Case Report

Pericarditis and Pleuritis Caused by Extramedullary Plasmacytoma

by

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Abstract

The following illustrates a case study of a 9 years-old girl with combined pericarditis and pleuritis caused by solitary extramedullary plasmacytoma. Pericardiocentesis and permanent thoracocentesis were performed, both yielded serohemorrhagic and serous fluid in succession.

In the beginning etiological diagnosis was made on the basis of the clinical pattern for tuberculosis infection and growth of three species of bacteria for bacterial infections and candida species for candidiasis. The initial treatment was in accordance with the etiological diagnosis mentioned above.

The final diagnosis was establish in the fourth month after the discovery of plasmacytoma in the pleural fluid and CT scan examination disclosing masses in the right lung.

Accordingly, cytostatic therapy was started. The result of therapy was very good, exudation into the pleural and pericardial sac regressed gradually and eventually ceased completely. According to the literature the prognosis of these neoplasma is good.

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Introduction

Plasmacytoma as an uncommon tumor. Martinson and Pulvertaft in 1961, could find only about 240 cases in the world literature. Plasmacytoma can be either a generalized disease involving the medullary portion of long bones, or else extramedullary. More than 90% of these solitary extramedullary plasmacytomas have been reported to develop in the head and neck (Batskis et al., 1964; Poole and Marchetta, 1968), whereas the extramedullary plasmacytoma of the upper respiratory tract comprised only 0.4% of malignancies in the area in a review of 2825 tumors at the Mayo Clinic (Webb et al., 1962).

Willis (1961) classified plasmacytoma into three groups: Group I: multiple myeloma, characterized by generalized bone involvement and characteristic X-ray findings, with frequently abnormal serum protein and Bence-Jones proteinuria. Group II: solitary plasmacytoma of the bone, with no evidence of a generalized disease. Group III: primary plasmacytoma of the soft tissue which can be single or multiple. Helmus (1964) stated that there may be an inter-relationship between these groups.

Pericardial disease is usually a manifestation of some systemic condition; except for rare traumatic pericarditis of the presence of an isolated pericardial cyst, it never occurs as an isolated disease. The most often cause of pericardial involvement in children is rheumatic carditis, whereas neoplastic disease is number six after rheumatic disease (Nadas and Fyler, 1972).

Many physicians consider the respective of a pleural effusion as an indicator of the cause of effusion. Weiss and Spodick (1953) reported in patients with pericarditis, that unilateral pleural effussion tends to be left sided and bilateral effusions to be larger on the left. Although the X-ray film does not show pleural fluid under a certain minimum, it is clear that whatever the level, left pleural effusions dominate the picture in pericarditis, particularly in the absence of cardiac disease. This conclusion is based on the result of 35 cases studied in 1983.

A case of severe exudative pericarditis in patient with exudative pleuritis caused by solitary extramedullary plasmacytoma is described. Due to the very complicated case especially in determining the diagnosis a long hospitalization was needed.

Case Report

A 9 years old girl was admitted to our hospital on 3rd January 1989, referred by a private hospital with left pleural effusion and pericardial effusion. The patient had been cared for 46 days at this hospital and had 8 times thoracocentesis, tuberculosis treatment and broad spectrum antibiotics.

On admission to our hospital, the patient revealed a history of breath shortness, mild fever, cough, and night perspiration, since two months before admission and had a contact with tuberculous patients.

On examination she was weak, without fever nor edema of the feet. Her position was limited with a tendency to lean forward. She was also undernourished, with a body weight 66% of standard (Havard P_{50}). Her pulse rate was 100 minutes, feeble and regular; blood pressure was 90/60 mmHg and a pulsus paradoxus of 15 mmHg was found. The jugular venous pressure was 10 cm saline. Respiratory rate was 32/minute, and the axillar temperature was 36.6°C. The apex beat was not palpable, the first and second heart sound were feeble, physiologically split

second heart sound, and no detectable murmur or rub were noted. Lung examination revealed dullness and hyporesonant at the caudal three fourth of the left lung. Small lymphnodes, as big as soy beans, were palpable on the left side of the neck. The liver was palpable as far as the umbilicus and tender. No ascites nor edema was present.

