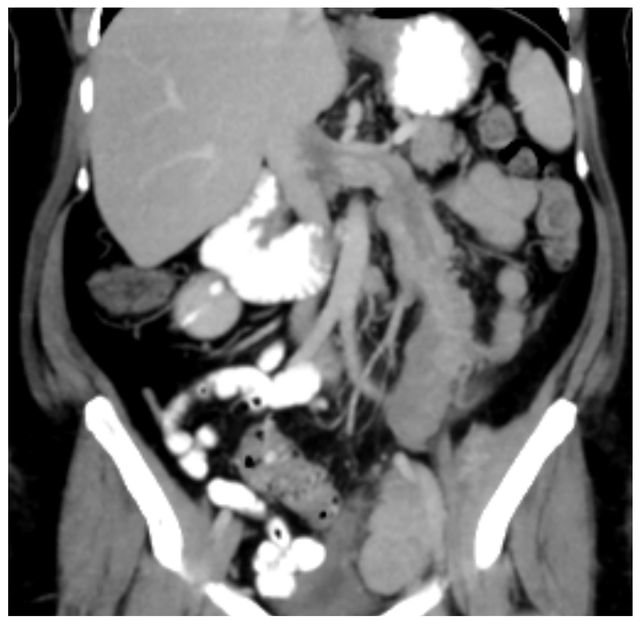

Figure 3: Coronal CT approximately 3 years after diagnosis shows continued marked expansion of the left gonadal vein with relatively hypodense intravascular tumor probably mixed with bland thrombus extending into the inferior vena cava; additional intravascular tumor involving the left iliac veins and branches in the left pelvis.

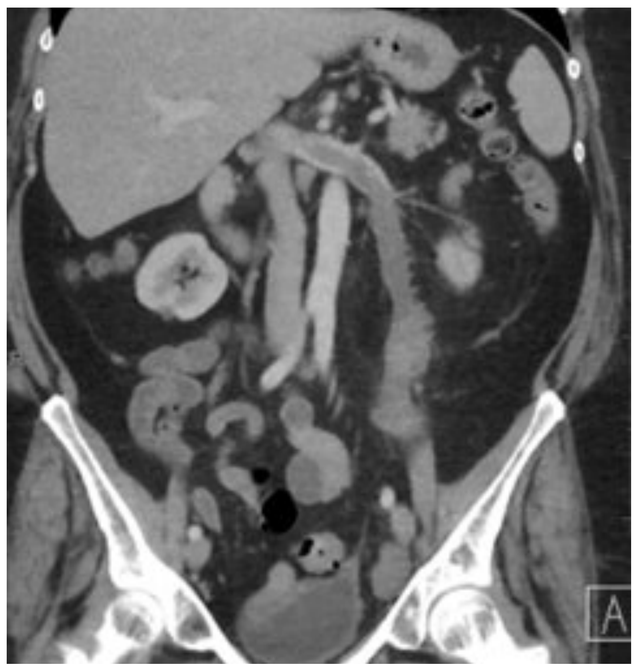

Figure 4: Coronal contrast-enhanced CT after treatment shows marked regression of the intravascular tumor in the left gonadal vein.