

## RETROPERITONEAL FIBROSIS IN YOUNG FEMALE

Pokhraj P Suthar<sup>1</sup>, Shreyans Mayur Doshi<sup>2</sup>

<sup>1</sup> Department of Radiology, SSG Hospital, Medical College, Vadodara, Gujarat, India

<sup>2</sup> Government Medical College, Surat, Gujarat, India

Correspondence to: Pokhraj P Suthar (pokhraj\_suthar@yahoo.co.in)

DOI: 10.5455/ijmsph.2014.140320144

Received Date: 03.03.2014

Accepted Date: 14.04.2014

### ABSTRACT

A 26 years old female presented with dull aching flank pain with elevated ESR, C - reactive protein and positive anti-nuclear antibody (ANA). Hypo echoic thin strip was noted in bilateral perirenal region without hydronephrosis on ultrasonography. Delayed enhancing perirenal rim of hypo dense soft tissue in bilaterally perinephric space on CT abdomen. Histological diagnosis of retroperitoneal fibrosis was confirmed.

**Key Words:** Retroperitoneal Fibrosis; Ultrasonography; CT Scan

### Introduction

Retroperitoneal fibrosis is characterized by the development of extensive fibrosis throughout the retroperitoneum. The estimated annual incidence varies from 1 per 200,000-500,000 population.<sup>[1,2]</sup> Association of retroperitoneal fibrosis with other connective-tissue diseases and reported familial occurrences suggest genetic factors may also play a role.<sup>[3]</sup> The symptoms and signs associated with retroperitoneal fibrosis are nonspecific, and diagnosis requires a high degree of suspicion. Although a definitive diagnosis can only be made based on biopsy findings, intravenous urography may provide further support for the diagnosis of retroperitoneal fibrosis, particularly if the classic features are present. CT scanning or MRI is essential for evaluating the extent of the disease process.

### Case Report

26 year old female presented with dull non colicky pain in flank, back and lower abdomen. Also had nonspecific symptoms like nausea, vomiting and malaise. On examination soft, non-tender abdomen without organomegaly. On routine investigation Haemoglobin 9.1 gm% with normocytic normochromic anaemia, total WBC count 8200 cell/cu.mm, ESR was 65 mm (elevated) in one hour, raised C- Reactive protein level and positive antinuclear antibody titre (ANA). X - Ray Chest, X - Ray Abdomen (standing) and X - Ray KUB were normal.

On ultrasonography study, hypoechoic thin strip noted in bilateral perirenal region without hydronephrosis. On CT scan Abdomen, perirenal rim of hypodense soft tissue was seen bilaterally within perinephric space showing delayed enhancement. It measures maximum 10 mm on right side and 8 mm on left side. Extensive fat stranding seen in the

bilateral perirenal and pararenal spaces with thickening of bilateral Gerota's, lateral conal and Zukercandl's fascia. Minimal fat stranding seen in pre and para aortic region and bilateral upper ureter. However both kidneys appears normal in size in density without calculus/hydronephrosis. Both ureters were well opacified with contrast. Multiple subcentimeter size aortocaval, paraaortic and bilateral common iliac group lymph nodes were noted. As an incidental finding cholelithiasis, bilateral polycystic ovary and bilateral pleural effusion were noted.



**Figure-1: Axial non contrast CT abdomen:** Perirenal rim of hypodense soft tissue bilaterally. It measures maximum 10 mm on right side and 8 mm on left side. Both kidneys appear normal in size without calculus

Histopathological study showed an inflammatory infiltrate contains few macrophages, lymphocytes, plasma cells with fibrosis. The macrophages were lipid-laden and had areas of perivascular lymphocytic infiltrate composed of T cells and B cells. Then patient was referred to Urologist for further management.