Laboratory investigations revealed a hemoglobin level of 15.9 gram/dl, with a hematocrite of 49%; the white blood cell count was 12.600/mm^3 with 73% segments 22% lymphocytes and 5% eosinophyls; erythrocyte sedimentation rate was 5 in the first hour and 15 in the second (Westergren). Blood chemistry revealed SGOT 22 U/dl (N: 5–15 U/dl), SGPT 9 U/dl (N: 5–19 U/dl), and LDH 365 U/dl (N: 80–240 U/dl), ureum 18 mg/dl, creatinine 0.84 mg/dl. Total serum protein 4.8 gram/dl, with albumin 3.0 gram/dl and globulin 1.8 gram/dl. Serum electrolytes were 132 meq/dl for sodium, 4.2 meq/1 for potassium and 104 meq/l for chloride. Sinus tachycardia, diminished QRS voltage, prolonged of PR and nonspecific ST-T wave changes were noted on initial ECG examination (Fig. 1). Chest X-ray demonstrated an enlarged cardiac silhouette with unmeasurable cardiothoracic ratio due to a half full pleural effusion on the left, and a few pleural effusion on the right (Fig. 2A).

An echocardiogram revealed a large anterior and posterior pericardial effusion with diastolic collaps on the right atrium (Fig. 3A).

The patient was immediately treated with furosemid, prednison and potassium chloride. Three days later isoniazide, ethambutol and vitamins were given. After two weeks medication no improvement was seen despite an increased urine output up to 900–1000 ml per day. Diagnostic and therapeutic pericardiocentesis were performed. Five hundred ml of pericardial fluid consisting in the begining of 300 ml serohemorrhagic and afterwards 200 ml serous fluid was aspirated. Microbiological studies of the fluid including examination for acid and alcohol fast bacilli as well as culture were negative. Cytologic examination revealed some erythrocytes, a large number of lymphocytes and a few polymorphonuclear leuchocytes.

Following the procedure a dramatic improvement in the patients symptoms along with the resolution of pulsus paradoxus and neck vein distention was noted. The echocardiogram showed marked reduction in the amount of pericardial fluid (Fig. 3B). Pleural effusion, however increased considerably.

Due to the increased pleural effusion, permanent left thoracocentesis (Underwater seal drainage = USD) was performed, and a drainage catheter was left in place. On the first day it yielded 2 liters of fluid and then the fluid production was about 500 ml per day. The total fluid yielded during 42 days was about 21 liters. On the 43rd day onwards no more fluid appeared from the drain. The first two liters was serosanguineous but further it was serous.

Analysis of pleural fluid revealed 100% polymorphonuclear leucocytes, protein content 3.6 gram/dl, NaCl 696 mg/dl, glucose 87 mg/dl. Test for Takahasi was negative. The first culture of the fluid yielded Pseudomonas sp which was sensitive to gentamycin, urotractine and amikacin. Anerobic, mycobacterial and fungal cultures, however, were all sterile. She was treated, therefore, with gentamycin in addition to the prior treatment.

Ten days afterwards, the condition of the patient was not improved and the second fluid culture was taken again and disclosed *Proteus* sp that was sensitive to gentamycin as well. Production of pleural fluid did not decrease until two weeks later and for that reason the third culture of

fluid was performed; the result was *Bacillus* sp. It was sensitive to gentamycin and chlorampenicol. Accordingly, chloramphenicol was added on the prior treatment. The response was very good, three days afterwards fluid exudation diminished and eventually ceased (Fig. 2B). PPD test was performed after two weeks free of prednison treatment with negative result. The patients was discharged from the hospital. Two weeks later the patient was rehospitalized with shortness of breath and a tendency to lean forward. 800 ml serous pericardial fluid and 700 ml serous pleural fluid was aspirated was also drained by thoracocentesis.

Fluid analysis revealed that microscopic and chemistry pattern showed the same results as before, but positive for *Candida* species. For that reason ketokonazol was added to the prior therapy.

All these therapy, however, failed to stop pleural and pericardial effusion. Fluid should be aspirated every three days, and the average fluid produced was about 800 ml from the pleural and 600 ml from the pericardial sac.

On April 3rd, 1988, the pathologist was able to identify plasma cell in the left pleural fluid, but not in the pericardial fluid. On the following day the second CT Scan examination disclosed a mass pattern in the right lung (Fig. 4) (The first CT scan was performed 3 months before with no abnormalities). Accordingly, we performed diagnostic right thoracocentesis and yielded 250 ml fluid with positive plasma cells. From these evidences, plasma cell tumor in the right lung as the cause of these effusions was established.

