Case Report

Metanephric Stromal Tumor: An Unusual Presentation of a Rare Paediatric Renal Neoplasm

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Abstract

A 2-year-old boy with prenatal diagnosis of a malformation uropathy was referred to paediatric surgery department. On systemic examination, there were no palpable masses. An ultrasonography of abdomen with color Doppler, a renal artery angiographic and scintigraphy revealed a preostial aneurysm at the left renal artery. The patient had a left nephrectomy. Grossly, the specimen measured 75 mm × mm 50 × 20 mm with renal artery aneurysm measuring 30 mm × 35 mm. On cut section, the renal parenchyma contained a whitish tumor that measured 35 mm × 10 mm. Histopathologically, this tumor was diagnosed as metanephric stromal tumor.

Keywords: Aneurysm, metanephric stromal tumor, paediatric neoplasm

Introduction

Metanephric stromal tumor (MST) is a rare, benign paediatric renal neoplasm that was reported for the first time by Argani and Beckwith in 2000 through a series of 31 cases.[1]

It occurs in the 1st year of life. The mean age at diagnosis is 24 months and it has rarely been described in adults.[1]

MST represents a spectrum of well-differentiated nephroblastic lesions with metanephric adenofibroma and metanephric adenoma that appear to be related to Wilms' tumor (WT).[2,3]

Its characteristic microscopic appearance and immunohistochemical profile helps to distinguish between MST and clear cell sarcoma of kidney (CCSK) and congenital mesoblastic nephroma (CMN).[1]

Recently, a cytogenetic characterization was described and it consists of a complex 17q rearrangement.[4]

A case operated for renal artery aneurysm and an incidental diagnosed MST will be presented herein and discussed.

Case Report

A 2-year-old boy with prenatal diagnosis of a malformation uropathy was referred to paediatric surgery department. On systemic examination, there were no palpable masses. An ultrasonography of abdomen and color Doppler were performed and they showed a vascular mass of the left renal helium measuring 25 mm × 21 mm with a vascular flow in the renal artery which pushed down the renal vein. These two diagnoses proposed a renal artery aneurysm and an arteriovenous malformation. An angiography of the renal arteries revealed at the left renal artery, a true preostial aneurysm measuring 7 mm × 12 mm. On renal scintigraphy, the left kidney had an altered function (glomerular renal function at 29%) with normal drain and the right kidney showed a good capture function (glomerular renal function at 71%). An endovascular treatment was attempted but it failed. Likewise, no vascular bypass gesture was possible; the patient had a left nephrectomy. Grossly, the specimen measured 75 mm × 50 mm × 20 mm with renal artery aneurysm measuring 30 mm × 35 mm. On cut section, the renal parenchyma contained a whitish area that measured 35 mm × 10 mm. Histological examination showed an unencapsulated tumor infiltrating the renal parenchyma and involving focally the renal sinus. It consisted of a proliferation of spindle cells with scanty

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cytoplasm and no nuclear atypia nor mitosis. Hypocellular myxoid areas were seen around blood vessels and tubules, forming concentric "onion skin" rings. Some intratumoral arterioles had myxoid changes of medial smooth muscle, characterizing angiodysplasia [Figure 1]. The glomeruli showed juxtaglomerular hyperplasia [Figure 2]. The wall of the aneurysm showed fibrosis with dystrophic calcifications. There were no nephrogenic rests. Immunohistochemically, tumor cells were diffusely immunoreactive for CD34, and there is no immunostaining with PS100, desmin, and cytokeratin [Figure 3]. This tumor was diagnosed as MST. There was no local recurrence after a following up of 4 months.

Discussion

MST is an uncommon benign tumor in children. It occurs in early infancy with a median age of 13 months and a peak at 2 years.[1] MST has occasionally been described in adults.[5] No bilateralism was reported.[6]

Its origin is not yet clear. Beckwith has suggested that MST may represent the result of maturation of intralobar nephrogenic rests with the loss of active blastemal component.[1,7]

Recently, some new kidney-specific entities have been identified (MST, metanephric adenofibroma, and anaplastic sarcoma of the kidney), and it is considerate that metanephric neoplasms represent a spectrum of differentiated lesions that seem to be related to WT.[2] In 2004, the WHO subdivided these tumors into a separate subhead based on cell of origin.[8]

The main presentation of this tumor is an abdominal mass followed by hematuria, recurrent urinary tract infection, fever, anemia, and hypertension.[1] It is rarely described as incidental finding like in our case where the patient was operated for a renal artery aneurysm.

Radiologic findings of MST are not detailed enough in the literature, and there is a difficulty in interpretation of ultrasonography because of the heterogeneity in this lesion that can be solid, cystic, or mixed.[3]

In the present case, the tumor was not identified on radiological examinations, indeed only an aneurysm of the renal artery was highlighted which is a consequence of angiodysplasia that is observed in MST.

MST is usually centered in the renal medulla;[7,9] however, in rare cases, it seemed to originate from renal cortex.[3]

Grossly, MST mainly presents as a solid nodular lesion centered in the renal medulla containing cysts[9] and rarely as a large cystic mass with a peripheral solid component.[3,6]

The characteristic histological features of MST include alternating cellularity giving to the tumor a nodular low power appearance, onion skin cuffing around renal tubules, angiodysplasia of the vessel wall, juxtaglomerular cell hyperplasia, heterologous differentiation, and patchy positivity for CD34 which is an additional nonspecific diagnostic tool.[1,4,10] The last two features are not present in our case.

