Retroperitoneal hamartoma in a pediatric patient

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A 13-year-old boy presented with vague abdominal pain and a palpable mass in the right abdomen. US revealed a large heterogeneous retroperitoneal mass (*M*), displacing the right kidney superiorly (*K*) (Fig. 1). Subsequently, a coronal enhanced CT showed a large heterogeneously enhancing mass with internal vessels (*arrow*) and areas of low attenuation (*curved arrow*) indicative of adipose tissue (Fig. 2). Also noted is the superiorly displaced right kidney (Fig. 2). Surgical pathology was consistent with a benign hamartoma, consisting of disorganized fatty, vascular, and fibrous components. Genetic analysis revealed a mutation in the phosphatase and tensin homologue (PTEN) gene.

Fig. 1 US

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Fig. 2 Coronal enhanced CT

PTEN is a tumor suppressor gene recently implicated in a collection of disorders termed the PTEN hamartoma-tumor syndrome [1, 2]. Affected individuals are at risk for developing various benign and malignant tumors [1, 2]. In addition, PTEN has been linked with vascular anomalies [2]. Given the extent of the tumor and the potential for substantial morbidity with surgical resection, surgical removal of the mass is currently deferred in this boy.

References

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