A skeletal survey of the flat and long bones did not show any evidence of multiple myeloma; there is no plasma cells increase in bone marrow aspiration and the plasma proteins were within normal limits, whereas Bence-Jones protein was positive in urine.

The patient was treated with chemotherapy (Alexan 50 mg per m^2 of body surface daily in four days every two weeks and Vincristin 2 mg per m^2 of body surface every two weeks, for the first series, during six weeks). After the seconds series (in three months), fluid production regressed gradually and eventually ceased completely. These treatments were continued until plasma cells in the pleural fluid were negative.

X-ray examination and diagnostic thoraco and pericardiocentesis hardly showed fluid in the pleural and pericardial sac.

Discussion

A rare and complicated case of plasma cell pericarditis and pleuritis was studied. The initial diagnosis of pericarditis was made on the basis of the combination of shortness of breath, a tendency to lean forward, pulsus paradoxus and enlarge cardiac silhouette, while diagnosis of pleuritis was based on a dullness on the left lung and X-ray findings.

In the beginning this patient seemed to be infected by three species of bacteria and by *Candida* sp. which grew in succesion. The first was *Pseudomonas* sp., the second was *Proteus* sp. and the third was *Bacillus* sp., all of them were sensitive to gentamycin. *Bacillus* sp., however was sensitive to chloramphenicol as well. For that reason, etiological diagnosis was based on the findings of the above bacteria, annd due to the similarity of the characteristic of the fluid in the pericardium and the pleurae, it was considered that the etiology of both effusions was similar. Tuberculosis however, owing to its high prevalence in Indonesia was initially considered as the etiological diagnosis in this case; also taking

into account that in spite of all negative tests, she had a history of contact with tuberculous patients, besides a history of cough and mild fever since two months and the fact she was undernourished.

Initial treatments consisted of tuberculostatics, antibiotics and antimycotics, however with no response up to two months. Discovery of plasmacytoma in the pleural fluid in the fourth month revealed that there was extramedullary plasmacytoma somewhere in the lung or in the upper respiratory tract, and CT scan examination disclosed its existence in the right lung. With cytostatic therapy the clinical signs and symptoms were improved.

Extramedullary plasmacytoma is a rare tumor of the lung that can only be diagnosed by biopsy and histopatological section. The most frequent sites are the upper respiratory tract and oropharynx (Dolin and Dewar, 1956). The 50–60 years age group is most commonly affected (Stout and Kenny, 1949; Webb et al., 1962), but it has also been reported in patients of five years as well as 80 years old (Youssef, 1971).

The usual symptoms of plasmacytoma are those due to the pressure or obstruction caused by the tumor. The patients may have occasional epistaxis. Pain is generally absent unless there is secondary infection or bone destruction (Chaudhuri et al., 1988). Our case is a very rare manifestation of plasma cell tumor.

In gross appearence, plasmacytoma vary from yellow grey to dark red in colour. They are generally smooth, without ulceration. The consistency is firm or rubbery. Histological diagnosis should be established and thorough evaluation should be made to rule out systemic involvement (Youssef, 1971).

The diagnosis of plasma cell tumor in this patient was established on the basis of the plasma cell finding in the pleural fluid and the existence of lung mass on the CT-scan. Lung biopsy was not performed. The negative results of bone marrow aspiration, bone survey and other clinical signs and symptoms of plasmacytoma suggested that it is a primary extra medullary tumor. We expected that metastasis had not occured seeing that no regional lymphnodes were found either physically at the neck or at the lung hilus on chest X-ray.

The regional lymphnodes tend to be involved, thus suggesting spread by metastases rather than the development of a separate primary lesion. Widespread cutaneous metastases have also been described (Emslie-Smith et al., 1955).

Once the diagnosis of plasmacytoma has been made, the lesion should be treated as malignant tumor. This potentially malignant tumor is generally treated by radiation, with about 50 per cent five-year survival. The therapeutic response of our case was very good, but according to Chaudhuri et al. (1988) local recurrence or disseminated disease can occur many years after original lesion has been succesfully treated therefore long-term follow up is necessary (Caudhuri et al., 1988). Whereas according to Dolin and Dewar 1956 and reviews of the medical literature, this neoplasma is associated with a good prognosis.

