

A Case of Inferior Vena Cava Obstruction Associated with Budd - Chiari Syndrome

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The inferior vena cava obstruction is a relatively rare disease. The majority of the obstruction is located at the lower two third of the inferior vena cava, while the upper obstruction is frequently associated with hepatic vein obstruction (1,2). Approximately 200 cases of Budd-Chiari syndrome (3,4) have been reported in the literature (5). The association of inferior vena cava obstruction with Budd-Chiari syndrome has been occasionally reported, but an ante-mortem diagnosis of this condition is not necessarily easy. With the progress in the technic of angiography, venous catheterization or liver biopsy, successful ante-mortem diagnosis and surgical treatments increased (6-10), but the ideal treatment of this complicated disease has not yet been established.

Recently, we experienced a case of this rare condition and performed a shunt operation using an artificial vessel graft.

CASE REPORT

A 43-year-old male patient was admitted to our clinic, on August 27, 1964, with chief complaints of abdominal distension, cutaneous varices on the abdomino-lumbar region and edema of the legs. Past history was not contributory except for the first lumbar fracture at the age of 33.

Present Illness : The patient noticed edema of his legs in the middle of January, 1964, and was treated as nephritis without beneficial result. At the beginning of February, abdominal distension and cutaneous varices were developed without hematemesis, melena or jaundice. Because of colicky pains, he was admitted to Nagasaki City Hospital and then sent to the radiological clinic of Nagasaki University Hospital on August 23, 1964.

Clinical Findings : Physical examination on admission revealed a

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well-nourished man of medium stature without jaundice. His pulse rate was 72 per minute and regular. Blood pressure was 124/80 mmHg. No cardiac and pulmonary irregularities were noted. The lung-liver border was at the level of the fourth rib and the liver was felt straight underneath the right costal margin. The spleen was palpable one finger-breadth beneath the left costal margin. A definite fluctuation was demonstrated in the abdomen. Ascending varicose veins were noted on the abdominal wall and lumbar region as shown in Fig. 1. Swelling of the lower extremities was also remarkable.

Table 1. Laboratory Findings

	preop.	1 M postop.	5 M postop.	7 M postop.
1) RBC	539x 10 ⁴	424x 10 ⁴	390x 10 ⁴	440x 10 ⁴
Hb (g/dl)	13.4	12.0	8.7	11.7
Ht (%)	42	36	26	35
WBC	4,600	3,900	3,500	2,200
Thrombocytes	13x 10 ⁴	3x 10 ⁴		
Bleeding Time	1'	2' 30"		
Coagulation Time	5-19'	8-28'		
2) Urine Albuminuria	-	-	-	-
Glucosuria	-	-	-	-
Bilirubin	-	-	-	-
Urobilinogen	+	+	+	+
3) PSP 15 min. (%)	25	30		
120 min. (%)	70	60		
4) SMR (mm.)	9.3	10		
Seroreaction	-			
5) Icterus Index	4.7	5.0	7.3	5.9
BSP 45 min. (%)	11.2		15.8	
TTT	1.9	1.9	3.8	2.8
ZST	5.3	6.8	6.7	9.7
Total Cholesterol (mg/dl)	207	156	170	186
Alkaline P-ase (Bod. unit)	0-4	8.7	11.7	11.7
Cholinesterase (ΔpH/h)	0.45		0.27	0.28
A/G ratio	1.5	1.4	1.0	1.0
Total Protein (g/dl)	5.3	5.2	5.4	5.8
S-GOT (S. F. unit)	28	34	178	107
S-GPT (S.F. unit)	21	28	110	54
6) Serum Na (mEq/l)	139	137.5	138	135.5
K (mEq/l)	3.2	5.1	3.8	3.6
Cl (mEq/l)	101.2	106.2	98.2	99.0
Urea-N (mg %)	12.4	13		
Mucoprotein (mg %)	8.5			
Amylase (S. R. unit)	107.3			
Ammonia (r/dl)	212	213		
7) ASLO	50x		50x	
CRP	-		-	
RA-Test	+		+	

