Answer to Medical Quiz: Image - 1

DOI: https://doi.org/10.3329/bjm.v33i1.56805

Answer 1. Ectopic right kidney with multiple air pockets within it.

Answer 2. Radiological diagnosis is class 2 emphysematous pyelonephritis involving the ectopic/pelvic right kidney.

Answer 3. Resuscitation, intravenous broad spectrum antibiotic,glycaemic control using insulin and other supportive measures.

Review:

Emphysematous pyelonephritis is a rare, severe form of necrotizing infection involving the renal parenchyma, collecting system and peri-nephric area characterized by accumulation of gas.[1] Patients with diabetes mellitus and those having obstructive uropathy by stone or tumoursconstitute the predominant risk group. Clinical presentation is indistinguishable to those of acute pyelonephritis and include fever, loin pain and vomiting.[2]Pneumaturia is an uncommon but characteristic presentation.[3] Imaging can identify gas and CT classification is proposed by Huang and Tseng: class 1 – gas in the collecting system only, class 2 – gas in the renal parenchyma, class 3A – gas/abscess in peri-nephric area, class 3B – gas/abscess in para-nephric area and class 4 – bilateral disease or emphysematous pyelonephritis in single kidney.[1] The index case falls in class 2, though the initial assessment mimicked an appendicular lump. Rare case of class 4 emphysematous pyelonephritis with emphysematous cystitis is reported.[4] Treatment includes resuscitation, intravenous antibiotics, control of diabetes and surgery in selected cases. Class 1, 2 and less aggressive (one risk factor from shock, altered sensorium, renal failure and thrombocytopaenia) forms of 3 and 4 may be managed conservatively (including percutaneous drainage), while those with aggressive disease (2 or morerisk factors) merits nephrectomy.[1] A recent report revealed high radiological class and acute kidney injury were associated with nephrectomy.[2] The mortality of emphysematous pyelonephritis has fallen from 80% to less than 20% over last few decades because of high index of clinical suspicion, availability of CT scan facility, use of effective, broad-spectrum antibiotics and teamwork between nephrologists, radiologists and urologists.

Declaration of interest:

The author reports no conflict of interest.

Funding:

The author received no financial support for the research, authorship, and/or publication of this article.

References:

1. Huang, JJ, Tseng, CC. Emphysematous pyelonephritis: clinicoradiological classification, management, prognosis, and pathogenesis. Arch Intern Med 2000; 160: 797-805. https://doi.org/10.1001/archinte.160.6.797. PMid:10737279
2. Rahim MA, Ananna MA, Iqbal S, Uddin KN, Latif ZA.Emphysematous pyelonephritis: experience at a tertiary care hospital in Bangladesh. J Royal Coll Physicians Edinb2021 Mar;51(1):19-23. https://doi.org/10.4997/JRCPE.2021.106. PMid:33877129
3. Kelly HA, MacCallum WG. Pneumaturia. J Am Med Ass 1898; 31: 375-81. https://doi.org/10.1001/jama.1898.92450080001001
4. Rahim MA, Jahan I, Chowdhury TA, Ananna MA, Iqbal S. Class 4 emphysematous pyelonephritis with emphysematous cystitis: report of a rare case from Bangladesh.Trop Doct 2021 Jul;51(3):452-4. https://doi.org/10.1177/0049475520983641. PMid:33413031.

Answer to Medical Quiz: Image - 2

DOI: https://doi.org/10.3329/bjm.v33i1.56805

Answers: Question-1

a) FLAIR images show several large areas of hyperintense signal in the white matter of the right cerebral hemisphereand in the splenium of the corpus callosum.

b) Small sites of cortical involvement are present.

c) There is no abnormal enhancement

Ans Question-2 :

Progressive Multifocal Leukoencephalopathy (PML)

Ans Question-3 :

a) HIV encephalitis
b) Acute disseminated encephalomyelitis (ADEM)

Ans Question-4:

Antiretroviral therapy

Review:

- PML is a demyelinating process caused by the JC polyomavirus. Th is virus infects or becomes active in patients with severe immunodefi ciency, such as AIDS, lymphoproliferative or myeloproliferative disorders, immunosuppressive therapy, or congenital immunodefi ciency. Recently cases of PML have occurred in patients with multiple sclerosis undergoing treatment with natalizumab.
- Classic clinical presentation includes focal neurologic deficits, most commonly limb weakness or ataxia. This presentation helps to distinguish PML from HIV encephalitis, which usually presents with global cognitive decline.
- The process predominantly affects white matter, but some involvement of gray matter structures is present in 50% of cases.
- The lesions present on MRI are usually asymmetric and nonenhancing.
- In the setting of recently initiated highly active retroviral therapy (HAART), marginal enhancement is more common. Such patients may experience a marked clinical deterioration, termed “immune reconstitution inflamatory syndrome” (IRIS).

Management:

- The diagnosis may be confirmed by the presence of JC viral antigen in the CSF using PCR.
- The only known effective treatment is reconstitution of the immune system with antiretroviral therapy.

Declaration of interest:

The authors report no conflict of interest.

Funding:

The author received no financial support for the research, authorship, and/or publication of this article.

References:

1. Berger JR; Aksamit AJ; Clifford DB; et al. PML diagnostic criteria: consensus statement from the AAN Neuroinfectious Disease Section”. Neurology.2013; 80(15): 1430-8. https://doi.org/10.1212/WNL.0b013e31828c2fa1
2. Buckle C, Castillo M. Use of diff usion-weighted imaging to evaluate the initial response for progressive multifocal leukoencephalopathy to highly active antiretroviral therapy: Early experience . AJNR Am J Neuroradiol. 2010; 31 : 1031 - 1035 . https://doi.org/10.3174/ajnr.A2024
3. Weber T. Progressive multifocal leukoencephalopathy . Neurol Clin. 2008 ; 26 : 833 - 854 . https://doi.org/10.1016/j.ncl.2008.03.007
4. Muftuoglu, Muharrem; Olson, Amanda; Marin, David; et al. (2018-10-11). “Allogeneic BK Virus-Specific T Cells for Progressive Multifocal Leukoencephalopathy”. New England Journal of Medicine.2018; 379 (15): 1443-1451. https://doi.org/10.1056/NEJMoa1801540