Learning objectives to know:

• Congenital anomalies of the kidney and ureter

•Classification of renal cysts

• Classification, definitions, pathogenesis, and management of urinary tract infections

• Presentation and management of renal neoplasms

Introduction

The kidney as an organ has a lot of important functions some are secretory and others are excretory functions .the secretory regulatory functions of the kidney are more important than the execrtory functions ,in the fact that that humans can survive weeks of lack of excretory functions but losing the regulatory function cam be leathal if the patient lack it for hours.

The excretory function of the kidney include disposing nitrogemous waste products in the form of blood urea nitrogen,were as the regulatory function include regulating water balance in the body ,maintain electrolytes in the safe range in the circulation ,acid base regulation and control of blood pressure .

Anatomy

The kidneys are paired organs bean shaped roughly about the size of the fist of the hand measuring 10-12 cm length and 5-6 cm width ,covered by a tough connective tissue capsule the kidney capsule .the surface of the kidney is adherently covered on both sides by fat that cushon the kidney.the right kidney is lower down than the right ,the diaphragm cover the upper third of the kidneys were it is in close relation to the plura.

The blood supply to the kidney is by the renal artery in 75% of cases it is single and it arise from the lateral border of the aorta just caudal to the origion of the superior mesenteric artery.

CONGENITAL DISEASES

Renal agenesis Complete absence of one kidney occurs in 1 in 3000 live births. The other formed kidney is usually hypertrophic. Reproductive tract anomalies are common in females with unilateral renal agenesis. Bilateral renal agenesis is incompatible with life.

Ectopic kidney This occurs when the mature kidney fails to reach its normal location in the lumbar region. The incidence is 1 in 500–1200. An ectopic kidney may be found anywhere along the path of ascent: pelvic, iliac, abdominal, and rarely thoracic. When the ectopic kidney is located on the contralateral side to its ureteric insertion, it is called crossed ectopia. Renal ectopia may be associated with reflux in the ectopic or orthotopic kidney and with pelviureteric junction (PUJ) and ureterovesical junction (UVJ) obstruction.

Horseshoe kidney

This is the most common renal fusion anomaly, occurring in about1 in 400 live births with a male predominance. The isthmus lies at the level of the fourth to fifth lumbar vertebrae (fused lower poles). This causes failure to ascend and rotate so that the renal pelvis faces anteriorly and vertically with the malrotated calyces pointing postero-medially . The vascular supply is variable and the ureter may insert high on the renal pelvis. Most horseshoe kidneys are asymptomatic but they are associated with an increased incidence of genital anomalies, PUJ obstruction and stone formation. The incidence of Wilms' tumor is higher.

Multicystic dysplastic kidney

Multicystic dysplastic kidney (MCDK) is the second most common cause of an abdominal mass in newborns after hydronephrosis due to PUJO. The unilateral incidence is 1 in 1000–4000 live births. It has a 'bunch of grapes' appearance with multiple non-communicating cysts of varying sizes without identifiable renal parenchyma.
Presentation as abdominal mass

Ultrasound and isotop scan for diagnosis
Bilateral incompatible with life unilateral need no treatment in normal functioning kidney

Ureteral duplication

Duplication of the ureter and renal pelvis is a common anomaly, with an incidence of approximately 1 in 150 births. Unilateral duplication is six times more common than bilateral. It is more common in females. The duplication may be incomplete (Y-shaped ureter) or complete. It is associated with vesicoureteric reflux (VUR), PUJO and ureterocele. Incomplete duplex ureters with a 'Y' ureter arise when the ureteric bud bifurcates after its initial development from the Wolfian duct. Complete ureteric duplication occurs when there are two separate ureteric buds that develop into two separate ureters, which drain the upper and lower kidney moieties separately. The lower moiety ureter has a shorter submucosal tunnel and is prone to VUR. PUJO is more common with the upper moiety. The upper moiety ureter may be ectopic and is often associated with a concomitant ureterocele. The upper moiety of the kidney is often dysplastic.

