Proceeding S.Z.P.G.M.I. vol: 18(2): pp. 103-106, 2004.

Xanthogranulomatous Pyelonephritis (XGPN) with Fungal Infection: A Case Report

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SUMMARY

A case of xanthogranulomatous pyelonephritis with fungal infection is described. The patient was a known case of nephrolithiasis and thalassemia. He had developed pyrexia of unknown origin since two years. On further workup he was found to have hepatomegaly, para-aortic lymphadenopathy, massive left kidney and advanced renal failure with deranged renal function tests. The patient underwent left nephrectomy with the provisional diagnosis of advanced renal failure secondary to nephrolithiasis or renal Koch's. Histopathological examination revealed XGPN with numerous fungi present both in spore form and septate hyphae. Post operative recovery was un-eventful.

INTRODUCTION

Xanthogranulomatous pyelonephritis (XGP) is a rare type of pyelonephritis. We reported an interesting case of XGP with fungal infection, which one comes across once in life time.

CASE REPORT

A 25 years old young male presented with fever, dysuria, weakness, fatigue, weights loss and exert ional dyspnea of two years duration. He was a known case of thalassemia since childhood. There was a history of multiple blood transfusions. Ten years ago he had undergone spleenectomy. Two years after spleenectomy he developed urinary tract infection for the first time. Renal stones and gall stones were detected six years ago. Two years ago he underwent nephrectomy and percutaneous nephrolithotomy (PCNL). He had CRF since 2 years and had taken anti-tuberculous treatment (ATT) for six months.

Physical examination revealed a pale febrile individual with a longitudinal scar mark on the abdomen, due to previous operation. Liver was enlarged, with tender palpable left kidney. CBC showed Hb: 7.4 g/dl, TLC: 26,000/cumm, Platelets: 563,000/cumm. ESR was raised. He was positive for anti-HCV. Urinary sediment showed many RBCs and trace of protein, blood urea was 35 mg/dl and creatinine was 3.37 mg/dl. Ultrasonography revealed enlarged liver. Gall bladder revealed multiple stones. Right kidney was enlarged upto 14.3x5.2 cms with increased echogenicity, but no stones and hydronephrosis was seen. Left kidney was also enlarged and huge in size measuring 22x12 cms with bulbous upper pole. There was mild to moderate dilatation of pelvi-calyceal system with debris. Renal parenchyma was thick and hyperreflective with few enlarged para-aortic lymph nodes. A routine left nephrectomy was carried out through lumber approach and a huge kidney was excised with thick cord like ureter.

The specimen was received fixed in formalin, which measured on gross 24x14x12cms and weighed 2050 grams. The attached part of ureter measured 8 cms in length and upto 1.7 cms in width

The external surface of the kidney was well-encapsulated, bosselated, focally roughened and finely nodular. The kidney was surrounded by scanty perinephric fat. On sectioning the entire kidney was replaced by grey yellow nodular masses with focal necrotic areas. No normal appearing renal parenchyma was identified (Fig. 1).

The pelvicalyceal system revealed finely granular yellowish surface with focal blood clots. Multiple sections were taken from the ureter, hilar vessels perinephric fat and nodular areas in the renal

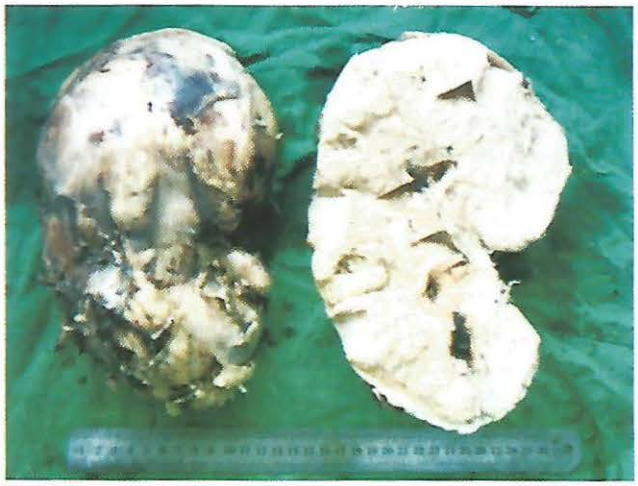

Fig. 1. Well encapsulated massive kidney weighing 2050 gms. Grey brown bosselated focal nodular serosal surface. Cut surface reveals dilated pelvicalyceal system fitted with blood clots.

