

Co-Existent Rhabdoid Tumor of the Brain and the Kidney in Newborn Girl: A Case Report

B. Zouita*, M. Ranib, B. Bannar, D. Basraoui, H. Jalal

Radiology Department, Hospital mère et enfant, CHU Mohamed VI, Marrakech

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*Corresponding author: B. Zouita

Abstract

Case Report

Malignant rhabdoid tumor of kidney (MRTK) is highly aggressive tumor of infancy and childhood. MRTK is unique in its significant association with primary brain tumours or brain metastases. We describe a case of malignant rhabdoid tumor of kidney, who first presented as posterior fossa brain tumor. Renal tumor was discovered incidentally during cerebral medullary MRI.

Keywords: Rhabdoid tumor, atypical teratoid rhabdoid tumor, imaging, mri.

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INTRODUCTION

Rhabdoid tumor was originally described by Beckwith and Palmer [1] In 1978 [2] as a variant of Wilms tumor with a rhabdosarcomatous component. Apart from kidney, rhabdoid tumors have been reported from many organs, including soft tissues and central nervous system (CNS) where it is referred to as atypical teratoid/rhabdoid tumor (ATRT)[1, 2].

Atypical teratoid/rhabdoid tumor (AT/RT) of CNS was first recognized in 1987, when it was simply referred to as “rhabdoid tumor”. It was defined as a distinct entity in 1996 and included in World Health Organization classification of CNS tumors in 2000. AT/RT currently constitutes one of three major CNS embryonal tumors in the 2007 World Health Organization classification of CNS tumors and is accorded World Health Organization grade IV due to its highly malignant nature [1]. We report a case of a newborn female with synchronous Rhabdoid tumor of the kidney and AT/RT.

CASE REPORT

Newborn female at day 20 of life, the mother had received no regular prenatal checkups, including

ultrasound examination; hence, no diagnosis was made during the prenatal stage. Pregnancy poorly followed estimated at term, referred to pediatric emergencies for a macrocrania evolving from birth, without other associated signs. A cerebral CT scan was requested which revealed a large infratentorial tumor seems to depend on the vermis, with a double cystic and tissue component, slightly enhanced, compressing the 4th ventricle and responsible for triventricular hydrocephaly (Fig 1), an MRI was requested to better characterize the lesion, which revealed an posterior fossa tumor originating from the vermis, with a tissue and fluid component, the tissue component of which is isosignal T1, intermediate signal T2, discreetly enhanced, in hyper diffusion with restricted ADC, measuring 59 x 62 x 64 mm, compressing the 4th ventricular with triventricular hydrocephaly, spectroscopy was in favor of a tumor profile with a peak in choline and decrease in NAA with a choline / NAA ratio > 2 (Fig 2), we completed with a medullary MRI to look for other localizations which revealed left renal tumor mass, heterogeneously enhanced, measuring 58 x 78 mm (Fig 3), hence a diagnosis of MRTK with coexistent AT/RT of the brain was established based on the clinical and the radiological presentation.

