Metanephric stromal tumor, a rare pediatric renal neoplasm in Iranian children

Abstract
Metanephric stromal tumor (MST) is a very rare pediatric renal mass most occurring during infancy and childhood. Because MST is extremely benign, the correct diagnosis of this tumor saves the patient from side effects of additional therapy. We report a case of MST in a 3-year-old Iranian girl. To the best of our knowledge, this patient is the first case of MST reported in Iran. A renal ultrasound and a computed tomography scan were performed revealing a left renal mass. The patient underwent total nephrectomy. She had a recovery period without any complication. Specimen evaluation and immunohistochemistry demonstrated evidence of MST. The patient did not receive any post-operative antiangiogenic therapy with Bevacizumab until 1 year. She experienced an event-free survival.

Keywords: metanephric stromal tumor, pediatric renal neoplasm, congenital mesoblastic nephroma, children

Introduction
Stromal tumors comprise less than 20% of the pediatric renal neoplasms.1 Metanephric stromal tumor (MST) is a very rare pediatric renal mass most occurring during infancy and childhood. Surgical excision of the lesion alone is curative in most patients.2 The correct diagnosis of this benign tumor is important because it saves the patient from side effects of unnecessary chemotherapy or radiation therapy. The most presentation of MST is an abdominal mass. In this report, a MST presented with an asymptomatic mass during vesicourethral reflux (VUR) follow up in a 3 y-old girl.

Case report
3 years-old Iranian girl delivered via cesarean section was brought to the hospital by her parents with the complaint of agitation and crying during urination. There has history of fever but, without night sweats, weight loss, poor feeding, and hematuria. Physical examination had left flank tenderness. A renal ultrasound was requested, revealing left kidney gravel with hydronephrosis grade 2. VCUG revealed vesicourethral reflux. Blood tests were normal. The patient started on antibiotic therapy and nocturnal prophylactic antibiotic. Serial renal ultrasound was performed every 2 months until resolution occurred. After 2 years follow up during serial sonography she presented with a mass on left kidney. This mass had both solid and cystic compartments with coarse calcifications in right and left kidneys. The lesions had heterogeneous echogenicity. Bladder and urinary tracts were normal. Because the findings were highly suggestive of malignancy, the patient underwent nephrectomy.

No lymphadenopathy was detected. The bony skeleton had normal density and shape. There was no sign of abdominal free fluid. There was no evidence of distant metastasis (Figure 1 & 2).

Figure 1 Bilateral metanephric stromal tumor in 3 years old girl (CT scan view).

Figure 2 Proliferation of spindle cells juxtaglomerular hyperplasia (H & E X 100).

The specimen examination demonstrated that the 5×4×3.5-cm mass had a well-defined grayish firm. The mass was solid with small cyst-like spaces and ossified areas. Microscopically, sections showed a hypocellular fibrotic and sclerotic lesion containing scatter bland

Abbreviations: MST, metanephric stromal tumor; VUR, vesicourethral reflux; CK, cytokeratin; SMA, smooth muscle antigen, CMN, congenital mesoblastic nephromas; MAF, metanephric adenofibroma; MA, metanephric adenoma

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spindle cells. Several irregular-sized blood vessels were seen but no evidence of angiodysplasia was detected. Aggregation of the smooth muscle cells and inflammatory cells was detected. Focal ossifications were seen which was in accordance with calcifications detected on radiographic images. The surrounded renal tissue showed atrophy, interstitial inflammation, and fibrosis. Other heterologous elements were not seen. No mitotic figure was present. In the immunohistochemistry panel, spindle cells in the surrounding areas showed focal positivity for smooth muscle antigen (SMA). The renal tubules showed positivity for cytokeratin (CK). In sections taken from the edge of the tumor, heterologous elements like mature bone and adipose tissue were present.

Final pathologic diagnosis was metanephric stromal tumor. After total nephrectomy she received adjuvant antiangiogenic agents (Bevacizumab 10mg/kg every month until 1 year after nephrectomy).

No evidence of recurrence or metastasis was detected during the follow-up period and 9 months after discontinuation Bevacizumab.

Discussion

Renal tumors comprise less than 10% of prenatal neoplasms. Among them, congenital mesoblastic nephromas (CMN) is the most frequent renal tumor. For a long time, metanephric stromal tumor (MST) was categorized as CMN until recently, when this tumor was reclassified in a review study carried out on National Wilms Tumor Study Pathology Center files. Stromal tumors comprise less than 20% of the pediatric renal neoplasms. MST along with metanephric adenoma (MA) and metanephric adenofibroma (MAF) is a member of the renal metanephric tumor group. MA and MST are the most and the least aggressive members of this group, respectively. MA and MST are purely epithelial and stromal tumors, while MAF has both components.

Until now, less than 50 cases of MST have been reported. MST mostly occurs during childhood (median age 13 months, range 1 day to 11 years, peak age at 2 years), however, a few cases of MST have been reported in the adult population.

It is almost impossible to diagnose MST by radiologic findings alone. The diagnosis is made based on gross morphology and histologic evaluation of excised mass. In gross morphology assessment, MST is a yellowish-brown, unencapsulated mass localized around the renal medulla. The lesion is lobulated, fibrotic, solid or cystic. The lesion diameter varies from about 1.6 to 21 cm. Microscopically, the mass has nodular appearance and one or more of following components:

a. Onion skin ring formation of stromal spindle cells surrounding tubules or vessels,

b. Changes in entrapped vessels such as angiodysplasia of intramural vessels,

c. Presence of nodules of heterologous tissue including glia, cartilage, bone and fat,

d. Entrained renal elements by tumor cells.

Immunohistochemistry of specimen confirms the diagnosis. MST is positive for CD34 and vimentin and negative for desmin, cytokeratins and s-100 proteins. For example, CD34 positivity differentiates MST from CMN or clear cell sarcoma. MST is associated with excellent prognosis and surgical excision of the lesion by nephrectomy alone is curative in most patients. There is no need for additional therapy. Only two patients with MST have been reported, who underwent further cystectomy because of the tumor extension to the bladder and prosthetic urethra and orchietomy because of tumor recurrence. Because MST is extremely benign, there is no need for adjuvant chemotherapy. The correct diagnosis of this benign tumor is important because it saves the patient from side effects of unnecessary chemotherapy or radiation therapy.

MST usually presents as an abdominal mass or accidental finding. To the best of our knowledge, this patient is the first case of MST reported in Iran. A neonate patient has been reported who was presented with extra renal symptoms including hypertension and hypoglycemia. This tumor is usually located in the renal medulla. Until now, only a case of MST has been reported which had ossification of cystic walls in radiographic images. The microscopic evaluation of our patient specimen also demonstrated focal ossifications. Recurrence due to incomplete resection of the tumor has been reported in only one patient, the MST recurred as a gonadal tumor almost six months after nephrectomy and chemotherapy. During the follow-up period, our patient was well and had no sign of recurrence or metastasis.

Conclusion

Although MST is a very rare pediatric renal neoplasm, clinicians must consider this benign tumor in the differential diagnosis of childhood renal masses. Surgical excision of the lesion alone is curative because the tumor is extremely benign. MST is associated with favourable prognosis, so correct diagnosis based on macroscopic and microscopic evaluation and confirmation of diagnosis by immunohistochemistry saves the patient from side effects of additional therapy, but diffused angiodysplasia in tumor may be need to antiangiogenic agents.

Acknowledgments

None.

Conflicts of interest

The authors declare there are no conflicts of interest.

References