INTRAABDOMINAL RETROPERITONEAL MALIGNANT FIBROUS HISTIOCYTOMA

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ABSTRACT

The malignant fibrous histiocytoma (MFH), a tumor also referred to as malignant fibrous xanthoma and fibroxanthosarcoma is a predominantly pleomorphic sarcoma usually occurring in the deep soft tissue of adults (1,2). This type of tumor also affects the bones (3). Most reports to date concern the soft tissues of the extremities; only 16% of the cases concern the abdominal cavity, where the retroperitoneum represents the most frequent site (1,4). Because of its highly variable morphologic pattern, this tumor has often been confused with other sarcomas such as pleomorphic rhabdomyosarcoma and liposarcoma.

This is the case report of MFH demonstrated by IVP, Barium bowel study, ultrasonography, CT scan and angiography.

CASE REPORT

A 50-year-old female patient, had weight loss of 4 kg, in one month. She had fever, anorexia. Physical examination revealed only thin woman without other abnormality. A previous history of adrenal mass, hypertension and hypokalemia 15 years ago. treated by medication. Plain KUB and IVP showed lateral displacement of left kidney and obliteration of the medial border of upper and middle pole of left kidney. The entire left psoas shadow was not seen, but there was no deviation of the left ureter, except at the proximal part (Fig. 1) UGI and small bowel series showed large LUQ mass, displacing the stomach medially and anteriorly. The DJ junction is also displaced medially; the jejunum is displaced inferiorly (Fig. 2). Pleated appearance of the splenic flexure was seen (Fig. 3). Ultrasonography showed a lobulated border solid low echoic mass, size 10 cm diameter at medioventral aspect of left kidney, obliterating ventral outline of left kidney. The echo of the mass was mildly inhomogenous, (Fig. 4) Noncontrast CT scan at the mass area showed a large isodensity mass (isodensity to the renal parenchyma). With contrast enhancement, the mass had central low density areas, in nearly entire mass. Other parts shows rather homogeneously enhancement. Invasion of the ventral part of left kidney, left psoas muscle. Left renal vein was included in the mass; however the rest of the left renal vein and inferior vena cava contained no thrombus. The left adrenal gland was displaced posteriorly. The pancreatic tail was attached to the mass. Separated enlarged nodes were not identified (Fig 5). Abdominal aortography and left renal angiography (Fig. 6) showed neovasculature to the mass from left renal artery. Retroperitoneal malignant fibrous histiocytoma, inflammatory type with invasion into the adjacent pancreatic tissue was found at surgery and pathology. The adrenal gland, left kidney and spleen were unremarkable.

DISCUSSION

In 1972, Kahn (5) reviewed a literature about retroperitoneal malignant fibrous xanthoma or xanthogranuloma, yielded 29 cases. The patient's ages ranged from 2 years to 73 years with a mean of 46 years. There were only three cases recorded in children

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UGI = Upper Gastro-intestinal, LUQ = Left Upper Quadrant,
DJ = Duodeno-Jejunal

