



Sarcoidosis of the Ureter: A Review of the Literature

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Abstract

Sarcoidosis of the ureter (SOU) is extremely rare. The aim of this article was to review the literature on SOU. Various internet data bases were searched for information on SOU. Four cases of sarcoidosis primarily involving the ureter and one case of retroperitoneal sarcoidosis with lymph node involvement encasing and obstructing the ureters bilaterally have been reported. SOUs have been reported in both males and females whose ages have ranged between 43 years and 73 years. SOU primarily involving the ureter tends to be associated with loin pain. Ultrasound scan would tend to show hydronephrosis of the affected side. Other radiological images including CT scan, MRI scan and intravenous urography would tend to show hydronephrosis as well as the site and extent of the lesion in the ureter as well as length and other features of any other lesion or lesions that are contiguous with the ureteric lesion but the findings are not specific. Raised levels of serum angiotensin converting enzyme as well as a history of sarcoidosis should alert a clinician to suspect SOU. Biopsies of ureteric lesions obtained at ureteroscopy, and excised ureteric lesions would show non-caseating granulomas which may be associated with inclusion bodies. Treatment of the four reported cases of SOU had included local excision of the involved segment of ureter and primary ureteric anastomosis to restore continuity of drainage down the upper urinary tract and ureteric stent insertion. A case of SOU was diagnosed after nephroureterectomy was undertaken following a provisional diagnosis of urothelial carcinoma of the ureter. Spontaneous resolution of retroperitoneal sarcoidosis obstructing two ureters without any treatment surgical treatment or use of steroids has been reported after laparoscopic biopsy of the lesion. There is a place for the use of steroid treatment in the case SOU that is associated with systemic sarcoidosis even though spontaneous resolution can occur. SOU is rare and its diagnosis may be delayed because of initial suspicion of other common differential diagnoses. Diagnosis of SOU may be established by histological examination of biopsy specimens of the ureteric lesion and exclusion of tuberculosis as well as fungal disease which should enable local excision of the lesion with or without treatment with steroids.

Key Words: Sarcoidosis of ureter; Sarcoid of ureter, Hydronephrosis; Serum Angiotensin converting enzyme, non caseous granulomas, Asteroid bodies, Schaumann bodies; Hamazaki Wesenberg bodies.

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Introduction

Sarcoidosis primarily affecting the ureter is extremely rare in view of this clinicians may be

unfamiliar with the manifestations and management of the disease.

Aims

To review the literature on sarcoidosis of the ureter.

Methods

Various internet data bases were searched including Google, Google Scholar, Educus, and PUB Med. The following key words were used: Sarcoidosis of ureter, Sarcoid of ureter, ureteric sarcoidosis. Information obtained from ten references was used in the write up of the review article which has been divided into (A) Overview and (B) Miscellaneous narrations and discussions from reported cases.