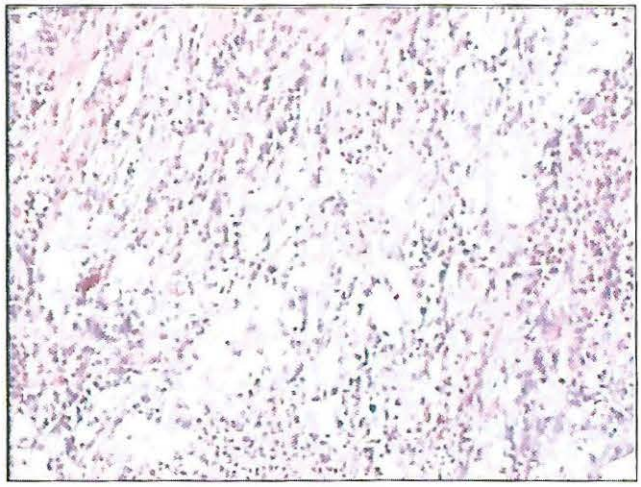

Fig. 2. Multiple granulomas with chronic inflammatory cell infiltrate (4x H&E stain).

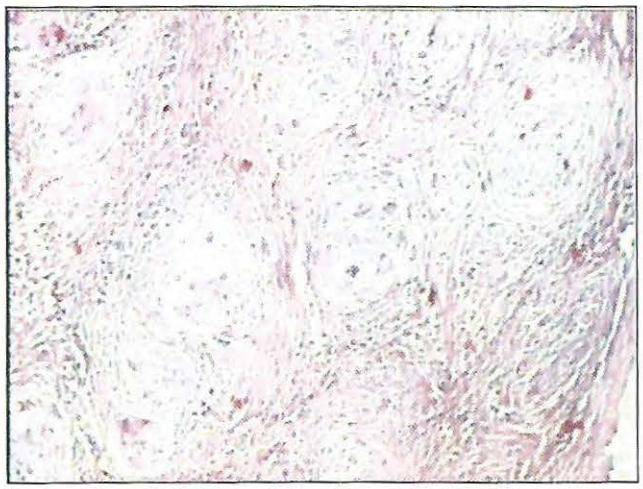

Fig. 3. Sheets of xanthoma cells and chronic inflammatory cell infiltrate (4x H&E stain).

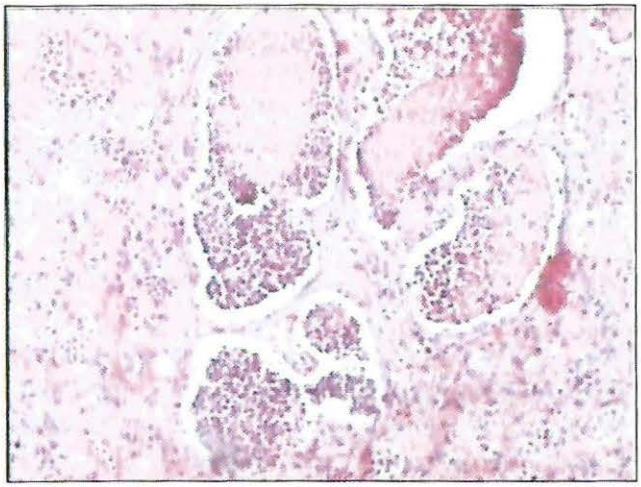

Fig. 4. Microabscesses (10x H&E stain).

substance. Microscopic examination revealed features of XGPN containing numerous granulomas composed of epithelioid cells with many multinucleated giant cells replacing most of the renal parenchyma. The intervening stroma revealed dense mixed but mostly chronic inflammatory cell infiltrate consisting of lymphocytes, plasma cells, many foamy and pigment laden macrophages and focally polymorphs forming micro-abscesses. The glomeruli in between this dense chronic inflammation revealed no significant pathology. Foci of proliferating renal tubules lined by benign epithelium were identified (Figs. 2-6. In addition numerous fungi were also present both in spore form and as septate hyphae confirmed on special stains *i.e.* PAS, Methenamine Sliver and Grocotte Gomori. Z.N (Fig. 7) stain for acid fast bacilli was negative. The case was diagnosed histologically as XGPN with fungal infection. Exact typing of the fungus can only be done after fungal culture.