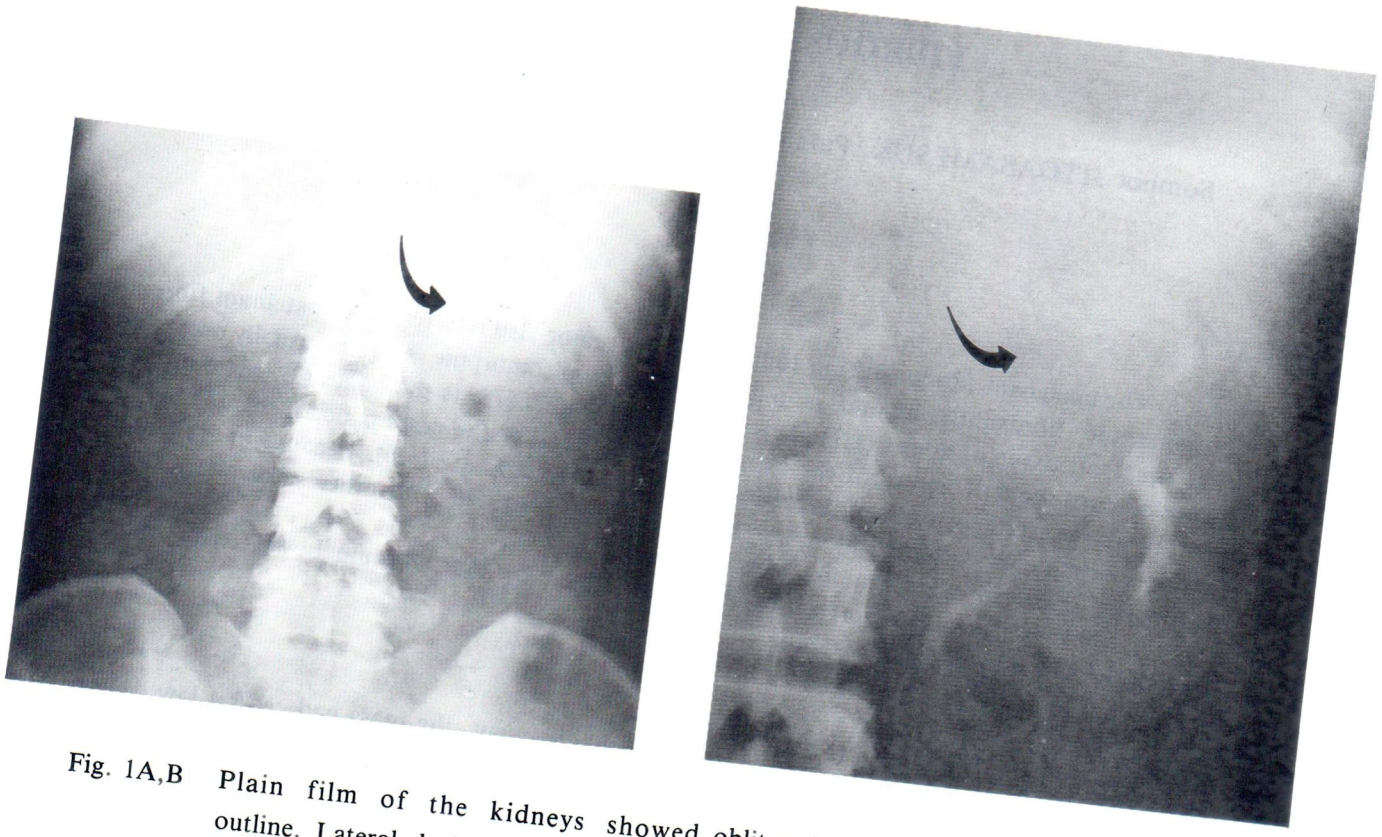

Fig. 1A,B Plain film of the kidneys showed obliteration of the upper pole-left renal outline. Lateral deviation of left kidney with loss of medial outline of upper and middle pole.

and only five cases in patients below the age of 30 years. The other cases were more or less evenly distributed over the next four decades with a peak in the sixth decade. Sixteen of the patients were males and 13 were females. Most of the lesions were circumscribed, non-encapsulated and lobulated retroperitoneal masses. In 13 cases, they were described as being large but no size was given. In the 10 cases in which the mass was measured, it varied in size from 6 to 20 cm in greatest dimension. It was occasionally adherent to adjacent retroperitoneal structures and complete excision of grossly visible tumor was not always possible.

Malignant fibrous histiocytoma is thought to originate from undifferentiated mesenchymal cells and is composed of fibroblast-like and histiocyte-like cell lines. Typically, these cells are arranged in a storiform pattern accompanied by pleomorphic giant cells and inflammatory infiltration. Fibrous, myxoid, giant-cell and inflammatory variants have been described (6).

