Wilms' Tumor with Metastasis to Duodenum – The First Reported Case in Thailand

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Abstract

A case of Wilms' tumor with gastrointestinal tract metastasis of the duodenum was documented in a 22 month - old - child. She had been diagnosed with Wilms' tumor stage III, treated surgically and by radiation and chemotherapy about 1 year previously. While she was still on chemotherapy, she developed hematemesis and clinical signs of partial gut obstruction. Gastroscopy and upper gastrointestinal series showed an intraluminal vascular mass in the duodenal bulb and histologically proved to be Wilms' tumor. We believe this is the first report in the world of Wilms' tumor with gastrointestinal tract metastasis.

Key word : Wilms' Tumor, Metastasis, Gastrointestinal Tract

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Wilms' tumor is the most common malignant renal tumor in children and adolescents and is second only to neuroblastoma for abdominal malignancy in childhood(1). For Thai children, it is the fifth most common malignancy following acute leukemia, brain tumor, retinoblastoma and neuroblastoma(2). Overall incidence in the USA was 7.6 / 1,000,000 in children below 6 years of age and peak incidence occurs between 2-5 years. Incidence rates in a number of Asian populations are less than half those in the population of most Western developed countries(3), regardless of sex(1). Unilateral involvement incidence (non heritable form) is 90-95 per cent compared to 5-10 per cent for

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the simultaneous bilateral (heritable form). Associated congenital anomalies (bilateral aniridia, hemihypertrophy, genitourinary anomalies) are 45 per cent in the simultaneous bilateral form compared to 4 per cent for the unilateral form[4].

Metastasis can occur by local invasion through perirenal fat to surrounding structures, through renal sinus, lymphatic and inferior vena cava. Distant metastasis by hematogenous route can spread to the lungs 85 per cent, liver 15 per cent, others which are considered rare such as bone marrow, salivary gland, tonsils, cortical bone and brain[5]. The most common presenting signs and symptoms are asymptomatic abdominal mass. Others are abdominal pain 25 per cent, hematuria 15 per cent, hypertension 25 per cent. Constitutional symptoms are fever, lassitude, anorexia and hypercalcemia. Treatment depends on staging and histological classification consists of surgery, radiotherapy and chemotherapy[6,7].

We report a case of Wilms' tumor with gastrointestinal tract metastasis which has never been mentioned before in the literature. Diagnosis was made initially by finding the tumor in the duodenal bulb and confirmed by histology later.

CASE REPORT

A 22 month - old - girl presented with vomiting for 10 days. She had been diagnosed with Wilms' tumor stage III when she was 10 months old presenting with a large abdominal mass. Ultrasound of the abdomen showed a solid tumor in the right suprarenal region with local invasion of the upper pole of the right kidney. An exploratory laparotomy was done with failure to resect the tumor due to a large suprarenal mass diameter 10X7X7 cms. which had invaded the upper pole of the right kidney. An attempt to resect it caused massive bleeding, so only biopsy could be done establishing histological diagnosis of Wilms' tumor and favorable histology. Treatment was initiated with irradiation and chemotherapy using Actinomycin-D and Vincristine for about 4 months until the mass was smaller. Reexploratory laparotomy and right nephrectomy was done successfully. After the operation, she was readmitted twice because of vomiting in the month following the operation, but her recovery went well. She was still on chemotherapy and her condition remained good until she developed vomiting with a small amount of coffee ground and was readmitted again at about 1 year after her successful operation.

Physical examination revealed a weak girl with mild pallor and no fever, The superficial lymphnodes were not palpable. The abdomen was flat with a surgical scar on her right side with no palpable mass. The rest of the examination was unremarkable.