**Figure-2: Axial arterial phase of CT abdomen:** Non enhancing perirenal rim of hypodense soft tissue bilaterally within perinephric space. Extensive fat stranding seen in the bilateral perirenal & pararenal spaces with thickening of bilateral Gerota's, lateral conal & Zukercandl's fascia. However both kidneys appears normal in size in density



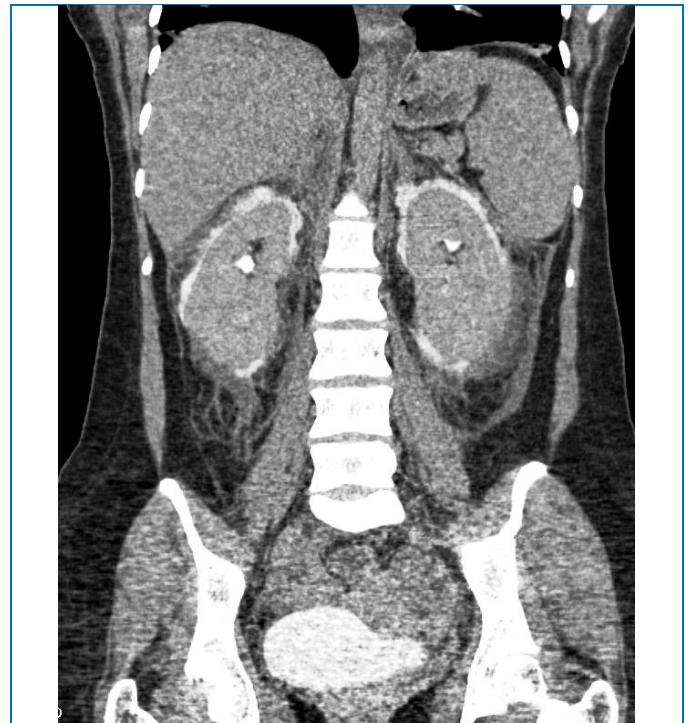
**Figure-3: Axial delayed phase of CT abdomen:** Delayed enhancing perirenal rim of hypodense soft tissue bilaterally in perinephric space. Both kidneys appears normal in size in density without hydronephrosis



**Figure-4: Axial delayed phase of CT abdomen:** Delayed enhancing perirenal rim of hypodense soft tissue bilaterally in perinephric space



**Figure-5: Axial delayed phase of CT abdomen:** Delayed enhancing perirenal rim of hypodense soft tissue bilaterally within perinephric space



**Figure-6: Coronal delayed phase of CT abdomen:** Delayed enhancing bilateral perirenal rim of hypodense soft tissue

### Discussion

Retroperitoneal fibrosis is characterized by the development of extensive fibrosis throughout the retroperitoneum. This fibrosis leads to entrapment and obstruction of retroperitoneal structures, notably the ureters. In most cases, the etiology is unknown. However, its occasional association with autoimmune diseases & its response to corticosteroids and immunosuppressive therapy suggest it is probably immunologically mediated.<sup>[4]</sup>

Approximately 8% of cases are associated with metastatic malignancy.<sup>[5]</sup> Retroperitoneal fibrosis is a relatively uncommon disease. The estimated annual incidence varies from 1 per 200,000-500,000 population.<sup>[4,2]</sup> It is twice common in male than female without racial predilection. The peak incidence of retroperitoneal fibrosis is in adults aged 40-60 years.<sup>[6]</sup> Association of retroperitoneal fibrosis with other connective-tissue diseases and reported familial occurrences suggest genetic factors may also play a role.<sup>[3]</sup> The human leukocyte antigen (HLA)-B27 cell marker has been demonstrated in several patients with retroperitoneal fibrosis.<sup>[7]</sup> Although the exact pathogenesis of retroperitoneal fibrosis has not been known, good evidence supports the suggestion that it develops as an immunologic response to antigens within atherosclerotic plaques.

The symptoms and signs associated with retroperitoneal fibrosis are nonspecific, and diagnosis requires a high degree of suspicion. Although a definitive diagnosis can only be made based on biopsy findings, intravenous urography may provide further support for the diagnosis of retroperitoneal fibrosis, particularly if the classic features are present. CT scanning or MRI is essential for evaluating the extent of the disease process.