Lane (7) described CT finding in 16 cases of retroperitoneal fibrous histiocytoma. The average tumor size on CT was 12 cm. Fifty-six per cents of the cases were seen on CT as muscle-density masses with regions of low density representing necrosis; 44 per cents showed smaller tumors with homogeneous muscle density; 25% contains areas of dystrophic calcification. From the work of various authors (8-11), it may be concluded that there are no computerized tomographic features specific for MFH or other soft tissue sarcomas except in tumors of high lipid content such as liposarcoma.

Sonographically, MFH usually presents itself as well-defined mass, the heterogeneity of the tumor giving rise to a variable echographic pattern (6). This variability seems to explain the different observations of Buecheler (12), Sarti (13) and Ros (9), who respectively describe the echographic aspects of MFH as mainly sonolucent (12), mainly solid-hyperechoic (13), and variable (9). Internal septation was also seen (6).

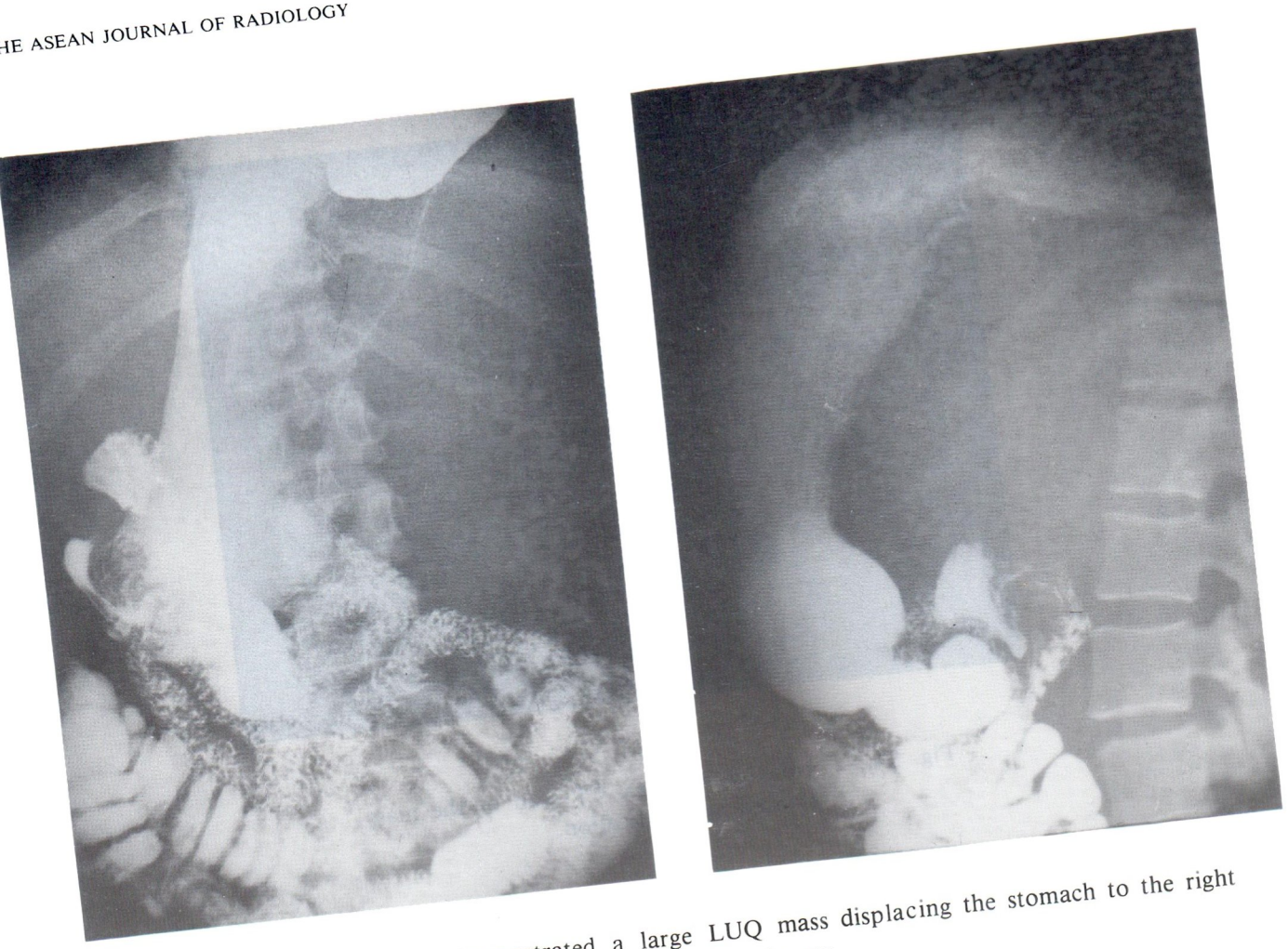

Fig. 2A,B UGI series demonstrated a large LUQ mass displacing the stomach to the right and anteriorly , indicating retroperitoneal mass.

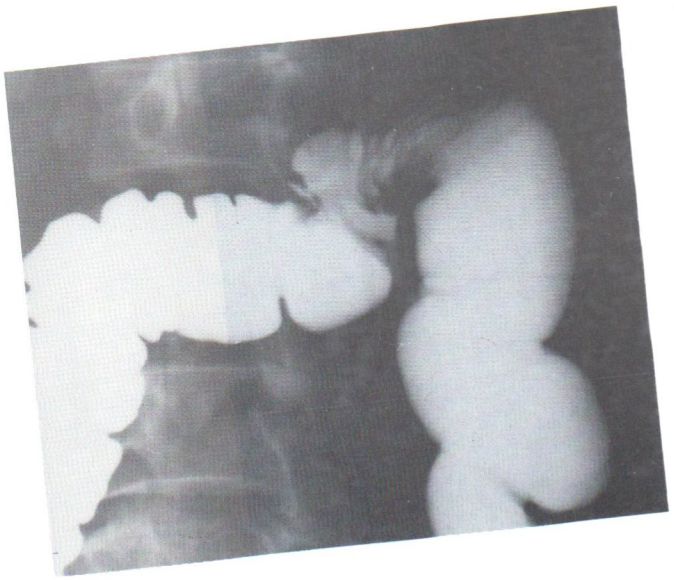

Fig. 3 Pleated appearance of splenic flexure due to infiltrated extrinsic mass was seen by Barium Enaema.

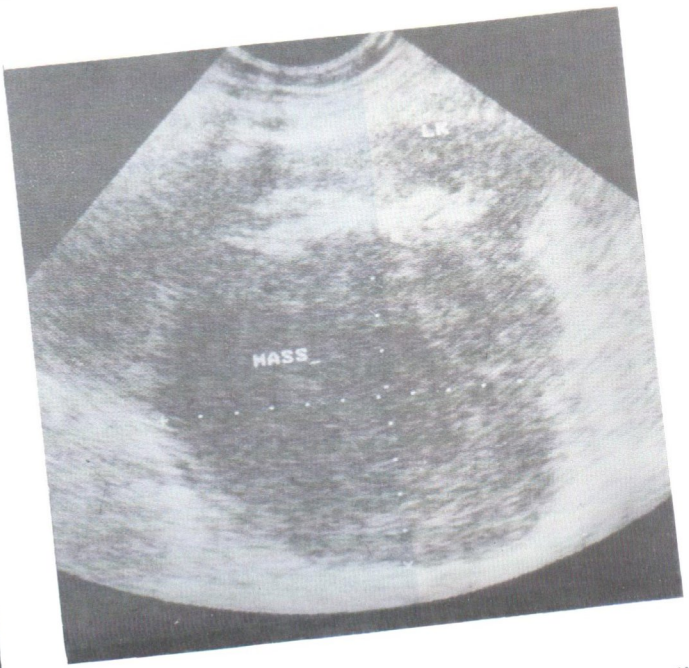

Fig. 4 An irregular border hypoechoic solid mass, ventral to left kidney was shown, obliterating ventral left renal parenchyma.