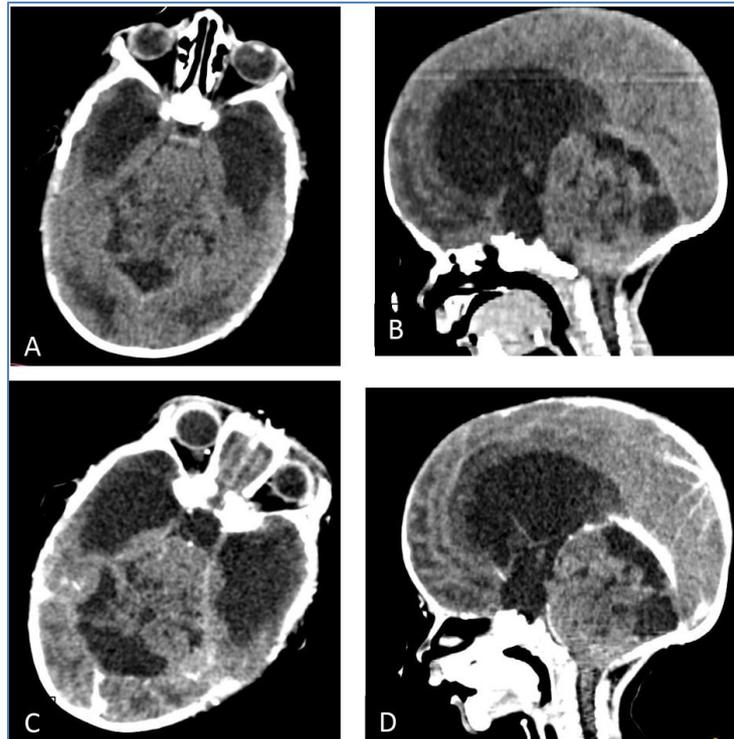


Fig-1: Computed tomography scan of the brain in axial (A and C) and sagittal (B and D) sections, without (A and B) and with contrast-enhanced (C and D) shows a large infratentorial tumor seems to depend on the vermis, with a double cystic and tissue component, slightly enhanced, with triventricular hydrocephaly.

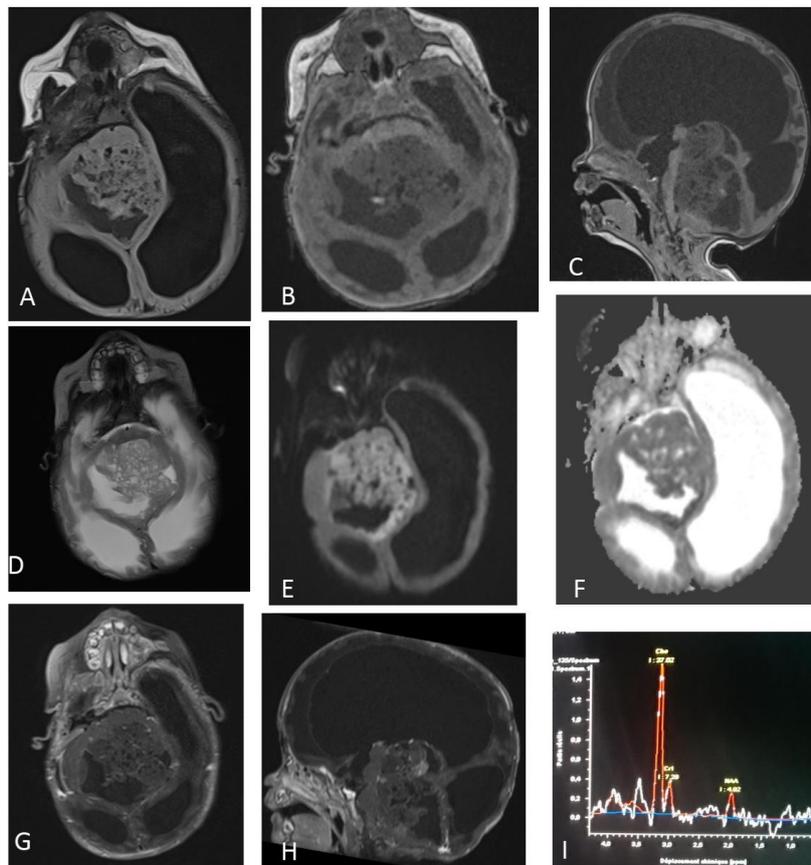


Fig-2: MRI brain images showing a tumor of posterior fossa with dual tissue and fluid components, the tissue part is in isosignal T1 weighted (B et C), intermediate signal in T2 weighted (D) and FLAIR (A), hyper diffusion (E) with restriction in ADC (F), heterogeneously enhanced (G and H), with triventricular hydrocephaly, spectroscopy is in favor of a tumor profile with a peak in choline and decrease in NAA with a choline / NAA ratio > 2 (I).