Reference year,	Age, sex, site	Presentation	Radiological findings	Treatment
Perimenis et al. [4] 1990	67-years, male, right	Right loin pain	Intravenous urography showed right hydronephrosis but did not show cause of right ureteric obstruction; right retrograde ureteropyelogram showed obstructing lesion in right upper ureter.	Exploration of right kidney and ureter and excision of 3 cm length of a hardened part of the right ureter with and re-anastomosing of the right ureter to the right renal pelvis. There was no obvious periuteteric lesion.
Mariano and Sussman [6] 1998	72-years, female, left	Left loin pain	Contrast CT-Scan showed left sided hydronephrosis associated with some irregular, soft-tissue density around the proximal ureter the left ureter; an intravenous urogram and retrograde left ureteropyelogram which confirmed a 3-cm narrowing of the proximal on third of the left ureter.	Insertion of left ureteric stent, exploration of left ureter, resection of the affected portion of the ureter and ureterolysis with omental wrapping of the ureter.
Hashimoto et al. [9] 2012	65 years; male, left	Presentation not available to author	Radiological investigation findings not available to author (Japanese literature)	Left nephroureterectomy and regional lymph node dissection under a clinical diagnosis of transitional cell carcinoma of the left ureter with lymph node metastasis.
Kalia et al. [10] 2010	43 years; male; left	Left loin pain	Ultrasound scan showed left sided hydroureteronephrosis and CT scan showed irregular soft tissue density in the upper ureter 12 mm distal to the left pelvi-ureteric junction as well as stranding in the periureteric fat. The CT scan also showed multiple lymph nodes in the retroperitoneum	He underwent cystoscopy and flexible left ureterorenoscopy and subsequently underwent laparoscopic excision of the ureteric lesion and left ureteropyelostomy and insertion of left ureteric stent which was subsequently removed.
Miyazaki et al. [8] 1996 (This case of sarcoidosis is not primary sarcoidosis affecting the ureter but a case of retroperitoneal sarcoidosis with lymph node involvement that had compressed the ureters)	53 years; female; bilateral hydronephrosis	Dysuria and urinary incontinence	She had intravenous urography which showed bilateral hydronephrosis; she also had computed tomography (CT) scan which showed a retroperitoneal mass surrounding enlarged retroperitoneal lymph nodes that had compressed both ureters and causing bilateral hydronephrosis. The CT scan also showed low-density areas both in kidneys which were not noted at the time the CT scan, was undertaken	She had laparotomy and biopsy of the retroperitoneal mass, and histological examination of the specimen showed noncaseating epithelioid cell granulomas A conservative treatment approach was adopted and subsequent CT scan did show resolution of the hydronephrosis and the lesions.

Table 1: A list of reported cases of sarcoidosis involving the ureter directly or indirectly from external compression

Results / Literature Review

(A) Overview

General definition

Sarcoidosis is a multi-system disease of unknown origin which tends to involve the lung or bilateral lymph nodes in 90% of cases [1] but can involve many other organs.

Pathophysiology

It had been suggested that sarcoidosis could be a manifestation of disordered immune regulations within individuals who are genetically susceptible to the disease pursuant to exposure to environmental agents. [1]

It has been stated that there is an increased risk of sarcoidosis with regard to some HLA genotypes [1]

) It has been iterated that there is an increased risk of sarcoidosis in cases of an increase in pulmonary T-cell derived CD4:CD8 ratio. [1]

) It had been intimated that Sarcoidosis tends to be associated with an increase in T-cell derived cytokines IL-2 and IFN-gamma. [1]

) Furthermore, it had been indicated that there is an increase of sarcoidosis in other cytokines (including IL-8, TN, and other cytokines) in the microenvironment. [1]

Clinical manifestations

Sarcoidosis generally tends to be associated with anergy to skin test antigens, polyclonal hypergammaglobulinemia [1] Diagnosis of sarcoidosis tends to be a diagnosis of exclusion in view of the fact that the clinical manifestations are non-specific and the pathological features tend to mimic other diseases and special stains and cultures from the specimen obtained from the organ involved tend to be required to help establish the diagnosis and a previous history of sarcoidosis involving another organ tends to be helpful in alerting the clinician to a possible diagnosis of sarcoidosis.

With regard to epidemiology, sarcoidosis tends to affect patients whose ages have ranged between 20 years and 40 years, and it tends to be more common in females than in males. The disease also tends to be more common in the black race in comparison with other races and it is rarely encountered in Chinese and in Southeast Asians. [1]

) The common types of sarcoidosis tend to present with peri-hilar node involvement by the disease, diffuse pulmonary disease, interstitial pulmonary fibrosis, localized bronchial stenosis, distal bronchiectasis and atelectasis. [1]

) Eighty percent of patients with sarcoidosis tend to have elevated serum angiotensin-converting enzyme which is not specific to sarcoidosis. Sixty-five percent of patients with sarcoidosis generally recover without any subsequent problems. Twenty percent of patients with pulmonary sarcoidosis have permanent pulmonary loss; 3% of patients with sarcoidosis tend to die as a result of pulmonary fibrosis or of congestive cardiac failure. [1]