Complete blood count revealed Hb 9.5 g/dl, Hct 29.9 per cent, WBC 11,700/mm^3, differential count and platelets were within normal limits. Urinary analysis revealed microscopic hematuria. Blood urea nitrogen was 21.08 mg/dl, creatinine 0.47 mg/dl. Liver function tests were also within normal limits. Electrolytes were sodium 138, potassium 3.31 chloride 88, calcium 2.5, bicarbonate 35.3 mEq/l. The chest X-ray had no infiltration. Gastroscopic findings on the second day of admission showed normal looking esophageal mucosa, the stomach contained a small amount of coffee ground content but no erosion or ulcer were seen. Looking through the pylorus, there was a round vascular intraluminal mass which occupied about three quarters of the luminal width of the duodenal bulb. The central ulceration was covered with exudate (Fig. 1). Advancing the endoscope further was unsuccessful due to the intraluminal mass. G-I follow through showed intraluminal filling defect as a round shaped mass in the duodenal bulb (Fig. 2).

Operation was unsuccessful in resecting the tumor mass due to its extension through the serosal and spillage into the subhepatic area. Only biopsy of the tumor and gastrojejunostomy were done. Histological findings of the duodenal tumor confirmed Wilms' tumor, blastemal cell type and all sections since the initial biopsy were reviewed

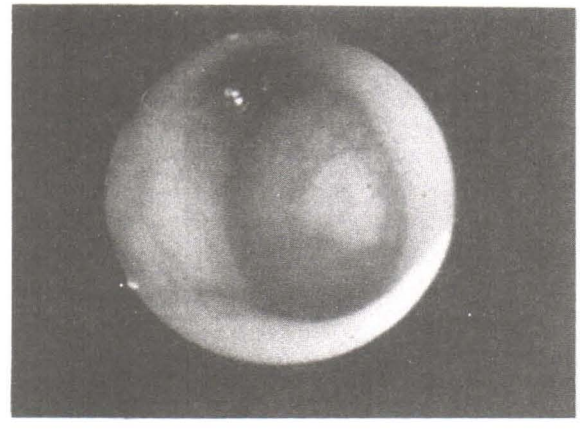

Fig. 1. Gastroscopic view tumor in duodenal bulb.

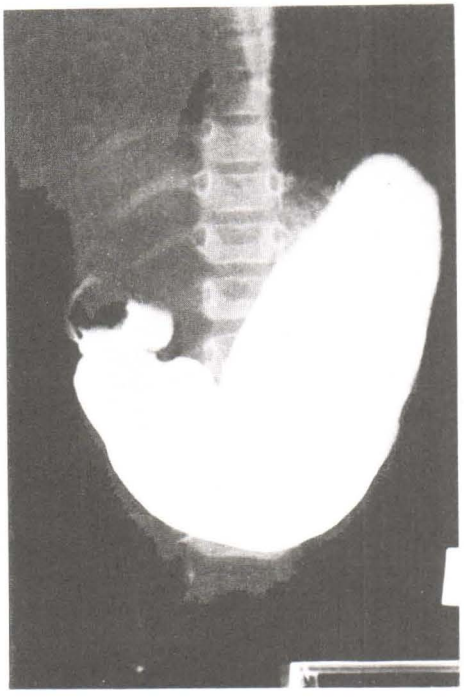

Fig. 2. Upper GI study filling defect in duodenal bulb.

again to confirm the diagnosis of Wilms' tumor with duodenal metastasis. The child's postoperative condition deteriorated and she eventually died.

DISCUSSION

Wilms' tumor originates from pluripotential mesenchymal cells of metanephric stroma, precursor tissue of mesoblastic stroma and primitive nephroblastic epithelium in embryonic staging consisting of blastemal, stromal and epithelial cell types (8,9). The National Wilms' tumor study divides the degree of cellular aplasia into favorable and unfavorable histology(7). This patient's histological classification was favorable. Neither bone, brain, lungs nor liver were found to be metastatic sites initially. Histological findings of the duodenal tumor was the same cell type as from the biopsied tumor at initial biopsy and from the nephrectomy at the second admission (blastemal cell type) (Fig. 3), so we confirmed this was not the second malignancy. Gastrointestinal tract metastasis has never been mentioned before for Wilms' tumor(10), so we believe this is the first report and the possibility of metastatic route is either hematogenous spread or seedling of tumor cells during the operation period.