A. Samik Wahab, MD.

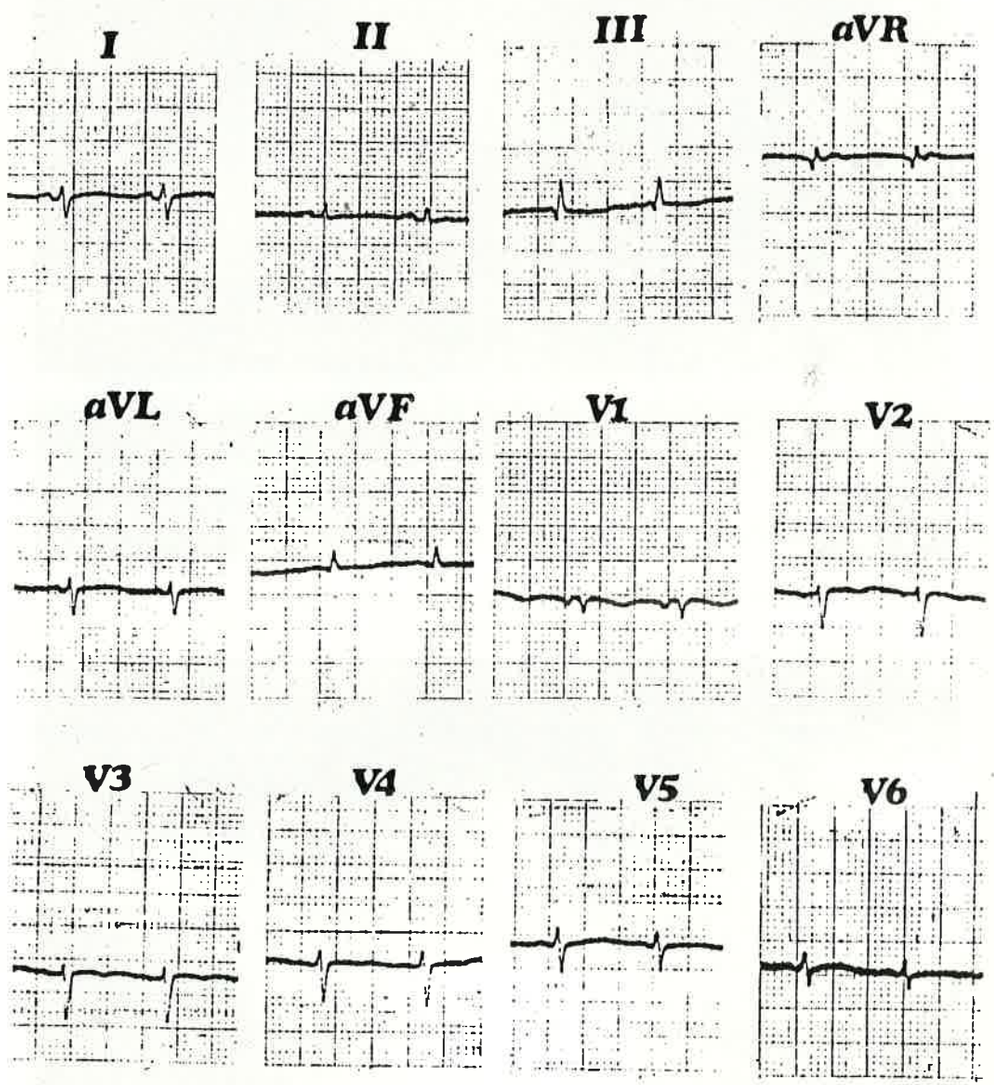

Figure 1 : *ECG - prior to tap demonstrating sinus tachycardia and borderline low voltage*