Ectopic ureters

An ectopic ureter is one that drains to regions other than the bladder. Ectopic ureters are almost always associated with ureteric duplication and are bilateral in 10%. The female to-male ratio is 7:1. In females, the ectopic ureter opens either into the urethra below the sphincter or into the vagina Such a child would complain of incontinence of urine despite normal voiding. In contrast, the male child is always continent as the ureter opens above the external urethral sphincter. Computed tomo-urography (CTU) or magnetic resonance urography (MRU) is diagnostic.

Ureterocele

Ureterocele is a cystic enlargement of the intramural ureter, which probably occurs as a result of atresia of the ureteric orifice. It has a female-to-male ratio of 4:1 and occurs bilaterally in 10%. Similar to ectopic ureters, ectopic ureteroceles frequently drain the upper pole and are often associated with dysplastic or non-functional renal tissue. In childhood, they usually present with

infection. When large, they can obstruct the bladder neck or even the contralateral ureteric orifice. The classic feature of a ureterocele on an intravenous urogram (IVU) is the 'cobra head' sign. The treatment of simple ureteroceles is surgical excision with reimplantation of the ureter. Endoscopic incision of a ureterocele is the preferred treatment method for simple ureteroceles in infants and small children, but may result in subsequent ureteric reflux. A non-functioning kidney may need nephrectomy.

Congenital megaureter

The normal ureteric diameter in children up to 16 years is 0.50–0.65 mm. If the ureter is dilated by more than 7 mm, it is classed as a dilated or megaureter. This may occur with or without obstruction or reflux. UTI is the presentation ,ultrasound and ivu diagnostic treated surgically .

Congenital pelviureteric junction obstruction

Congenital PUJO is the most common cause of unilateral hydronephrosis with an incidence of 1 in 500 live births. It may result from intrinsic obstruction secondary to an a peristaltic segment at the PUJ due to muscular hypoplasia. Other causes include a high insertion of the ureter into the pelvis and the presence of crossing aberrant vessels at the PUJ. The Anderson–Hynes dismembered pyeloplasty is the procedure of choice with a wide funneled, dependent anastomosis.

RENAL CYSTS

Renal cysts can be broadly classified into sporadic, acquired, and genetic causes.
Sporadic renal cysts

Sporadic renal cysts are usually benign. Cysts with thin, sharply defined walls and clear fluid content are known as simple renal cysts. This category of cysts may be diagnosed with certainty by US. Apart from a few thin septa, any variation in the nature of the fluid, thickness of the cyst wall or septa or the presence of either calcification or a solid nodule would require further imaging with either computed tomography (CT) scan or magnetic resonance imaging (MRI) to rule out cystic renal cell carcinoma (RCC). Bosniak proposed a four-tiered classification of the malignant potential of cystic renal lesions. Category I cysts represent benign lesions that require no further follow-up, whereas

categories III and IV have a higher probability of malignancy and require surgical excision. Category II cysts can be safely followed up.

Acquired renal cystic disease

Most patients on hemodialysis develop bilateral renal cysts after 10 years. On follow-up one-fifth of these patients with acquired renal cystic disease (ARCD) develop renal cancers.

Genetic renal cysts

These cystic renal lesions have a known genetic inheritance. They are usually accompanied by involvement of other organ systems and present earlier in life than sporadic renal cysts.

Autosomal dominant polycystic kidney disease

Autosomal dominant polycystic kidney disease (ADPKD) is the most common autosomal dominant genetic cystic renal disease causing chronic renal failure requiring dialysis and renal transplantation, the patient might be a symptomatic but 50% of affected individuals eventually develop end-stage renal disease

Risk factors for the development of ESRD are:

● early age of presentation; ● hypertension;

● male sex;

● ADPKD gene 1;

● African ethnic group.

ADPKD is associated with cysts in other organs, such as the liver, pancreas, arachnoid membranes and seminal vesicles. It does not usually manifest before the age of 30 years and in some patients, it is never diagnosed. Renal symptoms include abdominal pain, hematuria or a palpable mass. Most patients older than 20 years are hypertensive and good control of blood pressure can delay progression to renal failure.. Intracranial aneurysms occur in approximately 10–30% of patients with ADPKD and subarachnoid hemorrhage may cause sudden death in young adults.