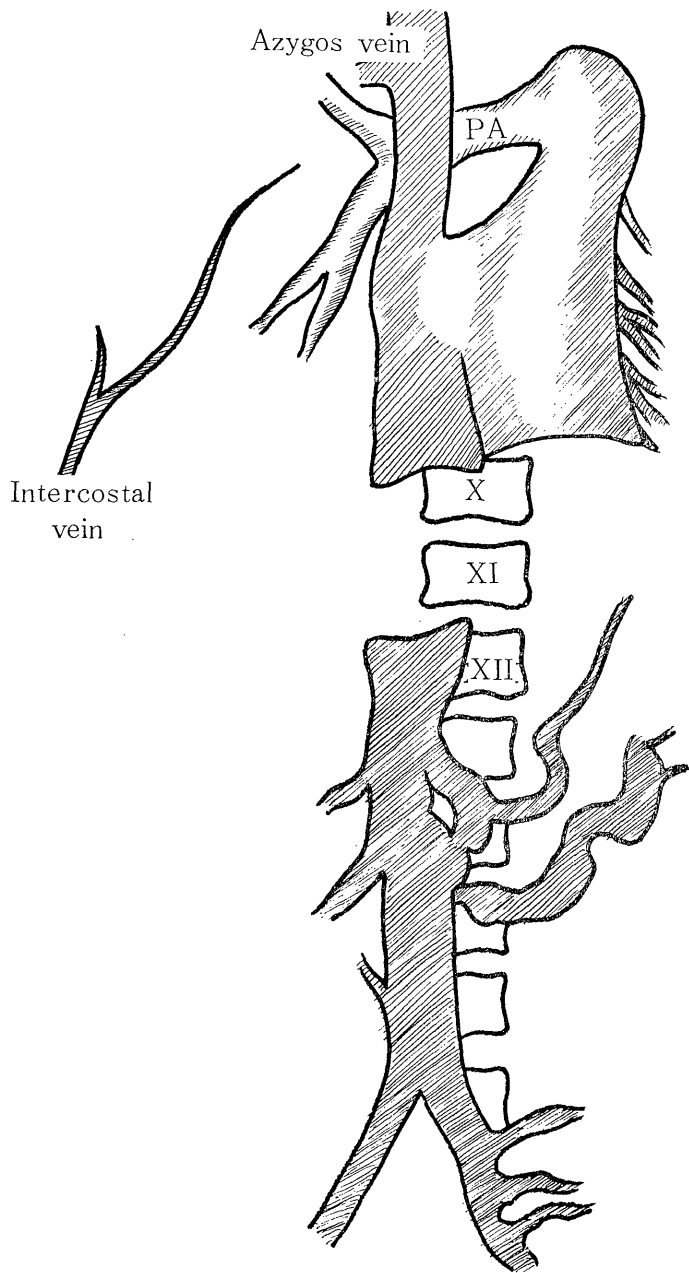
Laboratory Findings : Routine chest x-ray showed no abnormalities. Sinus tachycardia and low voltage were found electrocardiographically. Examination of the blood disclosed RBC 539×10^4 , WBC 4,600 with normal differential, Hb 13.4 g/dl, Ht 42%, bleeding time 1 min., coagulation time 5–19 min. Blood chemistry showed BSP 11.2 % in 45 min., cholinesterase 0.45 μ pH/h, serum proteins 5.3 g/dl, A/G ratio 1.5, total cholesterol 207 mg/dl, serum potassium 3.2 mEq/l, serum ammonia 212 μ g/dl, RA-test positive, ASLO 50x positive, serological reaction negative, PSP 25 % in 15 min. (see Table 1). Gastrointestinal series revealed moderate esophageal varices and displacement of the stomach due to hepatomegaly. Transsplenic portography revealed marked varices of the coronary vein but no infrahepatic portal obstruction (Fig. 2). Interrupted inferior vena cava at the 10th–12th thoracic vertebrae was revealed on venography. The interruption of the contrast media was 6 cm long and the hepatic veins were not visualized (Fig. 3, 4 and 5).

A diagnosis of the inferior vena cava obstruction associated with Budd-Chiari syndrome was made. High protein diet and administration of amino acids or bank bloods failed to improve hypoproteinemia. However, ascites was successfully controlled by administration of diuretics such as Rontyl and Diamox.