Lungs

In cases of sarcoidosis of the lung, there may not be any evidence of a gross lesion or nodules that measure between 1 cm and 2cm may be encountered and these quite often tend to be located in bronchial submucosa and histological examination of biopsies of these nodules tend to be helpful in establishing the diagnosis of sarcoidosis. [1]

Lymph nodes

Sarcoidosis involvement of lymph nodes tends to be related to hilar or mediastinal lymph nodes in almost all of the cases of sarcoidosis and the tonsils tend to be involved in 25% of cases of sarcoidosis. [1] The lymph nodes may also be calcified. [1]

Liver/Spleen

It has been stated that sarcoidosis involving the liver / spleen may be in the form of gross macroscopic involvement in 20% of cases and microscopic involvement of these organs in 75% of cases. [1]

Bone

It had been iterated that in sarcoidosis radiological changes in bone tends to occur in 20% of cases and usually these changes are seen in small bones of the hands and feet. [1]

Skin

It has been stated that sarcoidosis involvement of the skin tends to occur in between 30% to 50% of cases with erythema nodosum and that the mucus membranes can also be involved. [1]

Eye

It has been documented that in sarcoidosis iritis or iridocyclitis tends to occur in between 20% and 50% of cases. [1]

Necrotizing sarcoid granulomatosis

It had been documented that necrotizing sarcoid granulomatosis is extremely rare and that controversies exist whether the disease entity is a distinct form of sarcoidosis. [1]

It had also been stated that necrotizing sarcoid granulomatosis commonly affects women who either tend to have no symptoms or if symptomatic their symptoms tend to be mild. Necrotizing sarcoid granulomatosis tends to be associated with excellent prognosis following steroid therapy, use of immunosuppressive medicaments and surgical excision for localized disease. [1]

It had been intimated that necrotizing sarcoid granulomatosis mimics tuberculosis, fungal infections, Wegener's disease, and that it is important in view of the differential diagnoses to exclude infections by means of negative culture and histochemical staining of the specimens on pathological examination of the specimens. [1]

Sarcoidosis of the ureter

Sarcoidosis of the ureter is very rare and majority of clinicians would perhaps not encounter the disease and they would also be likely unfamiliar

with the presentation of the disease. Nevertheless, sarcoidosis of the ureter may present with:

-) Loin pain
-) Impaired renal function on biochemical tests
-) Hydroureteronephrosis of the affected ureter and upper renal tract

) It may mimic tuberculosis of the ureter or a malignant lesion of the ureter and the diagnosis may be ascertained as an incidental finding of an excised lesion of the ureter upon histological examination of the specimen without provisionally suspecting the lesion unless there is a history of previous systemic sarcoidosis.

Prognosis of pulmonary/systemic sarcoidosis

With regard to prognosis the best prognosis tends to be associated hilar lymphadenopathy alone and the worst outcome associated with pulmonary disease which does not have adenopathy. [1]

With regard to the few cases of sarcoidosis of the ureter which have been reported the prognosis so far, have been good pursuant to excision of the ureter and or following nephroureterectomy.

Investigations

Hematology tests

Full blood count

Full blood count and coagulation test may be undertaken as part of the general assessment of a patient with sarcoidosis of the ureter but the results would not provide any specific information that would be diagnostic of sarcoidosis of the ureter.

Biochemistry tests

Serum urea and electrolytes and liver function tests are tests that are carried out as part of the general assessment of a patient with sarcoidosis of the ureter but the results would not be specific to diagnose sarcoidosis of the ureter. However, if the serum urea and electrolyte results show there is impairment of renal function this may lead the clinician to undertake radiological investigations that may show obstruction of an upper urinary tract and this would help the clinician to undertake further investigations which may lead to the diagnosis of sarcoidosis of ureter.

Serum angiotensin converting enzyme levels could be raised in patients with sarcoidosis of the ureter but this would not be diagnostic of the disease.