INFECTIONS

UTI is very common and affects all ages and both sexes. It can cause significant morbidity and is a rare cause of mortality in patients with serious comorbidities or in patients with urinary tract obstruction. Recurrent UTI is more common in women, affecting 30–40% in the sexually active age group. It can be defined as an inflammatory response of the urothelium (host) to invading bacteria. Asymptomatic colonization or bacteriuria is also common and can be differentiated from a UTI by the absence of symptoms and pyuria (leukocytes in urine).

Classification

UTI is classified as uncomplicated when it occurs in an immunocompetent host with an anatomically normal and functional urinary tract.

UTIs may also be classified on their site of origin as pyelonephritis (kidney), cystitis (bladder), urethritis or prostatitis.

While acute pyelonephritis indicates an acute infection of the kidney, chronic pyelonephritis is only a morphological description of previous infection-related sequelae such as scarring in the kidney as seen on radiological or nuclear imaging.

Acute pyelonephritis

This commonly occurs as a result of ascending infection from organisms in the lower tract, usually caused by Gram-negative bacteria. Hematogenous spread may be seen in patients with diabetes and in immunocompromised hosts, people who inject drugs and patients with bacterial endocarditis. It is more common in females, especially during childhood, at puberty, after intercourse and during pregnancy. Acute pyelonephritis usually presents with fever, chills, flank pain, nausea and vomiting . Loin tenderness may be present. Symptoms may vary from mild to severe illness with septic shock and renal failure. Pyuria is almost always present and its absence in a patient with pyelonephritis may point towards an obstructed urinary tract. Urine and blood should be collected for culture. Escherichia coli and other Gram-negative organisms are commonly responsible. Imaging is necessary when the patient is not responding to antibiotics to rule out pyonephrosis, renal abscess and obstruction. Renal US is

often the first imaging modality used. Contrast-enhanced CT (CECT) typically shows decreased patchy opacification of the affected parenchyma. Pyelonephritis complicating pregnancy The relaxing effect of progesterone during pregnancy causes ureteral smooth muscle relaxation and dilatation, presumably predisposing pregnant women to ascending upper tract infections. It is associated with fetal growth retardation and preterm delivery. Therefore, all pregnant women must be screened in the first trimester for ABU because, untreated, a third of these patients will develop UTI. Lower tract UTI typically occurs in the first trimester whereas pyelonephritis most often presents in the second or third trimester with acute abdominal pain or premature labor. Pyelonephritis is more common in pregnant women with an underlying urological abnormality or diabetes. A renal US is indicated if response to treatment is poor. Antibiotic use during pregnancy is tailored to avoid fetal harm and typically includes fosfomycin, penicillin's or cephalosporins.

Renal and perirenal abscess

A renal abscess results from an ascending UTI in association with an underlying urinary tract abnormality such as obstructive uropathy or VUR. It is usually caused by common uropathogens such as E. coli and other Gram-negative bacilli. Renal abscesses may extend and perforate the renal capsule to form a perirenal abscess. Multiple renal abscesses may conglomerate into a solitary suppurative lesion called a renal carbuncle. This is usually caused by Staphylococcus aureus, which reaches the kidney by hematogenous spread. The clinical presentation may be insidious and non-specifc but patients usually present with persistent fever, back pain, abdominal pain and costovertebral tenderness. Urine examination may be normal if the abscess does not communicate with the collecting system. CECT scan is the investigation of choice to establish the diagnosis. Treatment with antibiotics without drainage may be effective in carefully selected patients when the abscess is small (5 cm and in patients not responding to antibiotics. Open surgical drainage is indicated when percutaneous drainage is inadequate.