In summary, we report the first case of Wilms' tumor with gastrointestinal tract metastasis in the duodenum to alert all paediatricians of the possibility of metastasis at this site.

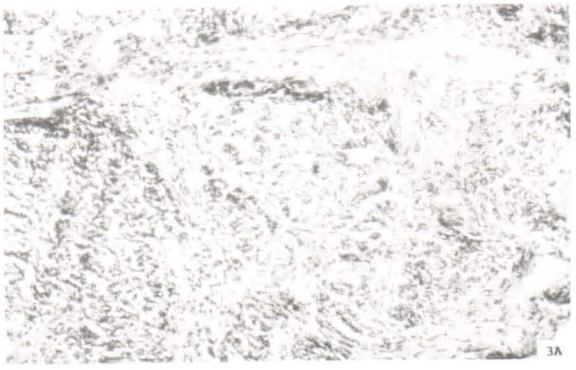

Fig. 3A. First biopsied tumor.

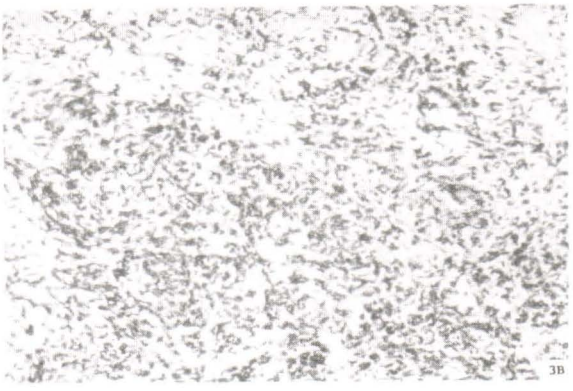

Fig. 3B. Nephrectomy.

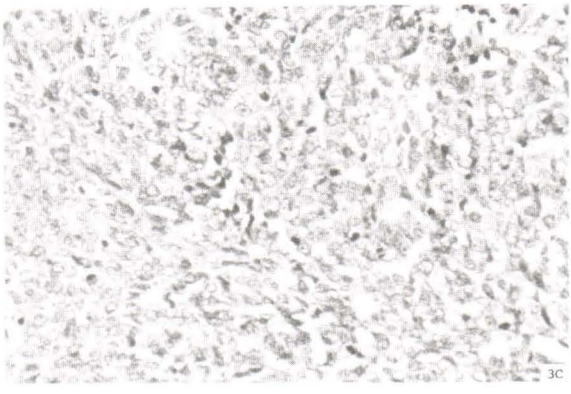

Fig. 3C. Tumor from duodenum.

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Wilms' tumor แพร่กระจายมาที่ระบบทางเดินอาหาร – รายงานผู้ป่วยรายแรกในประเทศไทย

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ได้รายงานผู้ป่วย เด็กอายุ 22 เดือน ซึ่งได้รับการวินิจฉัยว่าเป็น Wilms' tumor ระยะที่ 3 ซึ่งมีการแพร่กระจายไปที่ลำไส้เล็กส่วนต้น (duodenum) ผู้ป่วยได้รับการรักษาด้วยการผ่าตัด, รังสีรักษาและเคมีรักษาเมื่อ 1 ปี ก่อนและเริ่มมีอาการอาเจียนเป็นเลือดและลักษณะทางคลินิกของลำไส้เล็กส่วนต้นอุดตัน (duodenal bulb) ผลการตรวจทางพยาธิวิทยาของชิ้นเนื้อวินิจฉัยว่าเป็น Wilms' tumor ซึ่งน่าจะเป็นรายงานแรกในโลกซึ่งพบว่า Wilms' tumor แพร่กระจายมาที่ระบบทางเดินอาหาร

คำสำคัญ : Wilms's Tumor, แพร่กระจาย, ดูโอเดนัม

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