Operative Findings : On September 22, 1964, explorator, laparotomy was performed under G-O-F anesthesia. Ascites was minimal. The elastic firm right lobe of the liver had a nodular surface and appeared dark red in color. The left lobe atrophied to a size of a fist and the hypertrophied quadrate lobe projected to the small curvature of the stomach. The stomach and the biliary system had no abnormalities. The spleen was of a size of a palm. Retroperitoneum was incised at the anterior surface of the right kidney. There were diffuse fibrous proliferation rich of blood capillaries and the abdominal vena cava appeared to be thickened as like an artery (Fig. 6). Fibrous proliferation behind the liver was so pronounced that a retrohepatic approach to the vena cava was impossible. Thoracotomy at the right fourth costal bed revealed dense fibrous adhesion between the lower pulmonary lobe and the diaphragmatic pleura. The thoracic inferior vena cava was deformed to funnel-shaped due to fibrous narrowing of the caval hiatus of the diaphragm (Fig. 7). Incising the diaphragm, a cord-like mass, probably result of a thrombus in the right hepatic vein, was found on the superior surface of the liver. Using 30 cm long artificial vessel graft of Edward's woven Teflon with diameter of 1.4 cm, the abdominal vena cava was shunted to the right atrium passing through the anterior surface of the liver and the diaphragm incision (Fig. 8).

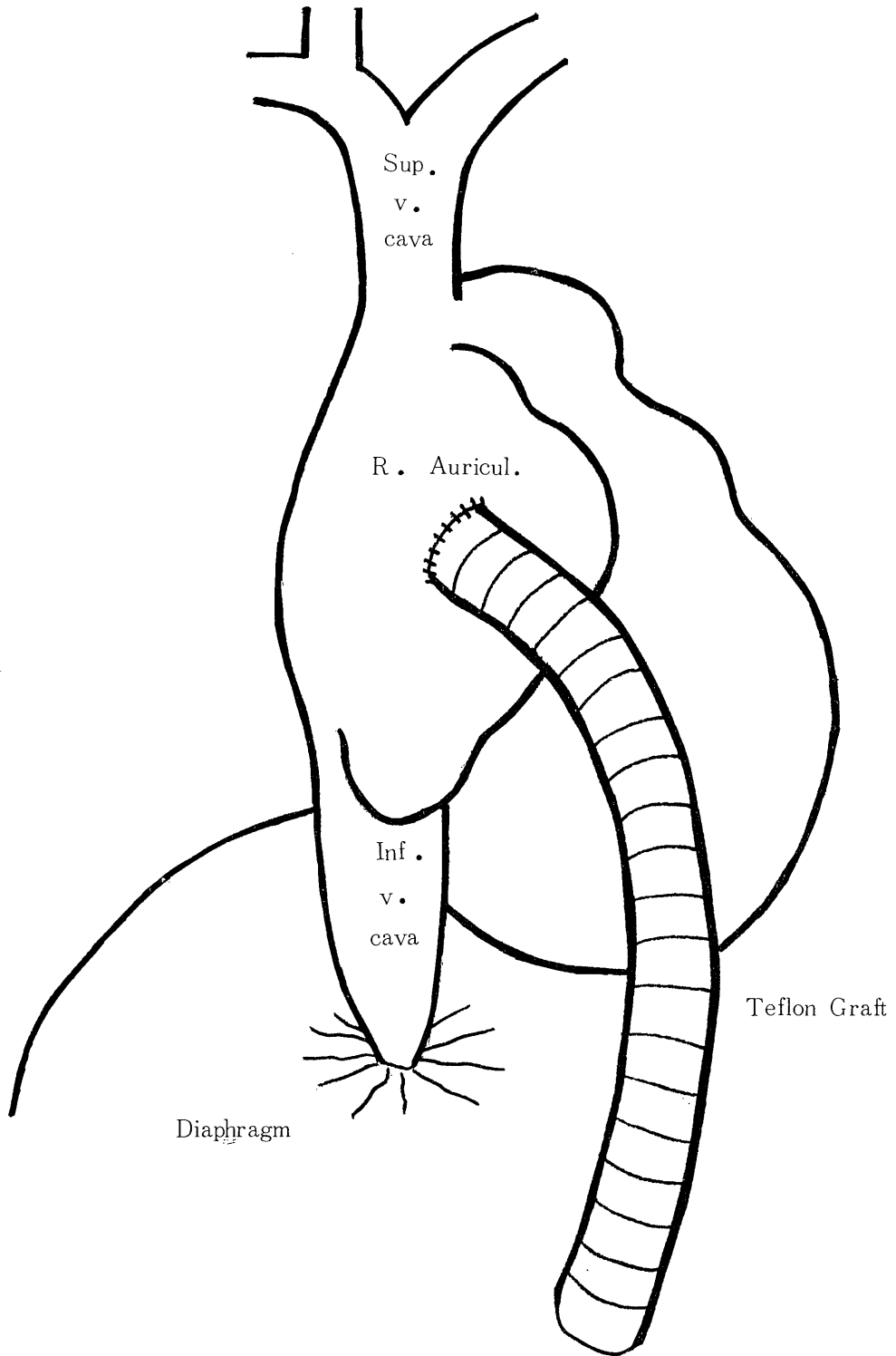
Portal pressure of 530 mmH₂O dropped to 420 mmH₂O after this shunting operation. Distension of the abdominal vena cava was markedly decreased. TALMA'S operation was also carried out in order