Microbiology tests

Bacteriology

Urinalysis, urine microscopy and culture are routine tests that are undertaken as part of the general

assessment of patients with sarcoidosis of ureter but these would not be diagnostic of sarcoidosis of ureter. If there is any evidence of urinary tract infection the infection would be treated prior to undertaking any endoscopic examination and surgical excision of the ureteric lesion.

Culture of urine to exclude Acid fast bacilli tends to be undertaken as part of the assessment of a patient when the diagnosis of sarcoidosis of the ureter is a possibility to exclude tuberculosis of the ureter.

Radiology

Ultrasound scan of renal tract, abdomen and pelvis may show evidence of hydroureteronephrosis and presence or absence of any mass lesion around the ureter or anywhere else in the abdomen and pelvis. If required percutaneous nephrostomy insertion and nephrostogram as well as insertion of ante-grade ureteric stent could be undertaken in cases of hydronephrosis due to sarcoidosis of the ureter initially to preserve renal function by means of the nephrostomy / ante-grade ureteric stenting and nephrostogram as part of investigation to establish the cause of the hydronephrosis.

CT scan

Contrast computed tomography urogram may be undertaken as an investigation to establish the cause of loin pain the patient presents with and this may show hydronephrosis and any abnormal looking areas as well as the extent of the abnormality of the ureter which would enable the urologist undertake further investigations including cystoscopy, retrograde ureteropyelogram, ureteroscopy and biopsy of any ureteric lesion found for histological examination. The CT urogram would also provide information whether or not there is any other lesion in the abdomen or pelvis.

MRI scan

Magnetic resonance imaging (MRI) scan of the renal tract, abdomen and pelvis may be undertaken as an investigation to establish the cause of loin pain the patient presents with and this may show hydronephrosis and any abnormal looking areas as well as the extent of the abnormality of the ureter which would enable the urologist undertake further investigations including cystoscopy, retrograde ureteropyelogram, ureteroscopy and biopsy of any ureteric lesion found for histological examination. The MRI scan would also provide information whether or not there is any other lesion in the abdomen or pelvis.

Radiological screening for retrograde ureteropyelogram, ureteroscopy and biopsy of ureteric lesion and insertion of retrograde ureteric stent.

As part of investigation of ureteric obstruction cystoscopy, retrograde ureteropyelogram, ureteric urine collection for microscopy and culture, ureterorenoscopy and biopsy of any ureteric lesion may be undertaken as part of the investigation of a patient with sarcoidosis of the ureter. These investigations are undertaken under radiological imaging screening. If insertion of ureteric stent is required by the retrograde approach radiological screening is required to confirm correct positioning of the stent.

Isotope Renogram

Isotope renogram may be required in cases of sarcoidosis of the ureter in order to assess for differential renal function and to confirm obstruction of the ureter. In the situation when the differential function of the affected ureter is very low then a nephroureterectomy would be carried out but if the percentage function of the affected upper renal tract is good enough then local excision of the affected ureter could be undertaken.

Microscopic examination findings of sarcoidosis

General description

Considering the fact that sarcoidosis of the ureter is rare, knowledge of the microscopic features of sarcoidosis in general especially would enable the pathologist and clinician to suspect and establish a diagnosis of sarcoidosis of the ureter.

Generally microscopic examination of specimens of sarcoidosis tend to show non-caseating granulomas that have tightly packed epithelioid cells, Langhans giant cells and lymphocytes (T cells) which are usually located within the interstitium adjoining bronchioles and surrounding and within vessel walls, pleura and connective tissue septa in cases of pulmonary sarcoidosis. [1]

It has been stated that in sarcoidosis there could also be hyalinization, diffuse interstitial fibrosis, fibrinoid - necrosis, fibrosis inside granulomas, intra- and extracellular inclusions. [1] It had also been stated that in pulmonary sarcoidosis there may be pleural involvement in 10% of cases [1]

