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CLINICAL QUIZ

Surprise diagnosis in an adolescent case with chronic kidney damage: Answers

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Answers

1. What is your diagnosis?

Histopathological evaluation resulted in signet ring cell carcinoma (SRCC) of the rectosigmoid region.

2. Is further investigation required for the diagnosis?

In the magnetic resonance evaluation of the abdomen, dilatation in the intrahepatic biliary tract, which is thought to be secondary to external compression, dilatation compatible with grade 2 hydroureteronephrosis in both kidneys, constriction secondary to external pressure or tumoral infiltration at the level of the ureter pelvis, a luminal mass lesion that holds the lumen in a 10-cm segment proximal to the rectum, and invasion of the mass lesion into the bladder posterior wall and distal segment of both ureters (Fig. 1).

Sigmoidoscopy and rectal endosonography revealed a malignant tumor occupying the lumen of the distal sigmoid colon and peritonitis carcinomatosa (Fig. 2).

Discussion

Hydroureteronephrosis is classified based on the presence or absence of reflux and obstruction. The variant with reflux but no obstruction is high-grade vesicoureteral reflux with a dilated ureter. In primary VUR, there is a failure of this anti-reflux mechanism because of a congenitally short intravesical ureter. Secondary VUR is frequently associated with an anatomic obstruction (for example, posterior urethral valves) or a functional

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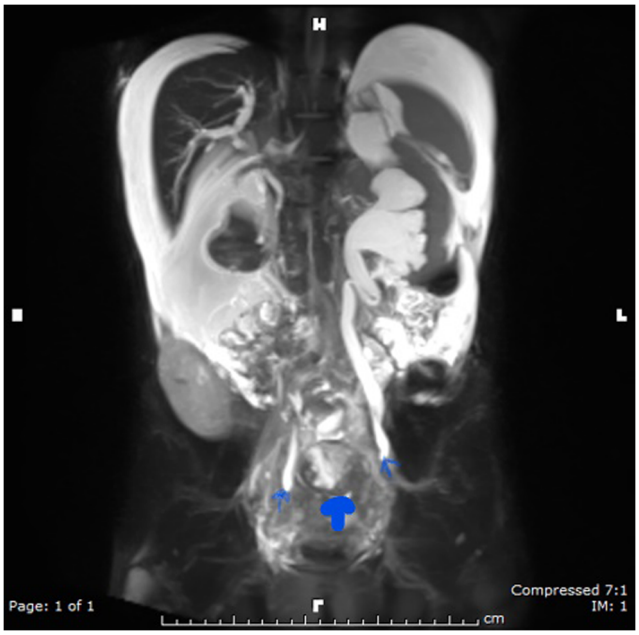

Fig. 1 Coronal MIP (maximum intensity projection) examination. A mass lesion narrowing the recto-sigmoid lumen was observed (thick arrow). Due to the spread of the mass to the mesorectum and ureters, there were stenosis and dilatation of both distal ureters (thin arrows). Intraabdominal widespread acid was observed

bladder obstruction (for example, bladder bowel dysfunction and neurogenic bladder). The severity of VUR can be influenced by the degree and chronicity of obstruction [1].

Colorectal signet ring cell carcinoma (SRCC) behaves aggressively and it is a poorly differentiated type of adenocarcinoma. The incidence of colorectal SRCC ranges between 0.1 and 2.6% for all cases according to the literature [2–5]. Most series report a predominance of the male sex, with ratios of 2:1. It is usually found in the right colon or the rectum [6]. At the time of diagnosis, around 20% of patients with colorectal

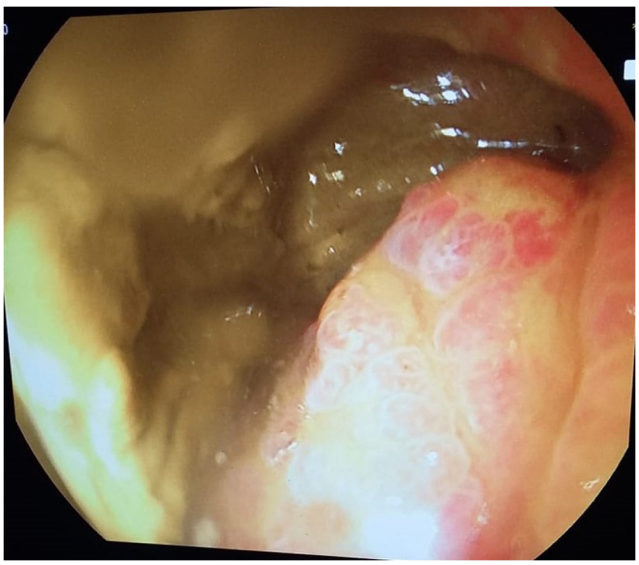

Fig. 2 A malignant tumor occupying the lumen of distal sigmoid colon

adenocarcinoma have distant metastases. The most common locations for the metastasis are the liver (77%), peritoneum (25%), and lungs (22%) [1]. Metastasis to the urinary system and particularly the ureter is rare. Less than five cases have been reported in the past 10 years [7].

Singh et al. [8] reported a 10-year-old patient who presented with abdominal distension, with a mass of distal sigmoid colon, diagnosed as SRCC. This is the youngest case of SRCC presented in the literature. The case is similar to our case regarding the location of the tumor, but hydronephrosis was not detected in that patient. Marone et al. [9] presented a 17-year-old patient with ascending colon involvement, who died 1 year after surgical treatment and chemotherapy. Another patient with peritoneal carcinomatosis similar to our case in terms of age and location of the tumor was reported previously [3]. A case diagnosed with chronic kidney disease (CKD), vesicoureteral reflux (VUR), hydroureteronephrosis, and subsequently diagnosed as SRCC has not previously been reported in the literature in any pediatric patient. While an adult patient was examined for hydroureteronephrosis, a sigmoid colon adenocarcinoma was diagnosed later. Hydronephrosis has been reported to develop due to the ureteral invasion of the mass [7]. Distant metastases at diagnosis in SRCC are usual. However, in our case, no metastasis other than in the peritoneum was found, suggesting direct peritoneal dissemination rather than hematogenous spread.

After histopathologic diagnosis of SRCC, chemotherapy was started as Modified FOLFOX 6 regimen that includes oxaliplatin 85 mg/m^2 day 1, leucovorin 400 mg total dose over 2 h day 1, fluorouracil 400 mg/m^2 bolus day 1, followed by 2400 mg/m^2 over 46 h and panitumumab 6 mg/kg every 2 weeks, was started. The patient is well after 3 months of follow-up. After chemotherapy, creatinine level decreased to 0.7 mg/dl and hydroureteronephrosis of the right kidney regressed, while hydroureteronephrosis in the left kidney persisted.

In summary, if progression in hydronephrosis is observed in patients with hydroureteronephrosis for a known cause such as VUR, intra-abdominal tumors should be kept in mind, including SRCC, a rare and rapidly progressing tumor in this age group capable of compressing the ureter.

Authorship contributions 1. Study conception and design—G.D., N.A., and R.S.; 2. Acquisition of data—G.D., N.A., R.S., H.Ş., and H.M.T; 3. Analysis and interpretation of data—G.D., N.A., F.B.Ç, and H.M.T; 4. Drafting of the manuscript—G.D. and N.A.; 5. Critical revision—G.D., N.A., F.B.Ç., H.Ş., and H.M.T.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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