Fig. 5 Caval Venography



1. Inferior caval vein obstruction
2. Collateral circulation
 - lumbar veins—azygos and hemiazygos
 - intercostal veins —azygos vein

Fig. 7 Operative Findings



to augment collaterals. Microscopic findings of the right hepatic lobe were as follows ; fibrous proliferation, pseudo-lobulus formation of minimal grade, dilatation of the central veins, hepatic cell degeneration and regeneration, diffuse sinusoidal dilatation, round cell infiltration and no thrombi (Fig. 9-11). Cirrhotic process was minimal.

Postoperative Course : Cutaneous venous dilatation of the chest diminished transiently. Seven days after operation, ascites began to leak from the operative wound and lasted for 10 days. Venography, performed one month after operation, failed to visualize the graft shunt. Ascites and edema of the lower extremities re-appeared and daily aspiration of ascites were needed for 2-3 months. Three months after operation, cutaneous venous dilatation of the chest wall were extended (Fig. 12) and ascites stopped to multiply. At the fifth month intermittent hemorrhoidal bleeding were ensued.

This patient is now ambulatory and exhibits no abnormal hepatic functions.

DISCUSSION

H. CHIARI(4) emphasized the syphilitic phlebitis as a cause of Budd-Chiari syndrome. It is widely accepted, however, that there are very few syphilitic phlebitis in Budd-Chiari syndrome (11). POPPER (12) classifies the caused of this syndrome as Table 2. Chief causes of

Table II Etiology of Budd-Chiari Syndrome

- I. Abnormal processes in the vicinity encroaching upon the hepatic veins
carcinoma, tuberculosis, syphilis, abscess, cyst and cirrhosis
- II. Primary vascular disease
syphilitic phlebitis, rheumatic phlebitis, tropical liver injury
- III. Spontaneous thrombosis
polycythemia vera, compression of the veins in cirrhosis by hyperplastic nodules, retrograde embolism, obliterative endophlebitis
- IV. Congenital defect or malformations of the veins

Budd-Chiari syndrome are thrombosis of the hepatic veins, congenital anomalies (13), compression by surrounding tumors or inflammatory processes (14-20). GIBSON (21) insisted that the sphincterlike structure was a predisposing factor to manifest a noninflammatory thrombus. The obstruction of the inferior vena cava arises from similar causes including neoplasm of blood vessels (22-26), especially leiomyoma (27) and GRAWITZ' tumor (28), trauma, phlebitis (21, 29) or persistent Eustachian valve. KIMURA (2) demonstrated from investigation of human fetal blood vessels that the membranous obstruction was formed at the point where the right hepatic vein or the inferior vena cava joined with the left hepatic vein or Arantius duct. Membranous obstruction

of the inferior vena cava accounts for only one third of all obstruction (31). A laminar membrane of several centimeters length was reported by SUNADA (9). Inferior vena cava obstruction associated with the left hepatic vein obstruction was more frequently reported than the association with the right hepatic vein obstruction (2,22). In our case, a circumscribed interrupted shadow on angiography was considered to be membranous obstruction and subsequent thrombosis, but thoracotomy revealed a funnel-shaped obstruction of the phrenic vena cava secondary to severe fibrosis (Fig.7). Hepatic cirrhosis, predominant in the left lobe, and compensatory hypertrophy of the quadrate lobe were assumed to be secondary to preceding left hepatic vein obstruction. The striking thickening of the caval wall and retroperitoneal diffuse fibrosis were suggestive of collagen disease such as ORMOND'S disease (33-36). EISEMAN (39) reported a case of portal hypertension caused by idiopathic retroperitoneal fibrosis, and BARTHOLOMEW (40) described a case of sclerosing cholangitis associated with RIEDEL'S struma and ORMOND'S disease. Laboratory studies of our patient revealed positive RA-test and 50 x positive ASLO but no struma and mediastinal fibrosis. The past history of lumbar fracture was suggestive of a cause of retroperitoneal fibrosis but not appeared to be critical.