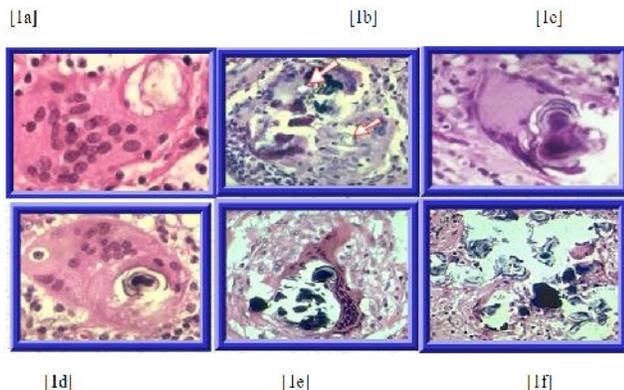
Inclusion bodies that could be seen on microscopic examination of sarcoidosis include Schaumann bodies which are laminated concretions of calcium and protein and asteroid bodies which are

stellate inclusions that are found in giant cells in 60% of granulomas. [1] Furthermore, it has been documented that neither Schaumann bodies nor asteroid bodies is specific to establish a diagnosis of sarcoidosis because they can also be found in differential diagnosis like berylliosis. [1]

It had been iterated that in necrotizing sarcoid granulomatosis microscopic examination of the specimen tends to show extensive, vascular, non-caseating sarcoid-like granulomas that invade pulmonary arteries and veins with diffuse necrosis of the parenchyma of the lung. [1]

Rosen [2] stated that the role of pathology in the establishment of a diagnosis of sarcoidosis is the identification of granulomas in tissue specimens and the exclusion of known causes of granulomatous inflammation and that the granulomas of sarcoidosis are non-specific lesions which by themselves and in the absence of identifiable etiological agent, would not be diagnostic of sarcoidosis or any other specific disease. Rosen [2] further stated that strict clinical, radiological, and pathological criteria must be used to establish a diagnosis of sarcoidosis. Rosen [2] also iterated that a diagnosis of sarcoidosis is most secure in the presence of compatible clinical and radiological findings collaborated by the demonstration of micro-organism negative, non-necrotizing granulomas in a biopsy specimen that is accompanied by biopsy evidence or strong clinical evidence of multi-system involvement, and negative cultures for bacteria, mycobacteria, and fungi.

Specifically in sarcoidosis of the ureter, microscopic examination of the excised ureter would tend to reveal non caseating granulomas involving the ureter and there may be associated inclusion bodies like Schaumann bodies, Asteroid bodies and or Hamazaki-Wesenberg bodies [see figures 1a, 1b, 1c, 1d, 1e, 1f, 2a, 2b, 3a, 3b, 3c, 3e, and 3f which show examples of various inclusion bodies related sarcoidosis and non-sarcoidosis lesions Reproduced from [3]: Rosen Y. Inclusions in granulomas Schaumann (conchoidal) bodies Atlas of Granulomatous diseases URL: granuloma.homestead.com/inclusions.html under copy right which states that the images may be copied and utilized for education or other non-commercial purposes. reference [3]



Figures 1a to 1f: [1a] Early Schaumann body forming around calcium oxalate crystals; [1b] Fragmented Schaumann body surrounding calcium oxalate crystals (at arrows) polarized; [1c]. Schaumann body; [1d] Schaumann body; [1e] Large fragmented, Schaumann body; [1f] Extracellular Schaumann body. Reproduced from: Rosen Y (Yale) inclusions in granulomas Schaumann (conchoidal) bodies Atlas of Granulomatous diseases URL: granuloma.homestead.com/inclusions.html under copy right which states that the images may be copied and utilized for education or other non-commercial purposes.

Kveim-Siltzbach test

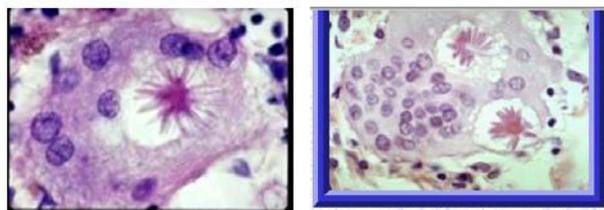
Rosen [2] stated that a positive Kveim-Siltzbach test would provide strong support in the establishment of a diagnosis of sarcoidosis.