The morphological differential diagnosis includes essentially CMN and CCSK.[4,7]

MST is differentiated from CMN by its scalloped, subtly infiltrative borders, in contrast with the deeply invasive character of CMN.[1,7] In addition, MST stains for CD34 while tumor cells in CMN are positive for desmin.[1,7] Cytogenetically, additional copies of whole chromosome 17, deletion on long arm of chromosome 3 (3q21q24), and duplication of the short arm of chromosome 11 (11p15) have commonly been reported in CMN.[11] In MST, a complex homogeneous gain of the segment between bands 17q22 and 17q25.3, resulting in partial triplication and duplication of the long arm of chromosome 17, was noticed.[4]

CCSK is characterized by the regular branching capillary vascular pattern which is absent in MST.[7] Furthermore, CCKS has a negative immunoreactivity for CD 34.[7] The CCSK has a cytogenetic characterization which consists of a recurring translocation between chromosome 10 and 17 (*t* [10; 17]) and a deletion on the long arm of chromosome 14 (14q).[12]

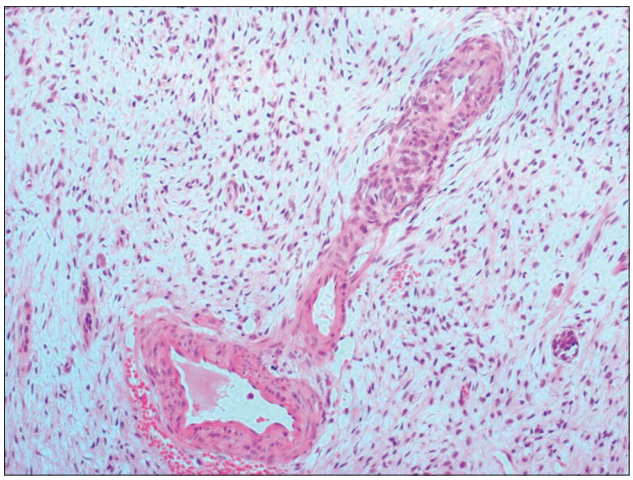

Figure 1: Intratumoral arterioles showing angiodysplasia (H and E, ×200)

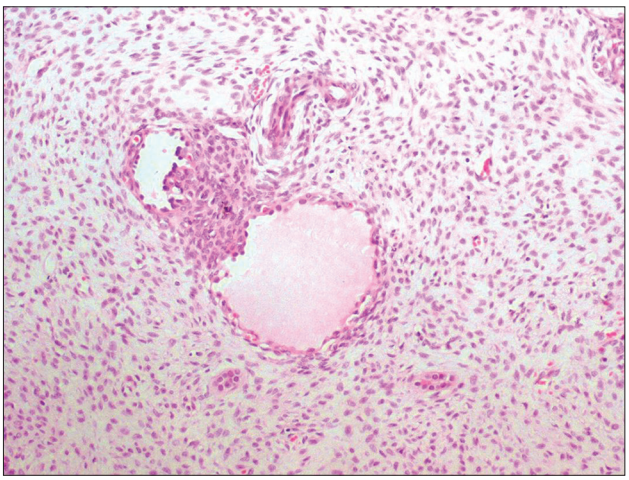

Figure 2: Proliferation of spindle cells with juxtaglomerular hyperplasia (H and E, ×100)

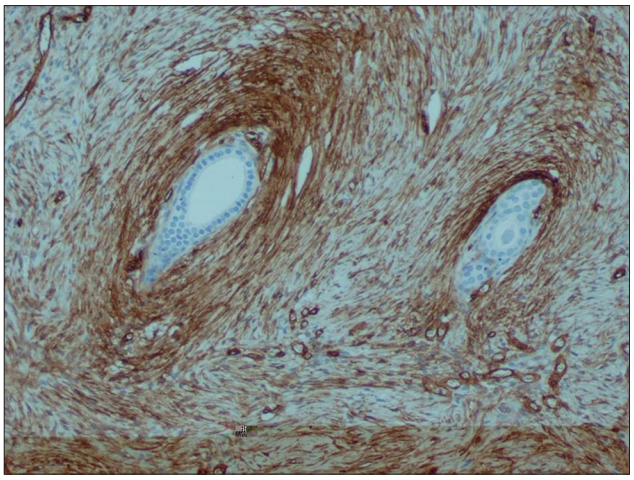

Figure 3: Tumor cells are immunoreactive for CD34 around tubules forming a concentric "onion skin" rings (×100)

To the best of our knowledge, the recently discovered complex cytogenetic rearrangement of 17q in MST has never been reported in other tumors.[4]

Indeed, it is very important to distinguish MST from these tumors because it has by far the most favorite outcome while CMN and CCSK are more aggressive and needs adjuvant chemotherapy in addition to surgery.[1,6] The CCSK metastasizes widely and cellular CMN occasionally metastasizes.[6]

The basic treatment of MST is a simple nephrectomy. Once the diagnosis is confirmed, no further adjuvant treatment is required.[7] There is no distant metastasis or local tumor recurrence so far.[6] The 5-year survival of this tumor following surgery is almost 100%.[6] The role of partial nephrectomy is not clear at this time.[7]

Conclusion

The incidence of MST is so miniscule. Its diagnosis is based on histopathological features and immunohistochemistry. Our report highlights an unusual case of MST that present as incidental finding when the child was treated for a renal artery aneurysm. This latter is a consequence of angiodysplasia which is observed in this type of tumor. The outlook following nephrectomy is excellent.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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