On a sonogram, retroperitoneal fibrosis appears as a retroperitoneal, extensive, well-defined, hypo echoic mass centred over the sacral promontory. Doppler ultrasonography has no role in differentiating benign from malignant retroperitoneal fibrosis.<sup>[8]</sup>

On IVP, the classic triad includes delay of contrast material with unilateral (20%) or bilateral (68%) hydronephrosis, medial deviation of the middle third of the ureters, as depicted in the image below, and tapering of the ureters at the level of L4/L5 vertebrae.<sup>[9,10]</sup> Up to 18%-20% of control subjects may show this triad.<sup>[11]</sup>

On unenhanced CT scans, retroperitoneal fibrosis appears as a plaque that is isodense with muscle and that envelops the aorta and inferior vena cava between the renal hila and sacral promontory and usually extends laterally to incorporate the ureters. Obliteration of the fat plane between the mass and the psoas muscle may be observed. After contrast injection, the plaque may show a variable degree of enhancement, depending on the stage of the disease. Enhancement is usually significant in the early active vascular stage. On the other hand, enhancement is

poor in the late vascular stage.

Retroperitoneal haemorrhage, primary retroperitoneal sarcoma, metastatic deposits to the retro peritoneum and retroperitoneal amyloidosis may show similar findings on CT scans. CT scan features that suggest malignant pathology include lateral displacement of the ureter, anterior displacement of the aorta, local bone destruction, and a large bulky lesion.

Idiopathic retroperitoneal fibrosis carries a good prognosis, with little effect on long-term morbidity or mortality.

## Conclusion

CT scanning or MRI is essential for evaluating the extent of the disease process of Retroperitoneal Fibrosis.

## ACKNOWLEDGEMENTS

The authors would like to acknowledge the help of Department of Radiology, Shree Sayaji General Hospital, Vadodara.

## References

- Higgins PM, Bennett-Jones DN, Naish PF, Aber GM. Non-operative management of retroperitoneal fibrosis. *Br J Surg* 1988;75(6):573-7.
- Kottra JJ, Dunnick NR. Retroperitoneal fibrosis. *Radiol Clin North Am* 1996;34(6):1259-75.
- Hatsiopoulou O, Irving S, Sharma SD. Retroperitoneal fibrosis in 2 brothers. *J Urol* 2001;165(1):182.
- Katz SM, Bates O, Yudis M, Falkner B, Griffith E. Immune complex glomerulonephritis in a case of retroperitoneal fibrosis. *Am J Clin Pathol* 1977;67(5):436-9.
- Lepor H, Walsh PC. Idiopathic retroperitoneal fibrosis. *J Urol* 1979;122(1):1-6.
- McDougal WS, MacDonell RC. Treatment of idiopathic retroperitoneal fibrosis by immunosuppression. *J Urol* 1991;145(1):112-4.
- Astudillo L, Alric L, Jamard B, Laroche M. Retroperitoneal fibrosis in an HLA-B27-positive patient. *Rev Med Interne* 1999;20(12):1149-50.
- Rubenstein WA, Gray G, Auh YH, Honig CL, Thorbjarnarson B, Williams JJ, et al. CT of fibrous tissues and tumors with sonographic correlation. *AJR Am J Roentgenol* 1986;147(5):1067-74.
- Koep L, Zuidema GD. The clinical significance of retroperitoneal fibrosis. *Surgery* 1977;81(3):250-7.
- Webb AJ, Dawson-Edwards P. Non-malignant retroperitoneal fibrosis. *Br J Surg* 1967;54(6):508-18.
- Saldino RM, Palubinskas AJ. Medial placement of the ureter: a normal variant which may simulate retroperitoneal fibrosis. *J Urol* 1972;107(4):582-5.

**Cite this article as:** Suthar PP, Doshi SM. Retroperitoneal fibrosis in young female. *Int J Med Sci Public Health* 2014;3:632-634.

**Source of Support:** Nil

**Conflict of interest:** None declared