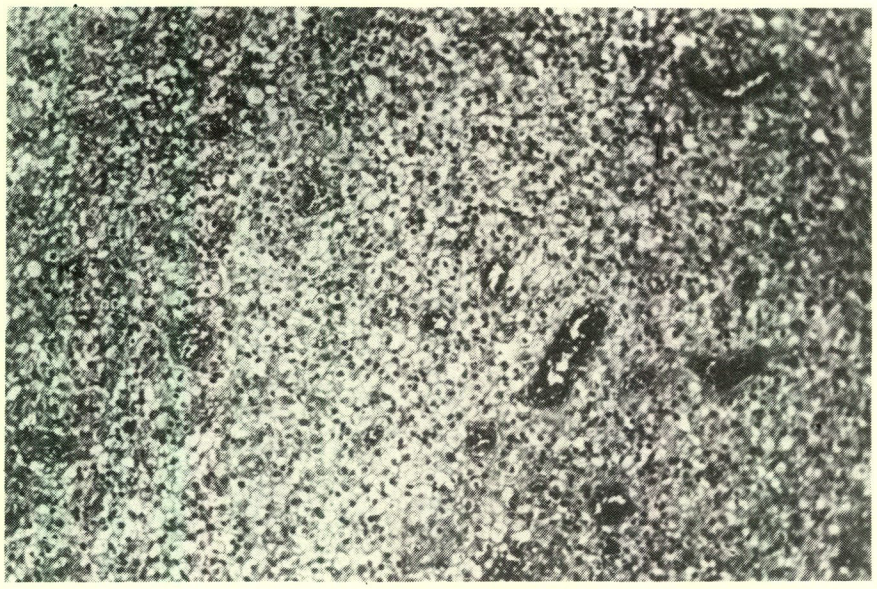

Fig. 4a :
Section showing replacement of renal tissue by sheets of foamy histiocytes, with vascular congestion.

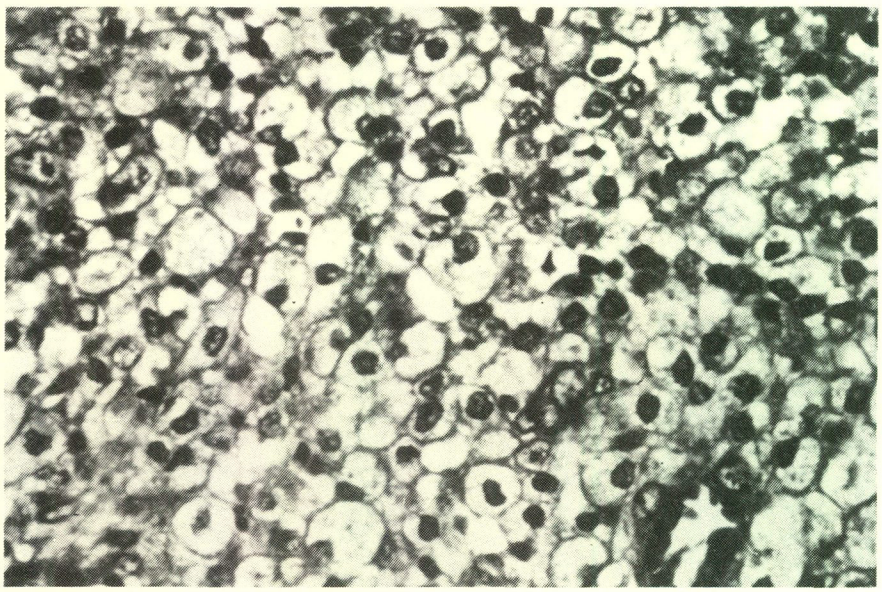

Fig. 4b :
Detail showing histiocytes with vacuolated cytoplasm containing lipid.

DISCUSSION

The recent increase in the reported cases of XGP is because of the advances in imaging technology, and an increased awareness in the medical community. The patients are of 5th and 6th decade of life; females to males ratio is 2 : 1[10] due to higher incidence of U.T.I. in females. Renal Cell carcinoma has occasionally been found to be associated with XGP[11,12,13].