Differential diagnoses

Some of the differential diagnoses to be considered with regard to systemic sarcoidosis include: atypical mycobacteria, Berylliosis for which a clinical history is required to establish the differentiation, extrinsic allergic alveolitis which tends to have loosely arranged epithelioid cells in granulomas, fungi, inhalation of talc and intravenous narcotism. [1]

With regard to sarcoidosis of the ureter some of the differential diagnoses include: tuberculosis of the ureter, all types of malignancies of the ureter, fungal infections.

Figure 2a: Asteroid body, sarcoidosis. Figure 2b: Asteroid bodies, foreign body granulomas



Figures 2a and 2b: Asteroid bodies: Stellate inclusions with numerous rays radiating from a central core. May be seen in granulomas of various entities but are most frequently encountered in the giant cells of foreign body granulomas. Structures strongly resembling asteroid bodies may be seen rarely in the cytoplasm of tumour giant cells

and in fibrin-rich exudates. Reproduced from: Rosen Y (Yale) inclusions in granulomas Schaumann (conchoidal) bodies Atlas of Granulomatous diseases URL: granuloma.homestead.com/inclusions.html under copy right which states that the images may be copied and utilized for education or other non-commercial purposes.

[B] Miscellaneous narrations and discussions from reported cases

Perimenis et al. [4] in 1990 reported a 67-year-old man who presented with a history of right loin pain and who was found to be tender in the right lumbar region. The results of his investigations including urine microscopy and culture, 3 early morning urine specimens for acid fast bacilli, and urine cytology, serum biochemistry test, and chest X-ray were normal. He had excretory urography which showed obstruction of the upper part of the right ureter and hydroureter without any cause identified but the left upper renal tract was normal. He had right retrograde ureteropyelogram which showed an obstructing lesion in the right upper ureter. He subsequently underwent exploration of right kidney and ureter and excision of 3 cm length of a hardened part of the right ureter and re-anastomosing of the right ureter to the right renal pelvis. There was no obvious peritubercular lesion. Histological examination of the excised ureter showed fibrous tissue that contained many noncaseating epithelioid granulomas which comprised of histiocytes and some Langhans giant cells. The giant cells were found to contain conchoidal bodies. Stains for fungi and acid fast bacilli were found on examination to be negative. He had a test for tuberculosis which was normal and calcium metabolism test was normal. He had Kveim test which was positive. Perimenis et al. [4] stated the following:

) The diagnosis of sarcoidosis in their case was unexpected and it had been based upon exclusion of other diseases, the pathology results and the positive Kveim test.

) The positive result of the Kveim test strongly indicated sarcoidosis but it could be regarded as definitive, even though false positive results are rare as intimated by Poole et al. [5]

) Fungal infections and tuberculosis had been excluded by means of negative stains and tuberculin test.

) One-organ localization of sarcoidosis is rare and one-organ localization of sarcoidosis raises diagnostic problems.

