Rajendra S. Garbyal · N. C. Aryya
A. N. Gangopadhyya · D. K. Gupta

Cystic atypical mesoblastic nephroma

Accepted: 27 November 2000

Abstract Cystic variants of atypical mesoblastic nephromas are very rare. The present communication deals with two such cases encountered in 3- and 6-month-old patients. The literature is briefly reviewed. The need for proper diagnosis of this tumor to distinguish it from cystic nephroma and Wilms’ tumor is highlighted.

Keywords Cystic atypical mesoblastic nephroma · Wilms’ tumor · Cystic nephroma

Introduction

Bolandé et al. [1] first recognized differentiated mesenchymal tumors of the kidney of infancy, which they designated as congenital mesoblastic nephroma (MBN). It showed a remarkably good prognosis compared to nephroblastoma (NB) when treated by adequate local excision [2, 3]. Series of retrospective and prospective studies on NB have expanded the concept of MBN, specifically as cases with more cellular or atypical histology are now being recognized. This raises the question of treatment modalities for MBN.

Due to the rarity of these tumors, most pathologists have limited experience and may encounter diagnostic problems with an atypical variant. We report two cases and our experience with atypical cystic variants of MBN.

Case reports

Case 1

A progressively increasing mass in the right abdomen was found in a 3-month-old male. Ultrasonography (US) showed a cystic renal mass, and IV pyelography (IVP) showed a non-functioning right kidney. Fine-needle aspiration cytology (FNAC) yielded only fluid. A right nephrectomy was done with a clinical impression of cystic nephroma/Wilms’ tumor (WT).

Case 2

A 6-month-old male presented with a right renal mass and failure to thrive. The mass was 14 x 10 cm and bimanually palpable. US showed a cystic right kidney mass with sepa formation. IVP showed a lower-pole cystic mass with distortion of the calyx. FNAC revealed fluid mixed with inflammatory cells. Exploration ended as a nephrectomy because of the impression of a WT.

Pathology

The specimens measured 10 x 8 and 14 x 10 cm respectively. The cut surface in both cases revealed solid and cystic areas with kidney tissue at the periphery. Small areas of hemorrhage and necrosis were also present (Fig. 1). Microscopically, hematoxylin and eosin-stained sections from both neoplasms showed similar findings. Solid areas exhibited cellular, plump, spindled-shaped atypical cells with high mitotic activity forming interdigitating fronds with entrapped glomeruli and tubules (Figs. 2–4). Cystic spaces of variable size and shape were lined by flattened epithelium (Fig. 5). Tumor cells were present between the cystic spaces (Fig. 6). Blastemal cells or immature tubules and skeletal muscle were not seen. Both cases were diagnosed as cystic atypical MBN.

Both patients underwent complete nephrectomy. In view of the atypical nature of the pathology, both patients received postoperative chemotherapy for 6 months. After follow-up periods of 3 and 2 years, respectively, there was no recurrence or metastasis.
**Discussion**

MBN accounts for nearly 90% of renal tumors diagnosed in the first 3 months of life, and the overall incidence is about 2.8% [4, 5]. The rarity of these tumors was assessed by Sandstedt et al., who reported 29 pure mesenchymal tumors in a total of 889 reviewed cases of renal tumors [6].

BNs are usually diagnosed in early infancy (<3 months), presenting as a renal mass and often detected at prenatal US examination [7], but cases are reported in older age groups [8, 9] and even in adults [10, 11]. Whether these patients had the tumor since birth, growing slowly, or acquired it later, is unclear. There are reports where these tumors were detected in young children and adults during routine physical examination [8, 12]. Clinically, they cannot be differentiated from WT and cystic nephroma (CN). Sometimes they may be confused with adrenal tumors because clinically extra-adrenal (predominant) development has also been reported [11]. A history of polyhydramnios in infants in whom MBN is diagnosed and hypercalcemia are known findings. Polyuria is believed to be the cause of the polyhydramnios, but has been proposed that hypercalcemia may be the main underlying mechanism, because neonatal hypercalcemia has been detected immediately after birth and levels decrease after removal of the tumor [13].