Emphysematous pyelonephritis

This is an acute-onset, rapidly progressive, possibly lethal form of pyelonephritis characterized by parenchymal necrosis and gas formation, caused

by organisms including E. coli, Klebsiella pneumoniae, Pseudomonas aeruginosa and Proteus mirabilis. Most patients have diabetes (up to 90%) and they may have obstruction secondary to calculi or papillary necrosis. Increased glucose levels in those with diabetes may provide a substrate for carbon dioxide production from fermentation. Symptoms are suggestive of pyelonephritis and an abdominal mass may be palpable. CECT of the abdomen is diagnostic and shows gas in the renal parenchyma, collecting system or both, along with other features of infection such as abscess, obstruction and perinephric stranding. Early diagnosis, intravenous broad-spectrum antibiotics and percutaneous drainage of the abscess and obstructed kidneys have improved outcomes in these patients. Emergency nephrectomy is rarely required and is reserved for patients who do not respond to the described measures.

Xanthogranulomatous pyelonephritis

Xanthogranulomatous pyelonephritis (XGP) occurs with severe renal infection in an obstructed kidney and is usually associated with calculi, causing loss of function and parenchymal destruction. Pathological examination typically shows accumulation of lipid-laden foamy macrophages. Patients may present with flank pain, fever with chills, persistent bacteriuria and a flank mass. A history of stone disease may be present. It is usually unilateral. CECT of the abdomen is diagnostic and shows a non-functioning enlarged hydronephrotic kidney around a shrunken pelvis with a calculus, also known as the bear's paw sign . Nephrectomy is the definitive treatment.

Tuberculosis of the urinary tract

Genitourinary tuberculosis (GUTB) accounts for 15–20% of extrapulmonary cases of TB. It is secondary and caused by hematogenous spread of tubercle bacilli from the thoracic lymph nodes or the lungs. GUTB occurs as a result of either reinfection or reactivation of old TB granulomas. Blood-borne organisms are deposited close to the glomeruli, causing an inflammatory reaction. Macrophages react and granulomas are formed. If bacterial multiplication goes unchecked, caseous necrosis results in the formation of tubercles. Multiple tubercles coalesce and rupture into the collecting system, causing intermittent tuberculous bacilluria and pyuria. The disease spreads through the collecting

system with ulceration initially. When bacterial multiplication is halted by the immune system, sequelae due to fibrosis appear. Tubercular obstructing or destructive lesions in the kidneys and ureters are responsible for renal function loss. Involvement of the bladder is secondary to renal disease. The disease gradually involves the bladder musculature, which is replaced by fibrous tissue, causing a decrease in the size and capacity of the bladder ('thimble' bladder). Urinary bladder involvement is responsible for urinary frequency, which is the most common symptom of GUTB. Epididymal tuberculosis presents as a painless epididymal nodule, usually involving the tail of the epididymis, or a chronic discharging sinus in the posterior scrotal wall. Patients may present with urinary frequency, colicky flank pain, hematuria and, rarely, fever and constitutional symptoms. They may also present with symptoms suggestive of recurrent UTIs and, rarely, calcified tubercular lesions may be misdiagnosed as urinary tract calculi. For microbiological confirmation, at least three consecutive early-morning specimens of urine are examined for acid-fast bacilli. The gold standard for microbiological diagnosis is urine culture. Nucleic acid amplification tests (NAATs) provide rapid diagnosis (within hours). When the diagnosis remains uncertain, bladder biopsy, tissue culture and tissue NAATs may be required. Imaging with CTU may also help and can show early signs such as calyceal distortion and papillary necrosis, hydronephrosis, poor function of renal segments secondary to parenchymal destruction, fibrosis and chronic obstruction. Ureteric strictures and proximal dilatation may also be seen. IVU can pick up the earliest signs of disease activity, such as calyceal distortion. Treatment involves short-course antituberculous therapy (ATT). Rifampicin, isoniazid and pyrazinamide are used sometimes with ethambutol as first-line drugs. The primary aim of therapy is preservation of renal function and avoidance of fibrotic sequelae. Ureteric strictures may require double J (DJ) stenting to preserve function until definitive reconstruction is attempted. Percutaneous nephrostomy (PCN) is recommended in obliterative strictures to achieve prompt decompression. Definitive surgery is usually done 3–6 weeks after starting ATT. The choice of reconstructive procedure depends on the type and location of sequelae. Open surgical repair is generally superior to balloon dilatation for tubercular ureteric strictures. Augmentation enterocystoplasty (usually using ileum) for small-capacity bladder ureteric reimplantation with or without a Boari fap (bladder tube) for lower ureteric stricture and ileal

