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# NEPHROCALCINOSIS / NEPHROLITHIASIS AS PRESENTATION OF MEN1 SYNDROME.

**Radiodiagnosis**

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## ABSTRACT

**Background-** Multiple endocrine neoplasia (MEN) 1 is characterized by hyperplasia /adenoma of parathyroid glands, pancreatic islet tumors , and pituitary gland tumors . We report a case of 44 year male who presented to surgical emergency with renal colic and NCCT KUB had shown nephrocalcinosis in B/L kidneys with left nephrolithiasis. We evaluated the patient by doing sonography and MRI which had shown almost all the feature of MEN 1. MEN 1 diagnosis and treatment requires multiple departmental and disciplinary approach. So it is very important for radiologist to know about all the causes and spectrum of appearance in MEN 1 .

## KEYWORDS

MEN 1, Nephrocalcinosis / Nephrolithiasis

## INTRODUCTION -

MEN 1, also known as Wermer syndrome, is an autosomal dominant condition with genetic defect located in chromosome 8 mostly presenting as the parathyroid, pancreatic, and pituitary tumors. Adrenal cortical tumors, carcinoid tumors, lipomatous tumors may also occur but are rare. Rarely can present as nephrocalcinosis and nephrolithiasis as there was in our case. In this article we found that nephrocalcinosis and nephrolithiasis are rare but important presentation in MEN 1 syndrome.

**Case Report –** 45 year male visited the surgical emergency with renal colic left side. NCCT KUB had shown B/L nephrocalcinosis with left nephrolithiasis .Patient had no other symptoms but clinical picture was showing raised calcium/ PTH level. This lead us to rule out possibility of MEN syndrome.

On evaluation sonography had shown B/L nephrocalcinosis / nephrolithiasis (figure 1a), pancreatic adenoma (figure 1b) and parathyroid adenoma (figure 2). MRI brain had shown lipoma scalp (figure 3a,b) & pituitary adenoma (figure 3c)

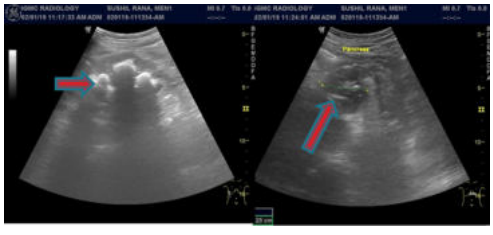

**Figure 1-a) Showing renal calculus with nephrocalcinosis . b) Showing pancreatic adenoma**

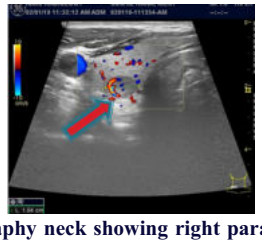

**Figure 2- sonography neck showing right parathyroid adenoma with arc and ring vascularity.**

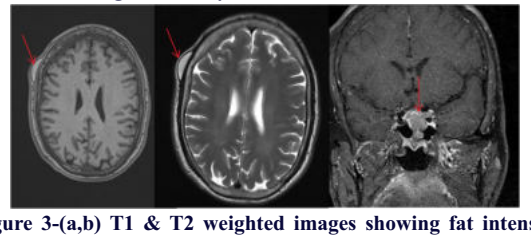

**Figure 3-(a,b) T1 & T2 weighted images showing fat intensity lesion in right frontal region (c) image showing non enhancing lesion in pituitary encasing the ICA on delayed T1 FAT-SAT coronal post contrast images**

## DISCUSSION-

MEN1 syndrome is most commonly inherited as an autosomal dominant condition. The diagnosis of MEN1 can be made by clinical criteria, on the basis of family history, or directly with genetic testing for a MEN1 mutation. Imaging plays a vital role for diagnosis of the clinical criteria's (parathyroid adenomas, enteropancreatic tumors, and pituitary adenomas.

**Andrew F et al 2006** had studied the spectrum of radiological appearances IN MEN syndrome and concluded that the understanding of the spectrum of disease and of the manifestations of each component is essential for accurate detection, staging, and surveillance in this diverse patient group.

**Mohamed N 2020** had reported a similar case who presented with renal calculi and was diagnosed as MEN 1.

Parathyroid adenomas are the most frequently occurring tumor in patients with MEN1, occurring in 90% of patients by age 40. Pancreatic neuroendocrine tumors (pNETs) are estimated to occur in 30%-80% of patients with MEN1. Pituitary tumors may occur in up to 60% of patients with MEN1. Furthermore, it is estimated that 1%-18% of patients diagnosed with primary hyperparathyroidism will have MEN1. The degree of hypercalcemia in these individuals is usually mild and patients may be asymptomatic There is a high prevalence of urolithiasis and early bone mineral loss in young individuals with MEN1-associated primary hyperparathyroidism.

## CONCLUSION –

Nephrocalcinosis / Nephrolithiasis is uncommon but can be a clue for diagnosis for MEN 1 syndrome in primary hyperthyroidism as in our case. Its very important for radiologist to know about the spectrum of MEN1 for the clinical management of patients.