The operative findings of a densely adherent kidney to the adjacent structures and the macroscopic and microscopic appearance of the kidney all strongly support the belief that the disease process had developed over years rather than months. It is surprising that there were so few urinary symptoms. The absence of these symptoms and the finding of a sterile urine can be misleading and results in errors of diagnosis and management.

Confident diagnosis can be made if sufficient characteristics are present pre-operatively[9]. The characteristics of the condition are :

1. It is always unilateral.
2. Absence or grossly impaired renal function on the involved side.
3. Large, frequently numerous renal Calculi are present.
4. Anaemia, raised ESR and leukocytosis are commonly present.

These criteria may not all be present in each case. Pathogenesis of the disease is still not clear.

An alteration in lipid metabolism with hypercholistrolaemia, was proposed[14] but the constant unilateral involvement and the infrequent finding of dyslipidaemia are against this theory.

Lymphatic stasis could be a co-factor[15], but is not the main mechanism of action. The presence of recurrent U.T.I. is a common finding in most series. The commonest germs were E. coli and proteous [16]. However, a number of cases fail to grow any organism.

A high incidence of renal lithiasis which may act through stasis, infection and irritation has also been documented[8,9]. Possible causes of sclerosing phlebitis[17] and ischaemic papillary nicrosis[18] have been proposed.

A more likely explanation about the pathogenesis is that there is a combination of different factors such as urine stasis and infection. Povisyl[19] was able to induce a typical XGP reaction in rats by ligating the ureter and intra parenchymal injection of E. coli culture.

Immunological factors[5] have also been suggested, as there is an increase in the levels of IgA, IgG & IgM, but the nature of the antigen is still unknown.

Clinically XGP presents a variable and a nonspecific picture, and is called "The great imitator". Fever in the majority of cases, lumber pain, malaise, weight loss are also common findings. Palpable mass may be present, while gross haematuria is rare. WBCs in the urine is 50% of cases, but microscopic haematuria is less frequent.

The role of urine cytology in the diagnosis of XGP has been stressed[20] where the characteristics of foam cells in the urine of patients with proven disease by examination of the morning voided urine or by ureteral catheter urine or urine from kidney puncture directly.

Non specific aspects of XGP are shown by the conventional radiological studies. Thus a KUB may show renal stones, enlarged kidney, obscured psoas shadow I.V.U. may show delayed or absent excretion[21,22].

Angiography usually shows avascular areas surrounded by basket-like arterial pattern[21,22], the main renal artery is normal or reduced in size.

Neovascularization may appear as an increased number of hilar or capsuler vessels. Arterial perfusion, and non homogeneous Nephrogenic effects may be present. However none of these features is typical of xanthogranulomatous pyelonephritis. Better diagnostic results by the advanced imaging technology of U.S., T.C. scan and M.R. imaging can be achieved.

Ultrasound reveals increased kidney size, renal stones as echo-reflecting central areas and hypoechogenic areas caused by suppurative foci[23,24,25], possibility of error with T.B. calcified kidney, and calculus pyelonephrosis.

C.T. scan reveals multiple low attenuation areas together with renal stones. The low attenuation areas are due to lipid rich XGP tissue and Calyces filled with pus, thickening of Gerota's fascia and perinephric abscesses can also be demonstrated.

Pathologically enlarged kidney, with irregular surfaces due to scarring and an adherent capsule was revealed. It is impossible to separate the cortex from the medulla, thinned parenchyma and dilated calyces with yellowish borders.

Histology reveals the characteristic foam cells which are Histeocytes replete with lipid droplets with inflammatory reaction of lymphocytes, plasma cells and giant cells. Glumeruli are fibrotic and hyaline together with fibrotic arterioles. Bacteria and fat droplets are present within the histocytes and neutrophils[26]. XGP tissue could arise from impaired drainage of suppurative foci or from the extension of the inflammatory process of the adipose tissue of the vascular adventitia[27]. It is currently believed that XGP starts as a localized reaction and then extends to the entire kidney[28]. Paediatric cases can remain limited[29] and at this age group diffuse form is less frequently associated with stones as opposed to adults.

Nephrectomy is the treatment of choice as in most cases the entire kidney is involved and there is destruction of the renal parenchyma. Surgery may