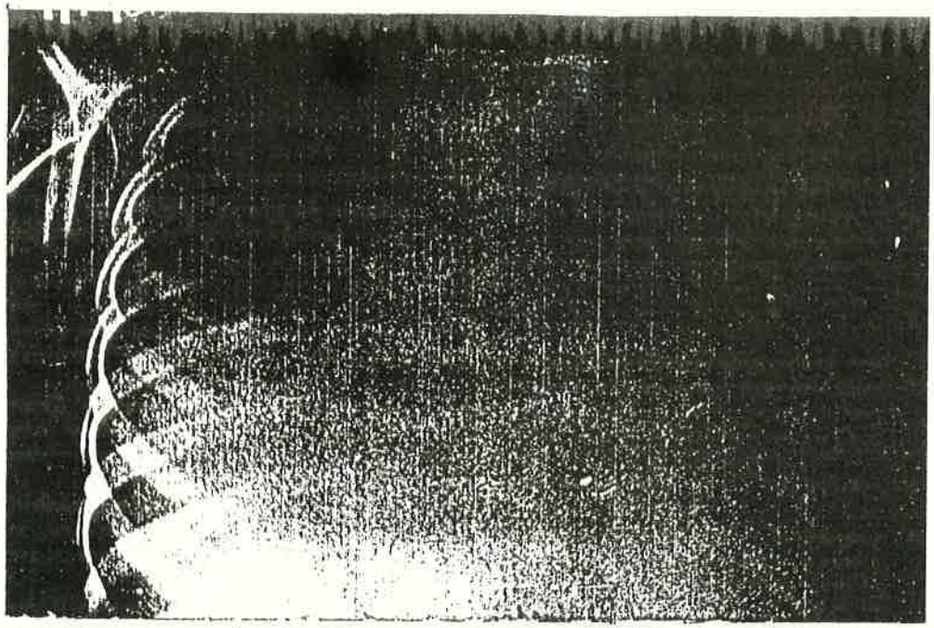

Figure 2A : *Chest X-ray AP view revealing large cardiac silhouette, central pulmonary arteries obscured by pericardial fluid, left and right pleural effusion*

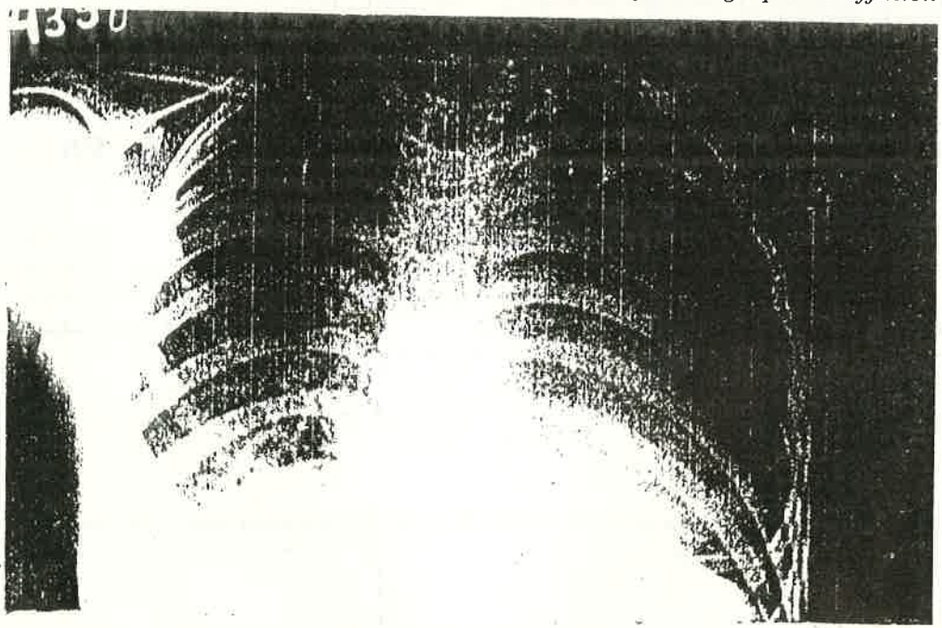

Figure 2B : *Chest X-ray AP view revealing large cardiac silhouette, taken immediately after permanent thoracocentesis*

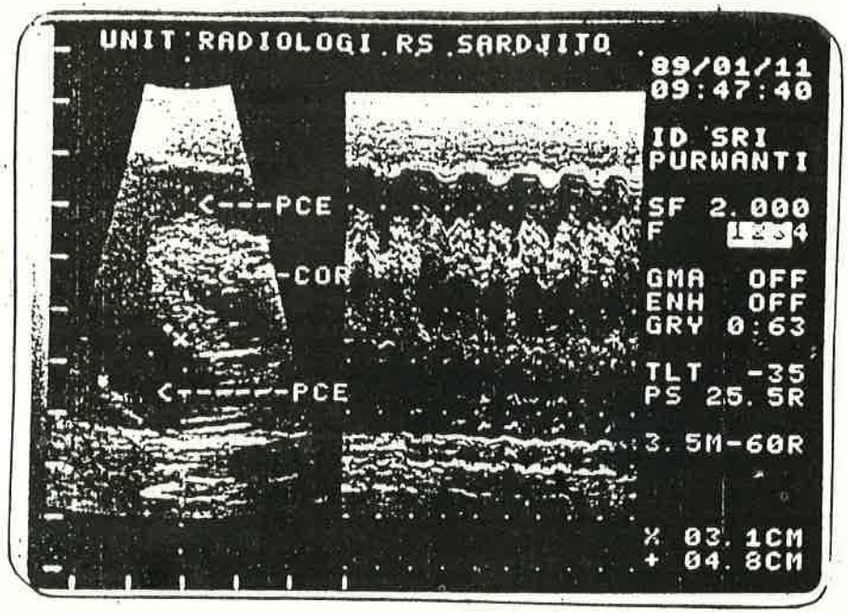

Figure 3A : *Echocardiogram demonstrating large anterior and posterior pericardial effusions prior to pericardial tap*
PCE = pericardial effusions Cor = Heart