Acute type of Budd-Chiari syndrome manifests nausea, vomiting, ascites, hepatomegaly and frequently ends in death by shock and coma. It is essential to remove hepatic stagnation in acute type and EHRLICH (42) proposed a portacaval shunt. Chronic type is much more frequent than the acute type. The signs and symptoms of the chronic type are prolonged dyspepsia, upper abdominal pain, leg edema, and occasionally hepatoencephalopathy or extra-pyramidal signs (43,44). Liver function tests of the chronic type are not characteristic but abnormal BSP is frequently observed, probably due to circulatory disturbances as well as hepatic cell derangement. Some authors (12, 45) insisted that fat metabolism is specifically disturbed and a case of pan-hypometabolism was reported (46). Our patient had a total cholesterol of 207 mg/dl preoperatively and 170-186 mg/dl postoperatively. Jaundice is usually latent as in our case and obvious jaundice indicates underlying hepatoma or hepatic cirrhosis.

The most characteristic findings of Budd-Chiari syndrome are :

- 1) cutaneous venous dilatation without forming "Caput Medusae" on the abdominal wall and lumbar region in which blood flows cranially,
- 2) leg edema which appears prior to ascites or concomitantly with it. Leg edema never disappears even after subsidence of ascites. THOMPSON (41) regarded thigh edema as an indication of the inferior vena cava obstruction in addition to the hepatic vein obstruction.

Diagnosis depends upon venography of the inferior vena cava or hepatic veins. Measurements of venous pressure or investigation of collaterals are often of diagnostic assistance. Liver biopsy reveals

hepatic fibrosis (1,47) caused by venous obstruction including fibrous proliferation connecting Glisson's capsule and central veins, dilatation of sinusoids and irregularly shaped thrombi, proliferation of KUPFFER'S cells and peripheral round cell infiltration.

Treatment in acute type consists of anticoagulant, glutaminic acids and steroids for coma. There is no other means but to wait collateral formation by combined use of high protein diet, vitamins and glucose (1, 48).

Surgical treatments are 1) endovenectomy (6, 7), 2) transcardiac membranotomy (49), 3) by-pass operation, 4) palliative operation. Endovenectomy is the most radical operation removing the membrane and thickened parts of the venous wall. Occasionally, it needs to use Dacron patch suture of the venous wall in order to prevent postoperative stricture (7) and to take a shortest approach, namely, a posterior approach. In this case, the posterior approach was impossible because of retroperitoneal fibrosis. Transcardiac membranotomy was also impossible because the obstruction in this case was several centimeters long. This procedure is a treatment of choice in the case of the lower third obstruction of the inferior vena cava and it has a risk of perforation, embolism or re-obstruction. KIMURA performed this operation successfully in 4 out of 6 cases. By-pass operation is suitable to the causes inadequate to direct procedures. There are two methods of approach, retrohepatic and antehepatic approach. The retrohepatic approach is apt to result in stenosis or obstruction by hepatic compression. KIMOTO et al. (7) performed two by-pass operation using homograft and Teflon graft which ended in death due to hemorrhagic shock. OHARA (10) succeeded in shunt operation through retrohepatic approach using Dacron tube of 2.5 cm diameter intending to make a larger stoma. Sunada failed to perform antehepatic shunt operation using Teflon tube because of uncontrollable hemorrhage.

Thrombus formation in the Teflon graft in our case was probably due to too long by-pass, Teflon graft of too small diameter and a large dose of anticoagulants.

Palliative operation, including omentothoracopexy, omentorenopexy, hepatopleuropexy (52) and TALMA'S operation, has a hope of improving venous return.

TALMA'S operation was performed additionally without beneficial result in our case.