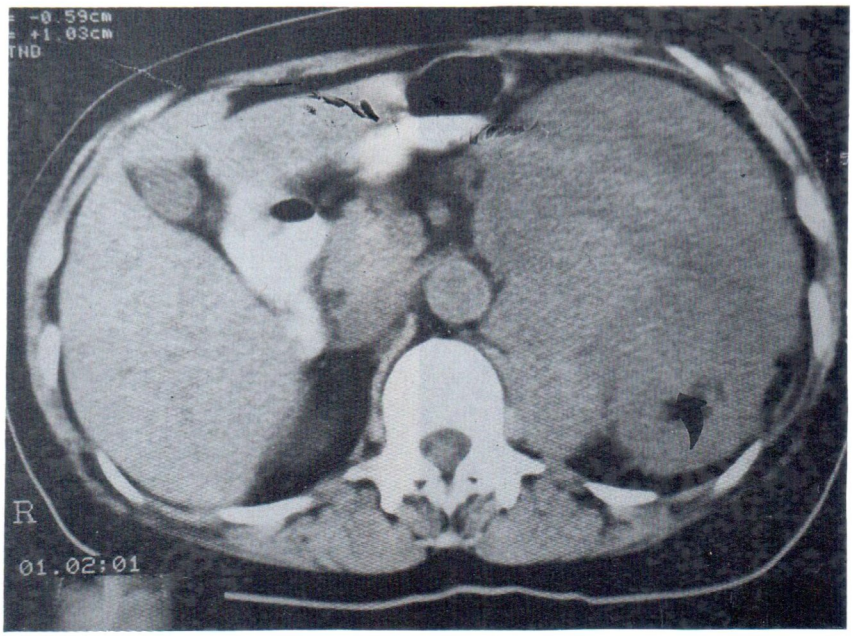

Fig. 5A. Non i.v. contrast CT scan of the mass showed no calcification.

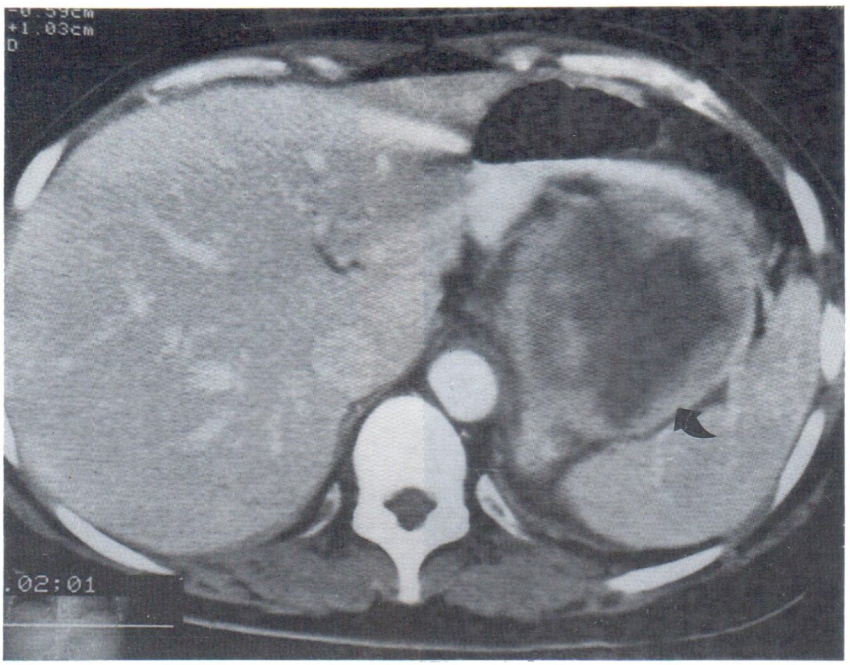

Fig. 5B. I.V. contrast CT scan of the mass showed rim enhancement, central necrosis and enhanced non-necrotic part.

