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Case Report

Cystic Nephroma Case Report

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Introduction

While cystic lesions of the kidney are common occurrence in the childhood, cystic neoplasms of the kidney are very rare and they present as a spectrum from benign to malignant lesions. Cystic nephroma is a renal tumour composed of multilocular cysts without blastemal or undifferentiated elements. It is considered to be related to cystic partially differentiated nephroblastoma and nephroblastoma (Wilms tumour). The clinical significance is its benign nature, which should be differentiated from other renal neoplasms of children [1]. They are difficult to diagnose radiologically and only histopathology of the tumour can differentiate between benign and malignant cystic lesions [2]. Here we report such a case of cystic nephroma and review the differential diagnosis.

Keywords: Nephroblastoma; Cystic; Pediatric

Case Report

A 13-month-old female child presented with a left sided abdominal mass since last one month. No other symptoms such as fever, vomiting, abdominal distension, abnormal bowel movements and respiratory distress were observed. There was no history of pain, rapid growth in the mass, haematuria and weight loss. On general physical examination vitals were within normal limits and no other syndromic features were present. On abdominal examination there was a large solitary, abdominal mass of size 8 x 7 centimetres, occupying left lumbar region and left hypochondrium not crossing the midline. There were no other masses in the abdomen and genitals and spine was normal. Ultra-sonogram was suggestive of a multiseptated cystic lesion adjacent to lower pole of left kidney of size 7.9 x 6.52 centimetres. Contrast enhanced CT scan of the abdomen showed a well-defined oval shaped cystic lesion of size 8.2 x 7.6 x 7.4 centimetres arising possibly from lower pole of the left kidney with thin rim of compressed renal parenchyma over the lesion. Multiple thin internal septation and loculations were present in the lesion. The lesion is reaching till the midline and displacing the bowel loops towards right side and compressing the upper ureter. Lymph nodes were not enlarged and renal vessel and vena cava were normal.

Considering the large size of the tumour and the possibility of malignant transformation, simple nephrectomy through anterior subcostal incision was performed. Grossly, the tumour was well circumscribed with a smooth surface and measured 9.4 x 8.5 x 7.4 cm in size. The cut surface revealed variable-sized cysts separated by thin intervening septa containing clear fluid. No solid component was found in it. The tumour did not communicate with the renal pelvis. The remaining compressed renal parenchyma was present as a thin rim around the mass. Microscopically, the tumour was well circumscribed and comprised of multiple cysts separated by septae in edematous myxoid stroma. The cysts were lined by flattened cuboidal epithelium. No atypia was seen. Final impression of left kidney cystic nephroma was made. Post-operative recovery was unremarkable and the child was discharged home after 4 days. Considering the benign nature of the mass no further treatment with chemotherapy or radiotherapy was given, however the child is in follow up since last one year and is in good health.

Discussion

Cystic nephroma is a rare, benign renal tumour. It has two peaks of age of onset, one in the childhood and the other in the adult population [1]. Most of the childhood cases have been reported in children between 3 months and 4 years of age, with a male predominance. The adult cases occur mostly between the 5th and 6th decade of life, with a female predominance [2]. The prevalence is difficult to determine due to the rarity and bimodal age of presentation. Most authors regard cystic nephroma of the childhood as a benign end of the spectrum of Wilms tumour with cystic partially differentiated nephroblastoma and nephroblastoma (Wilms tumour) on the malignant end. The terminology used in adult cases has the similar histopathologic features, but is considered to have a different origin and not associated with nephroblastoma or nephrogenic rests [2]. Cystic nephroma of childhood was first considered as a unilateral and sporadic neoplasm. However, bilateral and familial cases associated with pleuropulmonary blastoma have also been reported in recent decades [3]. The pathogenesis is uncertain. The DICER1 mutations may be the major genetic event in the development of cystic nephroma [4]. The clinical presentation is usually as a palpable abdominal mass incidentally found by parents or caretakers, or during routine physical examination. Ultra-sonographic findings are multiple anechoic spaces separated by thin septa. On computed tomography, it is a circumscribed and multilocular cystic tumour. However, the entire, or portions of tumour may appear solid on both ultrasonography and computed tomography, because of the aggregation of small-sized cysts. It is difficult to distinguish cystic nephroma from cystic partially differentiated nephroblastoma or other renal tumours with cystic change on radiological studies. The final diagnosis depends on the pathologic examinations [2-5].