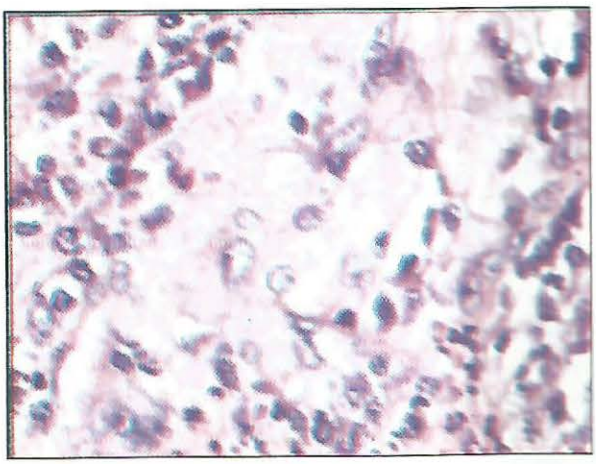

Fig. 5. Xanthoma cells, chronic inflammatory cells and few eosinophils with granular pink cytoplasm (40x H&E stain).

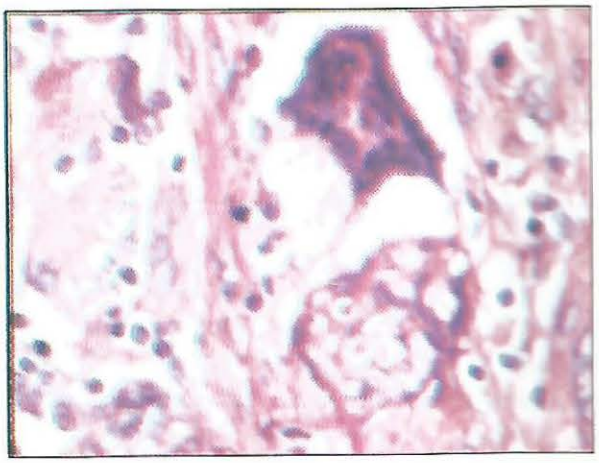

Fig. 6. Multinucleate giant cells with fungus in the giant cell (40x H&E stain).

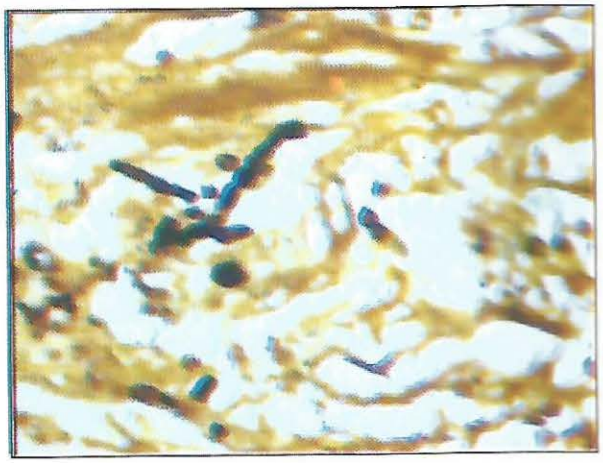

Fig. 7. Methenamine silver stained micrograph revealing dark brown fungus (40x).

DISCUSSION

XGPN represents an unusual suppurative reaction to chronic infection, often in the presence of chronic obstruction from a calculus, stricture or tumour[1]. The characteristics histologic feature of XGPN in the abundance of lipid laden macrophages (xanthoma cells)[1] as part of the inflammatory process resulting in the gross yellow appearance. Other components of the inflammation include lymphocytes, plasma cells, neutrophils and scattered multinucleated giant cells[2].

XGPN is a rare type of pyelonephritis which poses diagnostic difficulties on both clinical and histological grounds[3]. It is reported in literature under different designations i.e. staphlomycoses, foam cell granuloma, pyelonephritis xanthomatosa, renal xanthomatosis, chronic pyelonephritis with xanthomatous change, tumefactive xanthomatous pyelonephritis and XGPN[4]. Since no specific clinical picture exists so clinical history and imaging technique may prove to be unhelpful[5]. However biopsy of a renal mass diagnosed on ultrasonography may prove to be of some help. Histologically it closely mimics clear cell carcinoma (subtype of renal cell carcinoma). However a thorough histological examination fails to reveal any papillary or acinar arrangements of cells[6]. Furthermore nuclei are uniform in size, shape, stain-ability and usually reveal classical reniform morphology of histiocytic cells[1].

While diagnosing XGPN other differentials to be considered are hydronephrosis, avascular tumour, pyonephrosis, cystic renal carcinoma, lymphoma and renal tuberculosis[1].

In this particular case tuberculous infection and malignancy (clear cell carcinoma) was excluded by extensive sampling and thorough histological examination. The etiology of XGPN in this case seems to be fungal infection which is unique and we could not trace in literature any reported cases of XGP with fungal etiology.

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