CASE REPORT

A rare case of bilateral cystic partially differentiated nephroblastoma recurring as bilateral cystic Wilms tumour

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SUMMARY

Childhood cystic partially differentiated nephroblastoma (CPDN) is an uncommon renal neoplasm. Bilateral CPDN or CPDN co-existing with a cystic nephroma/Wilms tumour is extremely rare. Treatment of CPDN is by complete surgical excision. Although local recurrences are uncommon, distant metastases have not been described. We present a case of bilateral CPDN that, after complete excision, recurred as bilateral cystic Wilms tumour. To the best of our knowledge, this is the first reported case in the literature where a bilateral CPDN has recurred as bilateral Wilms tumour.

BACKGROUND

This is the first reported case in the literature where a bilateral cystic partially differentiated nephroblastoma (CPDN) has recurred as bilateral Wilms tumour.¹ This transformation suggests that there is a close relationship between cystic nephroma, CPDN and cystic Wilms tumour, where the degree of differentiation determines the type of neoplasm.

CASE PRESENTATION

A 9-month-old infant presented with fever and bilateral flank mass that was progressively increasing in size over the past 1 month. Examination revealed soft, cystic, mobile, non-tender, bimanually palpable and ballotable masses in the right and the left lumbar region.

INVESTIGATIONS

Imaging (ultrasonography and CT) revealed two well-circumscribed multilocular non-communicating cystic lesions arising from the upper and middle pole of the right kidney, and upper pole of the left kidney, with the right-sided lesion being the larger one (figure 1).

DIFFERENTIAL DIAGNOSIS

A provisional diagnosis of bilateral cystic nephroma/bilateral CPDN was made on the basis of imaging. Another differential that was considered was bilateral cystic Wilms tumour.

TREATMENT

The infant underwent bilateral partial nephrectomy in February 2013. Postoperative histopathological analysis revealed bilateral CPDN.

OUTCOME AND FOLLOW-UP

Postsurgery, the infant was well for 6 months, after which a follow-up scan revealed recurrent cystic mass in both kidneys. Excision biopsy revealed it to be cystic Wilms tumour. The infant underwent 18 weeks of postoperative chemotherapy and was well at the last follow-up.

DISCUSSION

Multilocular cystic tumours of the kidney in children include cystic nephroma and CPDN, which are two grossly and radiologically indistinguishable, histologically distinct tumours.² Bilaterality in either of the multilocular cystic tumours is extremely rare with less than 10 cases described.

Joshi and Beckwith proposed diagnostic criteria for cystic nephroma, which are as follows: (1) it forms a discrete mass; (2) the lesion is composed only of cysts and intervening septa; (3) these intervening septa are the only solid component of the tumour; (4) flattened, cuboidal or hobnail epithelia line the cysts and (5) the intervening septa are composed of fibrous tissue that may or may not contain well-differentiated tubules. The term CPDN was given for those lesions that resembled cystic nephroma grossly but contained immature/blastemal elements within the septa.³ The fact that the only solid component within these tumours was the intervening septum helped in its differentiation from cystic Wilms tumour, which had solid masses of nephroblastomatous tissue.⁴

Figure 1 CT of the abdomen showing bilateral tumours (pre-operative imaging).

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It has been proposed that cystic nephroma, CPDN and cystic Wilms tumour are different parts of the same disease spectrum with cystic nephroma and cystic Wilms tumour representing the benign and malignant ends, respectively, and CPDN being the transition between the two.³ This is analogous to the spectrum of differentiation described in the transition of ganglioneuroma to ganglioneuroblastoma and, finally, neuroblastoma. However, to date, the exact nature of the relationship between these three entities has not been elucidated.

Childhood CPDN is mostly seen in male children less than 2 years of age. Common modes of presentation include abdominal mass, abdominal pain and/or haematuria.¹ The treatment is partial nephrectomy without adjuvant therapy.⁵ Local recur-

rence without distant metastases has been described.⁶ Cases where CPDN has co-existed with cystic nephroma and Wilms tumour have been described in the literature.^{3 7} However, transformation of a CPDN into cystic Wilms tumour has not been described yet. This transformation suggests that there is a close relationship between cystic nephroma, CPDN and cystic Wilms tumour, where the degree of differentiation determines the type of neoplasm.⁸

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Learning points

- ▶ Multilocular cystic renal tumours of childhood include cystic nephroma and cystic partially differentiated nephroblastoma (CPDN).
- ▶ They are grossly and radiologically indistinguishable but histologically distinct.
- ▶ Bilaterality is extremely rare.
- ▶ Transformation of a CPDN into cystic Wilms tumour has not been described yet.
- ▶ This transformation suggests that there is a close relationship between cystic nephroma, CPDN and cystic Wilms tumour, where the degree of differentiation determines the type of neoplasm.

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