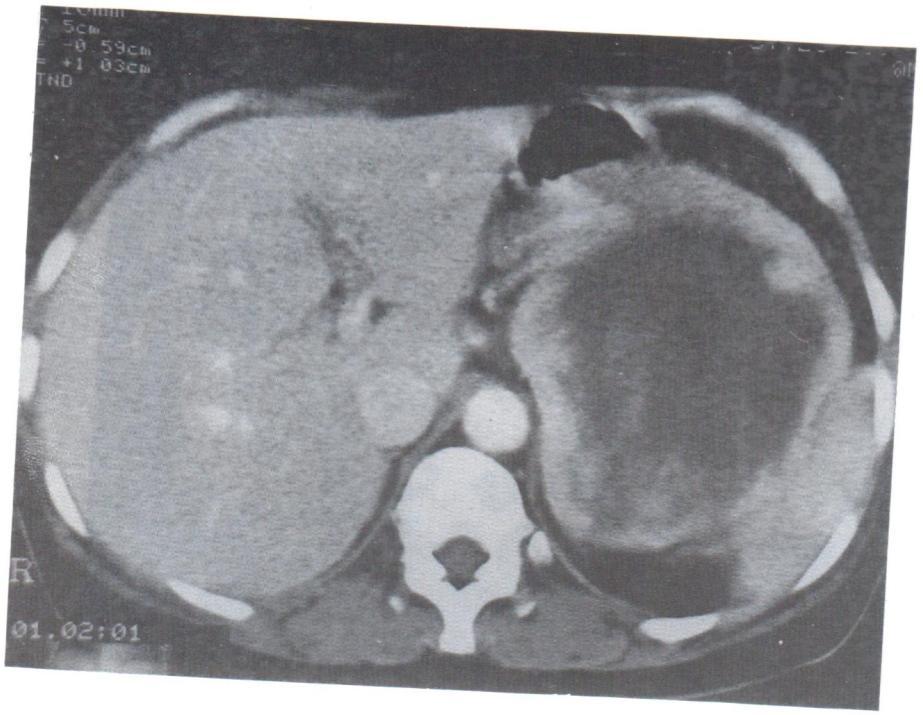

Fig. 5C. The unenhanced portion of the mass was at the central part.

