Fetal Rhabdomyomatous Nephroblastoma in a 31-Year-Old Woman: A Case Report

Ahlem Bellalah, Nouha Ben Abdeljelil, Manel Njima, Moez Hamdani, Wadii Hamdouni, Rym Hadhri, Adnene Moussa, Abdelfateh Zakhama, and Leila Njim

Fetal rhabdomyomatous nephroblastoma (FRN) is a rare variant of Wilms tumor with distinct morphologic features and biologic behavior compared to conventional nephroblastoma. It mainly occurs in patients under 4 years. In adults, extremely rare cases of nephroblastoma were reported. Among these cases, none has been interested a FRN. We report an exceptional case of a 31-year-old woman diagnosed with FRN discovered incidentally, to illustrate clinical and histopathological characteristics of this entity. UROLOGY 133: e5–e6, 2019. © 2019 Elsevier Inc.

Fetal rhabdomyomatous nephroblastoma (FRN) is a rare variant of Wilms tumor. In adults, extremely rare cases of nephroblastoma were reported. To the best of our knowledge, none of these cases was a FRN. We report an exceptional case of an adult diagnosed with FRN to illustrate clinical and histopathological characteristics of this entity.

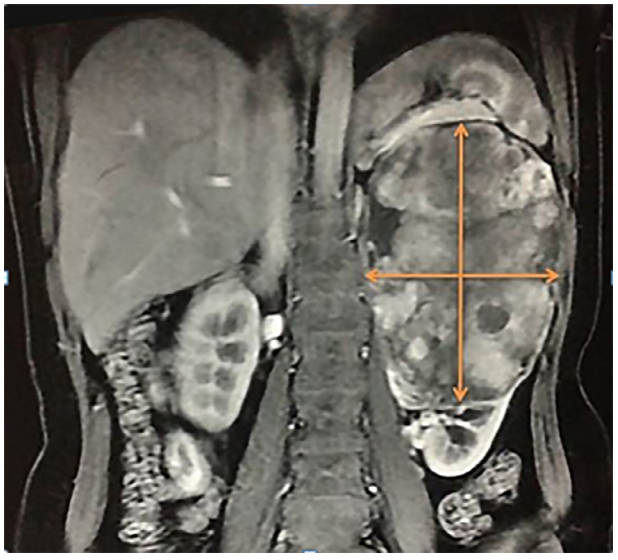

Figure 1. Magnetic Resonance Imaging (MRI): a solid left renal tumor with cystic changes.

CASE REPORT

A 31-year-old woman with no medical history was hospitalized for a left lumbar mass discovered incidentally by the patient after a slimming cure. Ultrasound examination revealed a well-circumscribed heterogenous tumor involving the upper renal pole. Magnetic resonance imaging showed a solid left renal tumor measuring 15 × 10 × 9 cm and containing cystic changes (Fig. 1). The diagnosis of nephroblastoma was suggested. A left nephrectomy was performed without neoadjuvant chemotherapy. The tumor was whitish on cut-surface with fibromyomatous appearance and several cystic and myxoïd changes (Fig. 2). Histopathologically, the tumor was well-circumscribed and mostly composed of fetal striated muscle cells in a myxoïd background (Fig. 3A). Both blastemal and epithelial components were absent. Rhabdomyomatous cells showed a

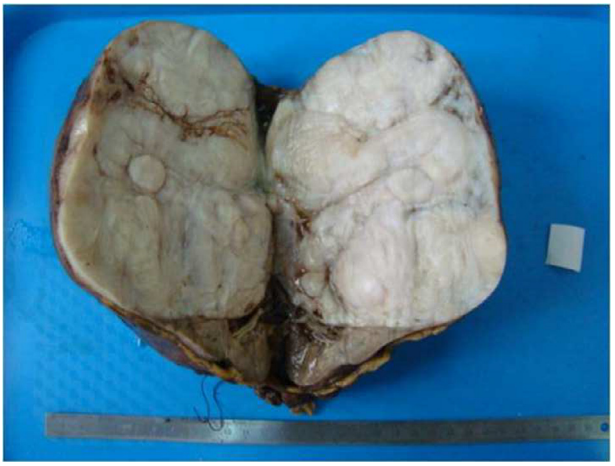

Figure 2. Macroscopic findings: a huge well limited mass with whitish and fibromyomatous appearance and cystic changes in the upper pole of the left kidney.

Conflicts of Interest: *None.*

From the Department of Pathology, Fattouma Bourguiba Hospital, Monastir, Tunisia; and the Department of Urology, Fattouma Bourguiba Hospital, Monastir, Tunisia

Address correspondence to: Moez Hamdani, M.D., Department of Pathology, Fattouma Bourguiba Hospital, 1st juin street, Monastir 5000, Tunisia.

E-mail: moez.hamdani@gmail.com

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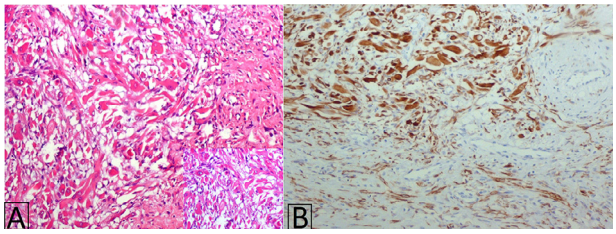

Figure 3. (A) Microscopic findings: the tumor is chiefly composed of fetal rhabdomyomatous cells with central nucleus loosely packed in a myxoid background (HE × 100). Higher magnification of rhabdomyomatous cells is at the right lower corner (HE × 400). (B) Immunohistochemical findings: strong immunostaining of rhabdomyomatous cells with Desmin.

strong immunostaining for desmin (Fig. 3B). The diagnosis of FRN was then confirmed. After a 30-month follow-up, no metastases or recurrences were detected.

CONCLUSION

FRN is a rare entity occurring exceptionally in adults. This variant is known to be chemo-resistant but to have a better prognosis compared with conventional nephroblastoma.