SUMMARY

A case of the inferior vena cava obstruction associated with Budd-Chiari syndrome was presented. By-pass operation between the inferior vena cava and the right auricle using Teflon graft by ante-hepatic approach and TALMA'S operation were performed,

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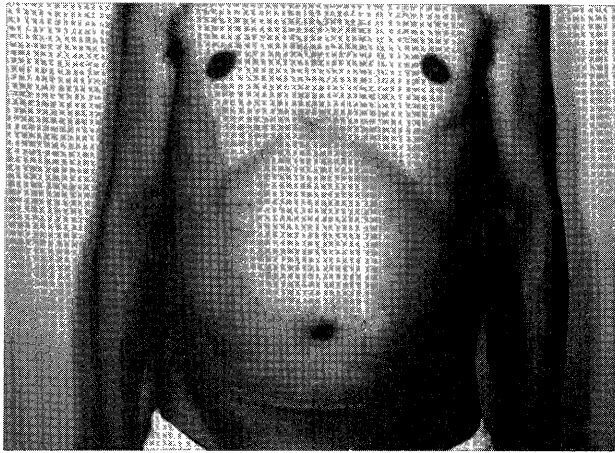


Fig. 1

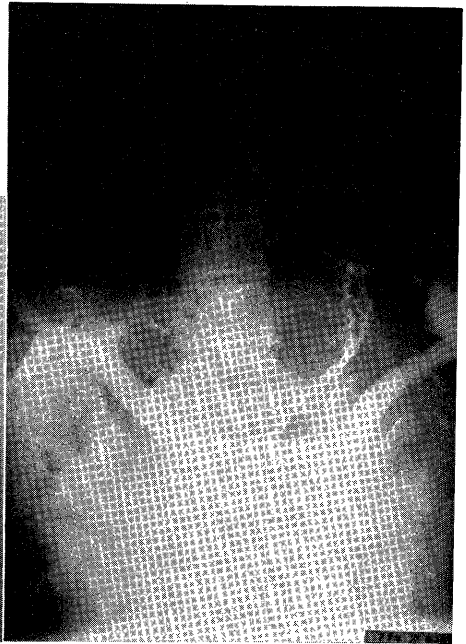