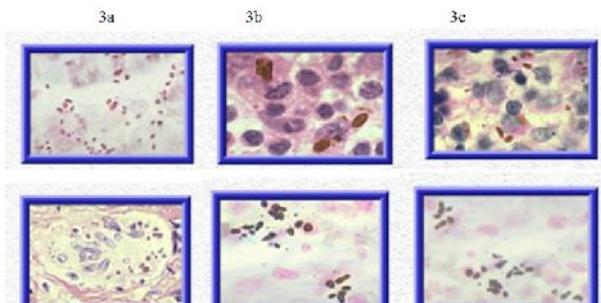


Figure 3a Hamazaki-Wesenberg bodies – lymph node – 40 X magnification; **3b** Hamazaki-Wesenberg bodies – lymph node – oil immersion; **3c** Hamazaki-Wesenberg bodies – lymph node oil immersion, **3d** Hamazaki-Wesenberg bodies in giant cell – lung, **3e** Hamazaki-Wesenberg bodies – lymph node methanamine silver – oil immersion. Note resemblance to yeasts, **3f** Hamazaki-Wesenberg bodies – lymph node – methanamine silver – oil immersion. Note resemblance to budding yeast. Reproduced from [3] Rosen Y. Inclusions in granulomas Schaumann (conchoidal) bodies Atlas of Granulomatous diseases URL: granuloma.homestead.com/inclusions.html under copy right which states that the images may be copied and utilized for education or other non-commercial purposes.

Mariano and Sussman [6] reported a 72-year-old diabetic woman who presented with a 3-week history of left loin pain but no nausea, vomiting or a history of nephrolithiasis. She had previously undergone cholecystectomy. Her urinalysis showed non visible haematuria and her urine cytology was normal. She had cystoscopy which was normal. She had contrast CT scan of her abdomen and pelvis which showed left sided hydronephrosis associated with some irregular, soft-tissue density around the proximal left ureter. She subsequently had an intravenous urogram and retrograde left ureteropyelogram which confirmed a 3-cm narrowing of the proximal one third of the left ureter. She had CT-guided needle aspiration from the site of the lesion; however, pathological examination of the specimen was non diagnostic. Gram staining and acid fast bacillus stain and cultures were negative. She had insertion of left ureteric stent, exploration of left ureter, resection of the affected portion of the ureter and ureterolysis with omental wrapping of the ureter. Pathological examination of the specimen showed noncaseating granulomas, which was consistent with sarcoidosis of the ureter. Mariano and Sussman [6] stated that their reported case was the second case of sarcoidosis of the ureter to be reported in the literature. Mariano and Sussman [6] stated that genitourinary involvement of sarcoidosis is not

common and this has been reported to occur in less than 5% of cases. [7]

Miyazaki et al. [8] reported a 53-year-old Japanese woman who presented with dysuria and urinary incontinence. She had urinalysis and cystoscopy which were normal. Her serum creatinine level was 1.5 mg /dl and her blood urea nitrogen was 30.6 mg / dl. She had intravenous urography which showed bilateral hydronephrosis and she also had computed tomography (CT) scan which showed a retroperitoneal mass surrounding enlarged retroperitoneal lymph nodes that had compressed both ureters and causing bilateral hydronephrosis. The CT scan also showed low-density areas in both of her kidneys which were not noted at the time the CT scan, was undertaken. She had laparotomy and biopsy of the retroperitoneal mass, and histological examination of the specimen showed noncaseating epithelioid cell granulomas. Examination of special stains and cultures did exclude tuberculosis, fungal infection and bacterial infection. She was referred to a different hospital 4 months later for further investigations where she had investigations including haematology and serum biochemistry tests most of which were within normal range. However, her serum angiotensin converting enzyme (SACE) and lysozyme levels were elevated at 27.9 IU/L and 33.0 g/dl respectively. Her serum calcium and calcitonin levels were within normal range but her urinary β_2 -microglobulin level was elevated at 3,156 microgram per litre. She had chest X-ray, lung function tests which were normal and trans-bronchial lung biopsies and histological examination of the specimen did not reveal any granuloma. She had CT scan of the abdomen 5 months pursuant to the biopsy which showed that the bilateral hydronephrosis had resolved spontaneously without any treatment and the retroperitoneal mass had reduced in size as well as that the bilateral low-density lesions in the kidneys had persisted. She had magnetic resonance scan which confirmed presence of multiple low-intensity areas in both kidneys and she also had Gallium scintigraphy which showed abnormal accumulation in both of the kidneys and in the retroperitoneal mass. She had renal biopsy and histological examination of the specimen did show noncaseating epithelioid cell granulomas and extensive interstitial fibrosis. Based upon a diagnosis of sarcoidosis with interstitial nephritis and retroperitoneal mass she was started on prednisolone therapy 60 mg orally daily and 3 months subsequently her renal function remained impaired but a new CT scan she had at that time did show complete resolution of the retroperitoneal lymphadenopathy. Subsequently at a time when she

was on 5mg per day treatment dose of prednisolone she had another CT scan which showed complete absence of retroperitoneal lymph node enlargement.