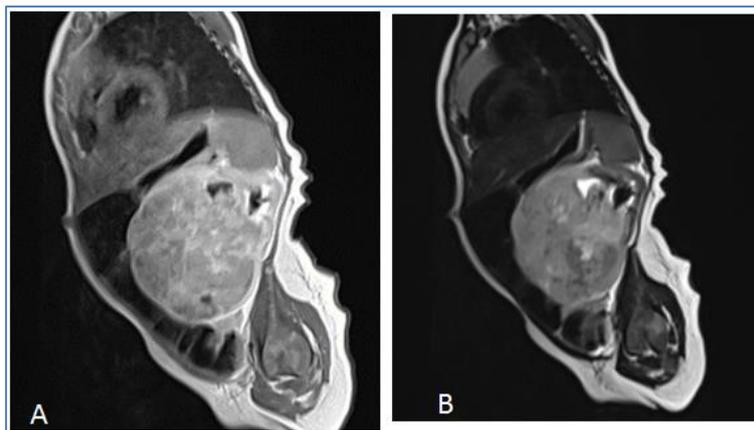


Fig-3: Medullary MRI on sagittal sections in T1 weighted (A) and T2 weighted (B) images shows an incidentally finding of renal tumor heterogeneously enhanced.

DISCUSSION

Renal tumors comprise 7-8% of all tumors under 15 year's children. Rhabdoid tumor of kidney (RTK) is a neoplasm of childhood once thought to be a sarcoma variant of Wilms' tumor [3]. It is now recognized as a distinct pathologic entity. RTK is a rare tumor, comprising about 2% of renal tumors of childhood. The tumor has a distinctive predilection for infants, with a median age of 11 months and over 80% of patients are under 2 years. A highly distinctive feature of this tumor is its association with early brain metastases [4]. Contrary to this, our patient had primary brain tumor AT/RT along with RTK. Thus, recommended brain imaging is crucial at the time of diagnosis. In the International Society of Pediatric Oncology, CNS involvement was only seen in 2.8% of the RTK patients because the intracranial evaluation was not a mandatory diagnostic procedure [6], in accordance with Reinhard *et al.*, only one patient was reported to have synchronous brain lesion among 32 patients of RTK [7] which adds rarity to our case.

Rhabdoid tumors can occur sporadically or as part of hereditary cancer syndrome known as Rhabdoid Tumor Predisposition Syndrome [4]. It has been suggested that most patients with both renal and central nervous system rhabdoid tumors have germ line mutations involving one copy of the hSNF5/INI1 gene [5].

Clinical features of AT/RT depend on the location of tumor and age of the patient. Patients usually present with symptoms of raised intracranial tension, including headache, vomiting, and lethargy, regression of developmental milestones, irritability, and macrocephaly in very young children. Computed tomography of brain demonstrates hyperdense lesion attributable to the high cellularity of the tumor and heterogeneous enhancement on postcontrast images. Calcification may be seen in up to 40% of tumors. Magnetic resonance imaging (MRI) is the imaging modality of choice in patients with AT/RT. On MRI,

AT/RTs show heterogeneous iso-intense signal on both T1- and T2-weighted MR images. Contrast-enhanced MRI shows variable enhancement (heterogeneous, peripheral nodular, intense, and mild). Peripherally located cystic components are commonly demonstrated on MR images and are a useful distinguishing feature. The main radiological differential diagnosis of AT/RT in the posterior fossa includes cerebellar astrocytoma, PNET/MB, brainstem glioma, and ependymoma [1-8].

Clinical presentation of RTK includes hematuria and loin/flank mass. Additionally, patients may develop hypercalcemia secondary to elevated parathormone levels [9], in CT, RTK appears as large, centrally located, heterogeneous soft-tissue masses, involving the renal hilum with indistinct margins, it is lobulated with individual lobules separated by intervening areas of decreased attenuation, relating to a previous hemorrhage or necrosis. Enhancement is heterogeneous. Calcification is relatively common, seen in up to 66% of cases, and is typically linear and tends to outline tumor lobules [10]. The main differential diagnosis of RTK is the far more common Wilm's tumor [11].

For RTK, Treatment consists of radical nephrectomy and resection of adjacent lymph nodes followed by chemotherapy. Survival is poor, with an 18-month survival rate of only 20%, and almost all patients succumbing before 5 years post-diagnosis [12], while for AT/RT Surgery with debulking can be offered in some cases [13].