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Grossly, cystic nephroma is a well-circumscribed tumour with a smooth surface. The cut surface reveals variable-sized cysts separated by fibrous septa. The cysts contain clear or yellow fluid. They may be herniated into the renal pelvis but do not communicate with it. No solid nodule should be found [5]. Tumour necrosis, haemorrhage, and calcification are extremely rare. The histologic diagnosis criteria were first established by Boggs and Kimmelstiel in 1956 and modified by Joshi and Beckwith in 1989 as multilocular cyst lined by epithelium in most of the part not communicating with the renal pelvis with essentially normal renal parenchyma and no mature elements should be present in the septa [6].

The differential diagnosis of other childhood neoplasms includes cystic partially differentiated nephroblastoma and solid tumours with cystic change such as nephroblastoma, clear cell sarcoma, congenital mesoblastic nephroma, and renal cell carcinoma. Occasionally, developmental disorder such as cystic renal dysplasia may also be mistaken as a cyst tumour [7]. Cystic partially differentiated nephroblastoma has a similar structure to cystic nephroma. However, blastemal elements, nephroblastomatous epithelial elements, and undifferentiated or differentiated mesenchyme are present in the septa. By definition, no expansive solid nodule exists in the tumour. Cystic partially differentiated nephroblastoma has a low malignant potential for local recurrence [7]. Surgery is curative in most of cases. By contrast, nephroblastoma is at the most malignant end of the spectrum with cystic partially differentiated nephroblastoma and cystic nephroma at the benign end. It is the most common renal malignant neoplasm of childhood and has the ability to metastasize and recur. Pre- and postoperative chemotherapy may be needed. Although haemorrhage or tumour necrosis can make cystic change, residual solid expansive areas can still be found [8]. Triphasic patterns composed of blastemal, epithelial, and stromal elements are the most characteristic pathologic findings. Sometimes, biphasic or monophasic tumour can also be observed. The prognosis depends on the stage and whether the anaplasia area is present or not.

Congenital mesoblastic nephroma may undergo cystic change, especially cellular type. Unlike cystic nephroma, the stroma is composed of interlacing fascicles of spindle cells with a much higher cellularity. Tumour cells dissect and entrap renal parenchyma with an infiltration border. Congenital mesoblastic nephroma has an excellent prognosis after complete excision. However, recurrence and metastasis have also been reported [9]. Clear cell sarcoma is a rare pediatric renal tumour with a propensity to metastasize to bone. Cystic change secondary to haemorrhage and necrosis has been reported [10]. Tumour cells can be epithelioid or spindled. Production of extracellular myxoid material that mimics clear cytoplasm is the characteristic of this tumour. Treatment requires nephrectomy followed by chemotherapy. Renal cell carcinoma occasionally occurs in children. The mean age at diagnosis is older than that of cystic nephroma. Similar to the malignant tumours described earlier, haemorrhage and necrosis can also make cystic change in renal cell carcinoma. However, it is usually not difficult to make a differential diagnosis of cystic nephroma based on the cytology and solid growth areas.

Cystic renal dysplasia is a disorder that occurs during embryo development. Most of the cases are unilateral. The affected kidney consists of irregular cysts surrounded by primitive mesenchyme. Cartilage and adipose tissue can be found. Glomeruli and renal tubules are scattered in the interstitium without an ordered distribution [10].

In conclusion, the clinical significance of cystic nephroma is its benign behaviour, and it should be distinct from other more common pediatric malignant neoplasms. Neither clinical presentations nor radiological studies can predict the histologic pictures of cystic renal tumour of childhood. Surgical intervention and histopathologic examination are necessary for the final diagnosis. Non-invasive follow-up is recommended after the complete resection.

Conflicts of interest – none to declare

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