Hashimoto et al. [9] reported a 65-year-old Japanese man who underwent left nephroureterectomy and regional lymph node dissection under a clinical diagnosis of transitional cell carcinoma of the left ureter with lymph node metastasis. Microscopic examination of the specimen showed non caseous epithelioid granuloma with large Langhans cells in the ureteric lesion and in the dissected lymph nodes which was adjudged to be diagnostic of sarcoidosis. He had computed tomography (CT) of thorax which did not show any pulmonary or intrathoracic lesion attributable to sarcoidosis. Hashimoto et al. [9] stated that even though sarcoidosis of the genitourinary tract is rare, it should be considered in the differential diagnosis of urological conditions.

Kalia et al. [10] reported a 43-year-old man who presented with left loin pain. He was asymptomatic otherwise. His general and systematic examinations were normal. The results of his serum biochemistry and haematological investigations were normal except for raised serum uric acid level. There was no growth in his urine culture but pathological examination of the urine revealed 10 to 15 red blood cells per high power field and 1 to 3 pus cells per high power field. He had ultrasound scan of abdomen and pelvis as well as contrast enhanced computed tomography (CT) scan of abdomen and pelvis which showed left sided hydro-uretero-nephrosis. The CT scan showed irregular soft tissue density in the upper ureter 12 mm distal to the left pelvi-ureteric junction as well as stranding in the periureteric fat. The CT scan also showed multiple lymph nodes in the retroperitoneum. He underwent cystoscopy and flexible left ureterorenoscopy and subsequently underwent laparoscopic excision of the ureteric lesion and left ureteropyelostomy and insertion of left ureteric stent which was subsequently removed. Histological examination of the ureteric lesion, showed: granulomatous inflammation with noncaseating granulomas, a few Schaumann bodies, and extensive fibrosis which was adjudged to be diagnostic of sarcoidosis of the ureter. He then had a CT scan of thorax which was normal and which excluded presence of any pulmonary sarcoidosis. He had a follow-up CT scan of abdomen pelvis and thorax which showed residual left sided hydronephrosis and few nodules in the chest. He was put on steroid treatment and had been under follow-up with regular ultrasound scanning of the abdomen and renal tract. Kalia et al. [10] stated that:

) Hydronephrosis in sarcoidosis could be either due to retroperitoneal lymphadenopathy or primary ureteric involvement by sarcoidosis

) Even though retroperitoneal lymph node enlargement is common in sarcoidosis, hydronephrosis due to lymphadenopathy is rare and primary involvement of ureter by sarcoidosis is rarer.

) Radiological imaging plays a pivotal role with regard to diagnosis of sarcoidosis of ureter and in such situations the imaging does show the level of obstruction and would illustrate the cause of obstruction when it is not apparent but diagnosis of the disease can only be established by pathological examination of the lesion showing noncaseating granulomas.

) The differential diagnosis of sarcoidosis of the ureter include: (a) infections such as tuberculosis, actinomycosis, syphilis, histoplasmosis, (b) primary ureteric malignancy, (c) retroperitoneal fibrosis, (d) metastatic disease

Conclusions

Primary sarcoidosis of the ureter is a rare disease which could affect one ureter or both ureters and it may present with loin pain and radiological evidence of hydronephrosis. Primary sarcoidosis of ureter tends to mimic other causes of ureteric obstruction and its diagnosis is usually achieved as a diagnosis of exclusion based upon clinical history, radiological imaging findings, histological examination findings, of the excised or biopsied ureteric lesion and special cultures and stains to exclude other granulomatous lesions and fungal infection as well as malignant tumours of the ureter.

Conflict of interest: None

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