Fig. 2

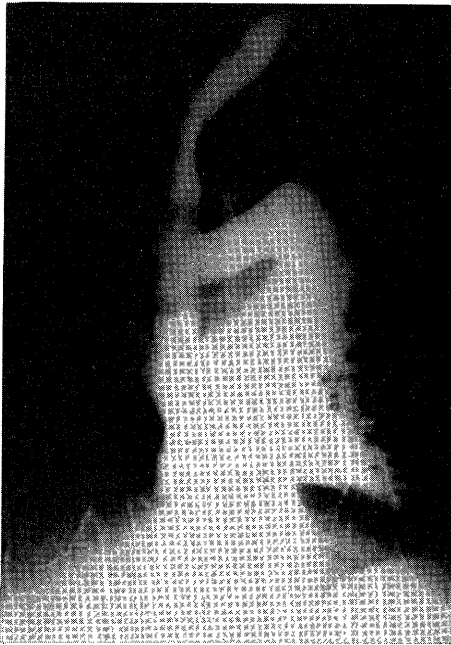


Fig. 3

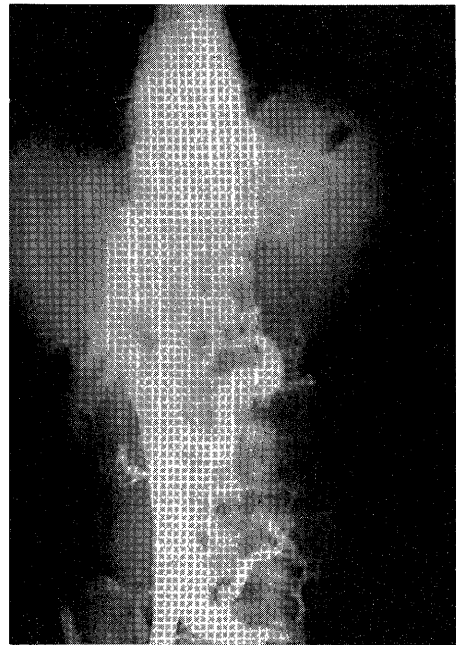


Fig. 4



Fig. 6



Fig. 8

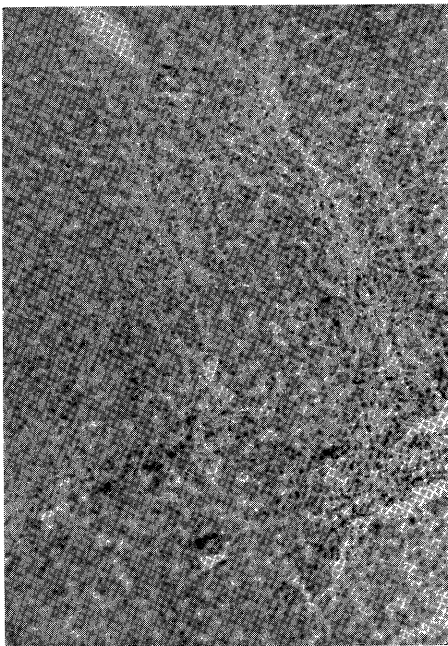


Fig. 9

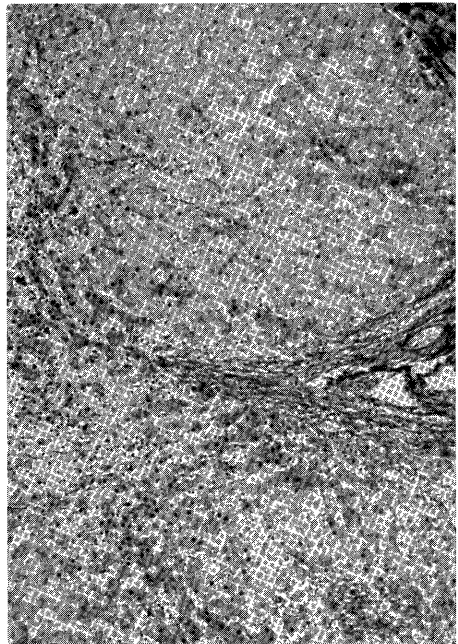


Fig. 10

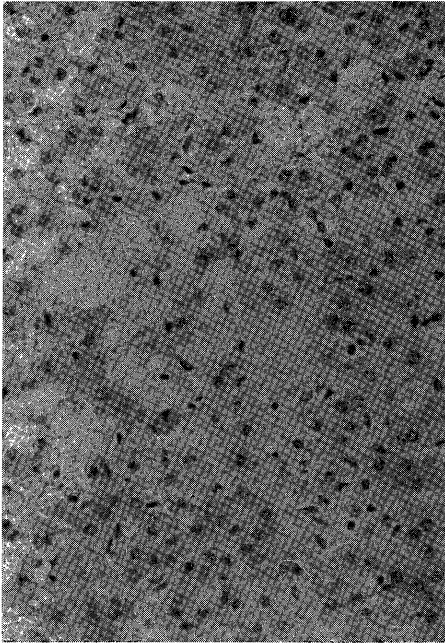


Fig. 11

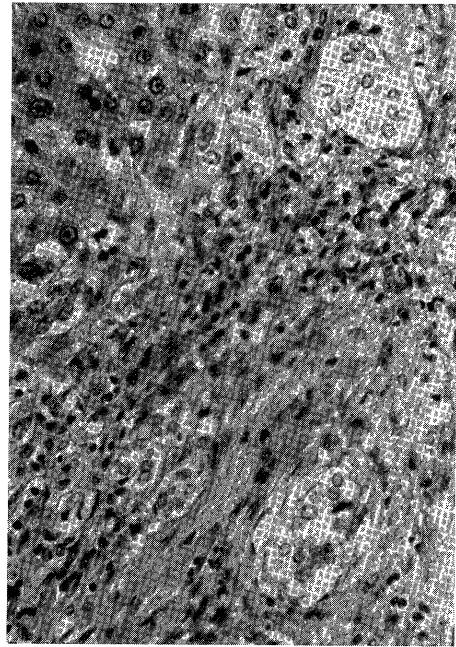


Fig. 12

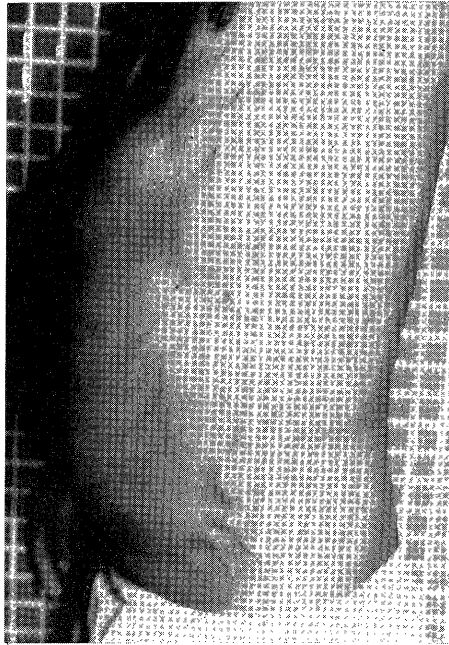


Fig. 13