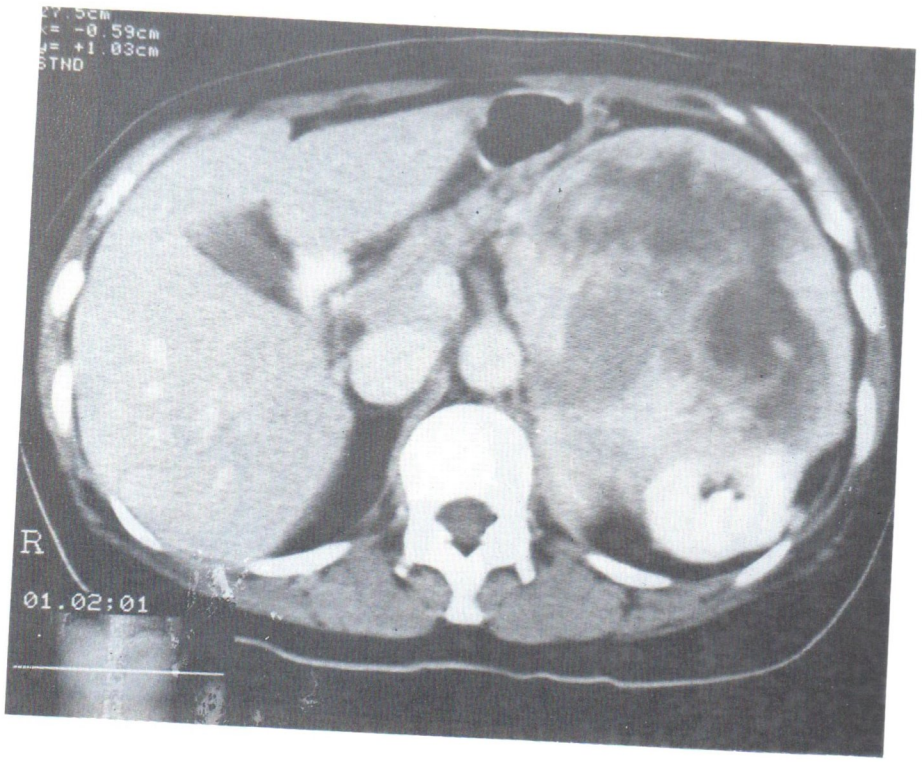

Fig. 5D. Left crus of the diaphragm, the pancreatic tail and ventral border of left kidney was infiltrated.

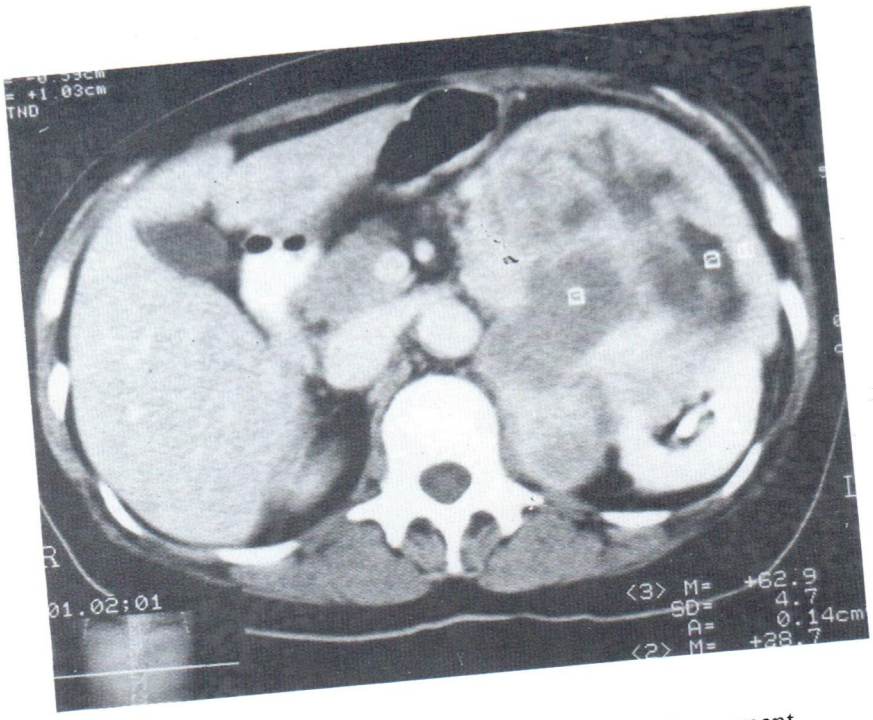

Fig. 5E. Inhomogeneity of tissue enhancement was shown.