replacement of the ureter for multiple long ureteric strictures may be required. Nephrectomy is done for major renal lesions with a poorly functioning kidney. Urinary infection in childhood and vesicoureteric reflux All children with UTI must be evaluated for underlying predisposing conditions as recurrent pyelonephritis can cause renal scarring and loss of renal function. UTIs account for 7% of childhood febrile illness. In the age group 1 year, it is more common in females. Structural and functional abnormalities of the urinary tract such as VUR and posterior urethral valves predispose to UTI. Reflux is considered primary when it is due to an incompetent UVJ and secondary when it is due to increased bladder pressure or outlet obstruction. Presenting symptoms in neonates and infants include febrile illness or sepsis and may not be localized to the urinary tract. The method of urine sampling, especially before toilet training, is crucial and may involve suprapubic aspiration or per urethral catheter collection. A bacterial count of 50 000 colony-forming units per milliliter is generally considered a positive culture result in children, although a lower count from a suprapubic aspirate in a symptomatic child is significant. The most important complication of UTI in a child is renal scarring secondary to renal parenchymal inflammation. US should be performed in all children, and children with recurrent UTIs or a first time UTI with pyelonephritis should be evaluated further. VCUG is the investigation of choice to diagnose reflux and should be performed in high-risk children. 99mTc dimercaptosuccinic acid (DMSA) radionuclide cortical scan is the best modality to detect parenchymal lesions. VUR is present in approximately 30% of children with UTI and in up to 90% of children with renal scarring. Renal scarring may cause hypertension in up to 20% and is an important cause of renal failure. The grades of VUR are summarized in Figure 82.9, with grades I–III generally resolving spontaneously. Low-dose nocturnal antibiotic prophylaxis to prevent scar-inducing pyelonephritis is the mainstay of treatment as the majority of reflux cases resolve with time. However, surgery (ureteric reimplantation, periureteric injections of Tefon or collagen) should be considered if episodes of acute pyelonephritis recur despite antibiotic therapy or if severe reflux is accompanied by a surgically correctable malformation such as a paraureteric bladder diverticulum.

TUMOURS OF THE KIDNEYS AND URETERS

Upper tract urothelial cancer Primary urothelial neoplasms of the renal pelvis and ureter are rare. They account for less than 10% of all urothelial tumors. They are more common in adult men. Important risk factors are tobacco consumption, occupations in the dye, petrochemicals and rubber industries, analgesic abuse, high arsenic content in drinking water, exposure to cyclophosphamide and the presence of chronic inflammation. Chronic infl ammatory conditions are also associated with squamous cell carcinoma.

Presentation

Patients commonly present with gross hematuria, with or without flank pain and occasionally clot colic. Passage of long, slender, worm-like clots is suggestive of upper tract involvement. Patients with known bladder tumors should always be screened for upper tract tumors. Very few present with advanced constitutional symptoms and a palpable mass. Microscopic hematuria should be evaluated to exclude urothelial malignancy in the high-risk adult (chronic smokers, occupational exposure, older age) population.

Pathology

Both the PCS and the ureter have a thinner muscular layer than the bladder. Therefore, aggressive tumors of the upper tract can easily invade the muscle layers; hence, the prognosis is poor. Urothelial tumors can invade surrounding tissues, metastasize to regional lymph nodes and spread haematogenously to lungs, liver and bones.

Diagnosis

Urinalysis may reveal numerous RBCs and white blood cells. Urine cytology should be obtained. The presence of atypical or malignant cells in a freshly voided sample has a high specificity for urothelial malignancies. CTU, cystoscopy, retrograde pyelogram and flexible ureterorenoscopy are required for diagnosis. CTU is the investigation of choice.

Benign renal tumors

Incidental detection of renal lesions has increased owing to the widespread use of abdominal imaging. The lesions may be cystic or solid. Solid renal tumors should be considered malignant unless proven otherwise.