Rhabdoid tumor has the worst prognosis of all renal tumors. It is highly aggressive and metastasizes early, with most patients presenting with advanced disease. Eighty percent develop metastases, most commonly to the lungs and less often to the liver, abdomen, brain, lymph nodes, or skeleton [12]. While for the AR/RT, the prognosis is much poorer than medulloblastomas, with little if any response to chemotherapy and death usually occurring within a year

of diagnosis [14]. The combination of the two tumors will definitely worsen the prognosis.

CONCLUSION

The association of an RTK with brain tumors either synchronously or secondary is described several times in the literature. Thus, infants and very young children presenting with kidney tumors should be investigated for synchronous presence of brain tumor or vice versa.

REFERENCES

1. Biswas, A., Kashyap, L., Kakkar, A., Sarkar, C., & Julka, P. K. (2016). Atypical teratoid/rhabdoid tumors: challenges and search for solutions. *Cancer management and research*, 8, 115.
2. Tahir, F., Majid, Z., Qadar, L. T., Abbas, A., & Raza, M. (2019). Co-existent rhabdoid tumor of the kidney and brain in a male infant: a rare case. *Cureus*, 11(8).
3. Vujanic, G.M. (2006). Renal tumors in early life. Current diagnostic pathology second edition. *Mosby, Philadelphia*, 12; 210-9.
4. Shukla, D., Pradhan, A., Bhardwaj, M., & Malhotra, V. (2015). Malignant rhabdoid tumor of kidney and brain in an infant. *Indian pediatrics*, 52(1), 65-66.
5. Mehrazma, M., HOUMAN, N., & SABER, A. M. (2010). Rhabdoid Tumor of Kidney Associated With Brain Mass; Case Report and Review of Literature.
6. van den Heuvel-Eibrink, M. M., van Tinteren, H., Rehorst, H., Coulombe, A., Patte, C., de Camargo, B., ... & Vujanic, G. M. (2011). Malignant rhabdoid tumours of the kidney (MRTKs), registered on recent SIOP protocols from 1993 to 2005: a report of the SIOP renal tumour study group. *Pediatric blood & cancer*, 56(5), 733-737.
7. Reinhard, H., Reinert, J., Beier, R., Furtwängler, R., Alkasser, M., Rutkowski, S., ... & Graf, N. (2008). Rhabdoid tumors in children: prognostic factors in 70 patients diagnosed in Germany. *Oncology reports*, 19(3), 819-823.
8. Cheng, Y. C., Lirng, J. F., Chang, F. C., Guo, W. Y., Teng, M. H., Chang, C. Y., ... & Ho, D. T. (2005). Neuroradiological findings in atypical teratoid/rhabdoid tumor of the central nervous system. *Acta Radiologica*, 46(1), 89-96.
9. Lowe, L. H., Isuani, B. H., Heller, R. M., Stein, S. M., Johnson, J. E., Navarro, O. M., & Hernanz-Schulman, M. (2000). Pediatric renal masses: Wilms tumor and beyond. *Radiographics*, 20(6), 1585-1603.
10. Chung, C. J., Lorenzo, R., Rayder, S., Schemankewitz, E., Guy, C. D., Cutting, J., & Munden, M. (1995). Rhabdoid tumors of the kidney in children: CT findings. *AJR. American journal of roentgenology*, 164(3), 697-700.
11. Chung, C. J., Lorenzo, R., Rayder, S., Schemankewitz, E., Guy, C. D., Cutting, J., & Munden, M. (1995). Rhabdoid tumors of the kidney in children: CT findings. *AJR. American journal of roentgenology*, 164(3), 697-700.
12. Lowe, L. H., Isuani, B. H., Heller, R. M., Stein, S. M., Johnson, J. E., Navarro, O. M., & Hernanz-Schulman, M. (2000). Pediatric renal masses: Wilms tumor and beyond. *Radiographics*, 20(6), 1585-1603.
13. Rorke, L. B., Packer, R. J., & Biegel, J. A. (1996). Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood: definition of an entity. *Journal of neurosurgery*, 85(1), 56-65.
14. Biswas, A., Kashyap, L., Kakkar, A., Sarkar, C., & Julka, P. K. (2016). Atypical teratoid/rhabdoid tumors: challenges and search for solutions. *Cancer management and research*, 8, 115.