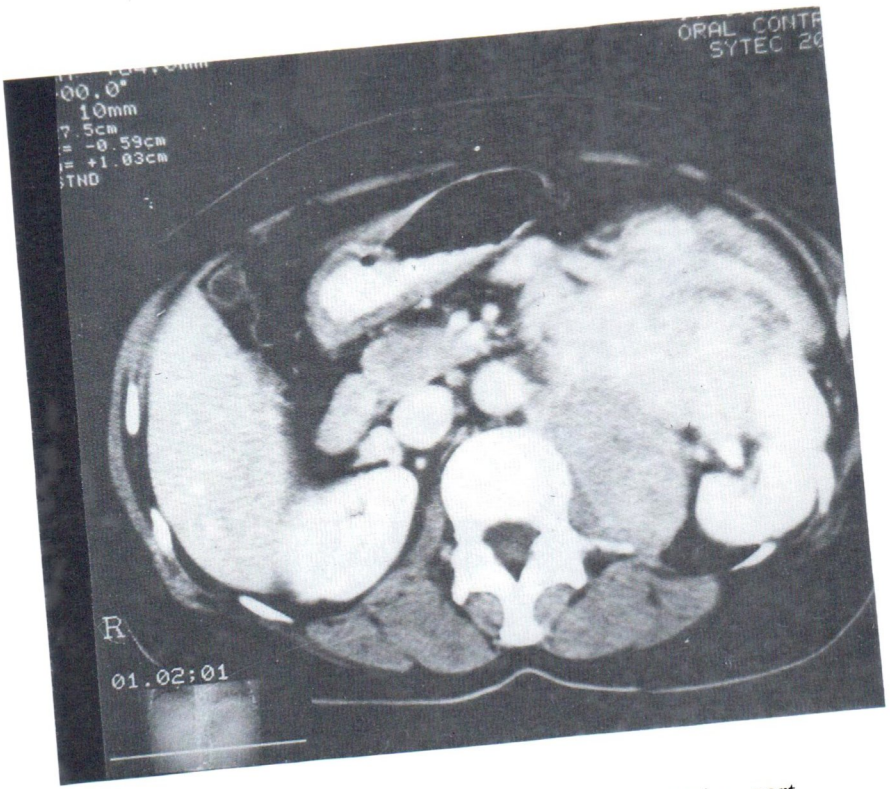

Fig. 5F. The mass extended to the anterior part of the abdominal cavity.

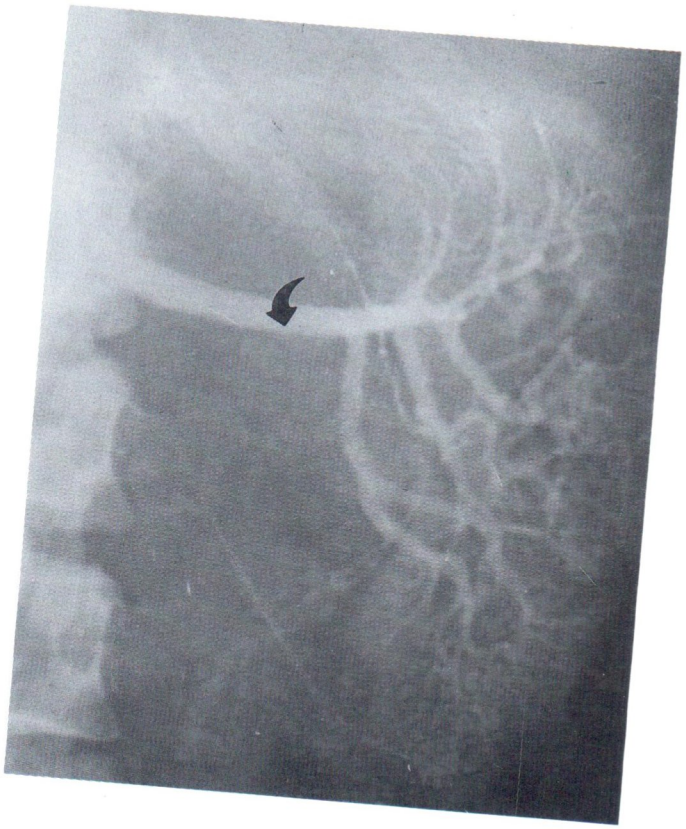

Fig. 6 Left renal angiography showed neovasculature arising from left renal A.

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