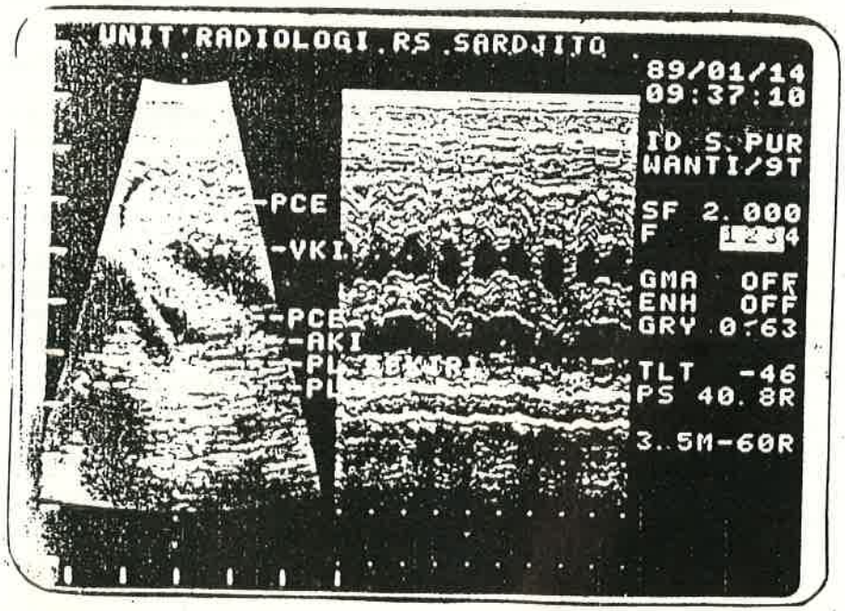

Figure 3B : *Echocardiogram taken immediately post pericardial tap, revealing considerable decrease of pericardial effusion*

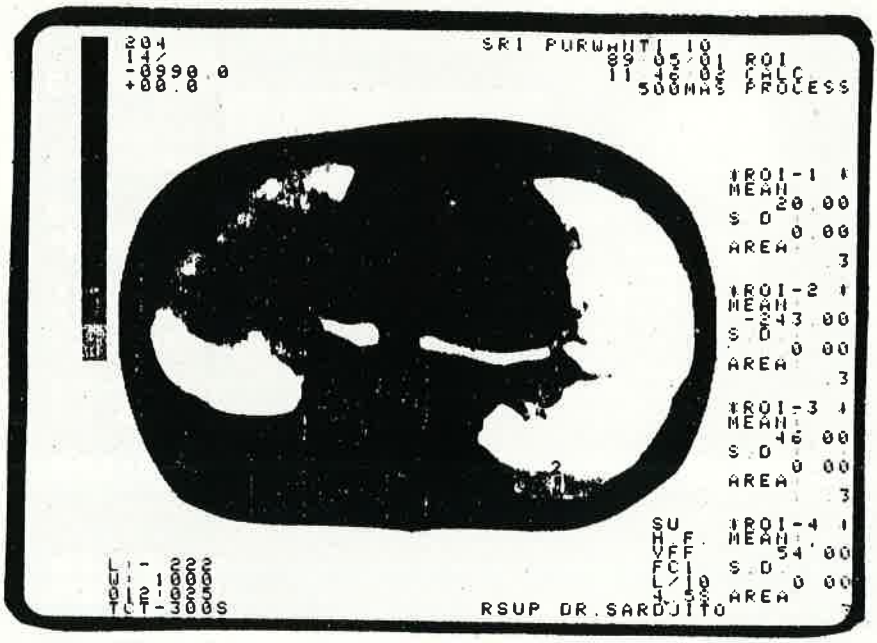

Figure 4 : *CT-scan X-ray revealing a mass pattern in the right lung*

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