Renal oncocytoma

This derives its name from its cellular appearance on histo pathology, where uniformly highly granular eosinophilic cytoplasm owing to abundant mitochondria (oncocyte) is seen. It accounts for around 5% of renal tumors. It appears as an enhancing mass on cross-sectional imaging and is difficult to differentiate from RCC.

Renal angiomyolipoma

Angiomyolipoma comprises a composite mix of fat tissue with dysmorphic blood vessels and smooth muscle. It is most often detected incidentally and has a female preponderance. Angiomyolipoma may be associated with syndromes such as the tuberous sclerosis complex or it may be sporadic in nature.

Renal cell carcinoma

RCC is the most common solid neoplasm of the kidney. It accounts for around 90% of renal tumors and constitutes 2–5% of all cancers in adult men and 1–3% in adult women. There has been a recent steady increase in the incidence of RCC. It may be sporadic or familial.

Familial renal cell carcinoma

von Hippel–Lindau syndrome is the most common familial syndrome associated with RCC. VHL disease is a rare autosomal dominant disorder that is characterized by multiple pathologies, including clear-cell RCC , phaeochromocytoma, retinal angiomas and haemangioblastomas of the brainstem, cerebellum or spinal cord.

Aetiology

Cigarette smoking, obesity and hypertension are the major risk factors associated with RCC. Others include diuretics, occupational exposure to petrochemicals and dyes and ARCD in patients on long-term hemodialysis.

Clinical presentation

The classic triad of flank pain, hematuria and a palpable mass is now uncommon as most renal masses are detected incidentally. Symptoms and signs may be non-specific. The most common presenting symptom is hematuria. Patients may have constitutional symptoms such as fever, malaise and weight loss in advanced disease. Advanced disease can present with bilateral lower limb oedema or recent-onset non-reducing right-sided varicocele owing to thrombus in the IVC. Paraneoplastic syndromes (PNSs) are found in up to one third of patients with RCC. The most common PNS is an elevated erythrocyte sedimentation rate (ESR) followed by hypertension, anemia and hypercalcemia. Up to a quarter of the patients may have evidence of metastatic disease on presentation. The most common site of metastasis is the lung and is classically described as cannon ball metastases. Metastases may occasionally present as pathological fractures.

Pathology

ccRCC is histologically an adenocarcinoma arising from the proximal renal tubular epithelium. They are slow growing and bulge out of the renal contour

The tumor can spread directly, invading the perinephric tissue through the capsule or at times directly extending into the renal vein as a tumor thrombus. Vein wall invasion is associated with poor prognosis

Diagnosis

blood count, ESR, serum creatinine, liver function tests, lactate dehydrogenase (LDH), corrected serum calcium, coagulation markers and urine analysis. Increased alkaline phosphatase should prompt further investigation to rule out liver and skeletal metastases. LDH is useful in risk stratification of metastatic disease.

Investigations

Ultrasound-CT scan- MRI

Treatment

Radical or nephron-sparing nephrectomy, immunotherapy, Tyrosine kinase inhibitors inhibit vascular endothelial growth factor

Wilms' tumour

This is the most common tumor of childhood, accounting for 5% of all childhood cancers. They are bilateral in 5% of cases and familial in 1%. The tumor has mixed elements derived from the embryonic nephrogenic tissue, namely blastemal or undifferentiated tissue, epithelial tubules and stroma. The typical presentation is a child aged between 1 and 4 years of either gender with a large,

palpable abdominal mass that may cross the midline. It may also be associated with hematuria, hypertension, fever and weight loss. Pain is relatively uncommon. The large tumor can rupture and present as an acute abdomen. Other causes of renal masses include neuroblastoma, congenital mesoblastic nephroma, RCC, clear-cell sarcoma and rhabdoid tumor. US can confirm the renal origin and solid nature of the mass. Further definitive imaging with either CECT or MRI is necessary to stage the disease. Up to 13% of patients have bilateral tumors. The tumors usually infiltrate the kidneys and normal renal parenchyma is compressed at the periphery around the tumor (claw sign). A CT of the chest should be obtained as the lung is the most common site of distant metastasis. Current treatment is nephrectomy with pre- or postoperative chemotherapy. Both regimes have a comparable survival of ~90%.