RADIOGRAPHY

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PROGRESSIVE SYSTEMIC SCLEROSIS

röentgenographic manifestations of two cases

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HINDARTO LUHUR
RUSTIADJI

Department of Radiology
F.K. Unair/RSUD dr. Soetomo
Surabaya

PROGRESSIVE SYSTEMIC SCLEROSIS (PSS)

ROENTGENOGRAPHIC MANIFESTATIONS OF TWO CASES *

HINDARTO LUHUR **
RUSTIADJI ***

INTRODUCTION :

Progressive Systemic Sclerosis (PSS) is a disorder of connective tissue leading to fibrosis, vascular lesions and immunologic abnormalities that involves the skin and a variety of internal organs (gastrointestinal tract, lungs, heart and kidney) (3,4,5,11).

Since Goetz used the term "Progressive Systemic Sclerosis" in 1945 instead of Scleroderma to indicate that this is not primarily involving the skin but a progressive and generalized disease; the role of radiologist is important for suggesting the correct diagnosis of this disease. Especially if there are characteristic visceral manifestations with little or no skin alteration (10) or the skin changes being preceded by involvement of internal organs (8).

The gastrointestinal tract was the most commonly involved visceral organs especially esophagus (3,5,8). After esophagus, lungs and heart are second in frequency of internal involvement (3,5). Esophageal manifestation of PSS may be demonstrated by roentgenologic examination during that ... phase of the disease when the patient still has no clinical symptoms on which a diagnosis can be based (5).

We present two cases of PSS with their roentgenographic manifestations.

CASE REPORTS :

Case 1 :

A 39 year old female, race Madura, was seen at the rheumatologic clinic on October 9th 1984, stated that she had been troubled with joint

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** Resident of department of radiology RSUD dr.Soetomo/FK Unair Surabaya

*** Moderator/Staff of department of radiology RSUD dr.Soetomo/FK Unair Surabaya.

aching for approximately 3 years. The pain involved wrists, elbows, ankles and knees with swelling and stiffness of these joints. The pain was felt especially in the morning, when the weather was cold or the hands contacted with cool water that it made her can hardly to wash. She had developed hardening of the skin over the face and around the mouth, that made her difficult opening her mouth. She hadnot experience of fever and difficulty in swallowing food. Occasionly she had cough and exertional dyspnea. There was no family history of this disease.

Physical examination : Blood pressure 110/70. Pulse rate 84/min. The skin over the face was thickened in a smooth and shiny manner and the lips drawn into a small slit not extend than 4 cm. There was also thickened, inelastic and tightly bound down skin over the arms, the hands and the legs. The muscles of the fingers were atrophic and the tips of the fingers were anemic. The arm and knee joints were stiff especially the knees showed flexion deformities. Raynaud's phenomenon test was positive.

Laboratory findings : Hb 10,2 gr %. Blood sedimentation rate 72/105 mm. Others are normal. L.E. cell : negative. ANA test : negative. Rose Waaler test : positive . Electrocardiogram : normal. Pulmonary function test revealed mild restriction and no obstruction.

Roentgenogram of the chest was normal. Roentgenogram of the esophagus, the stomach and the jejenum was no abnormalities. Hands and wrists roentgenogram revealed only osteoporotic appearance of the bones.

Pathologic examination : Skin biopsy showed hyperkeratosis and parakeratosis. In the dermis there were dnese collagenous layer with hyaline degeneration, atrophic dermal appendages and some fatty cells.
Conclusion : the typical pathologic appearance of scleroderma.

Case 2 :

A 73 year old female, race: Javanese, entered the hospital on November 28th 1984 because of stiffness on the hands and knees, which had been felt for one and a half year. There was also swelling of the fingers, wrists and knees. She could hardly grasping her hands and opening her mouth widely. Recently she felt tightness of the skin with shiny surface and some white spots. There were no history of fever, dysphagia and dyspnea.

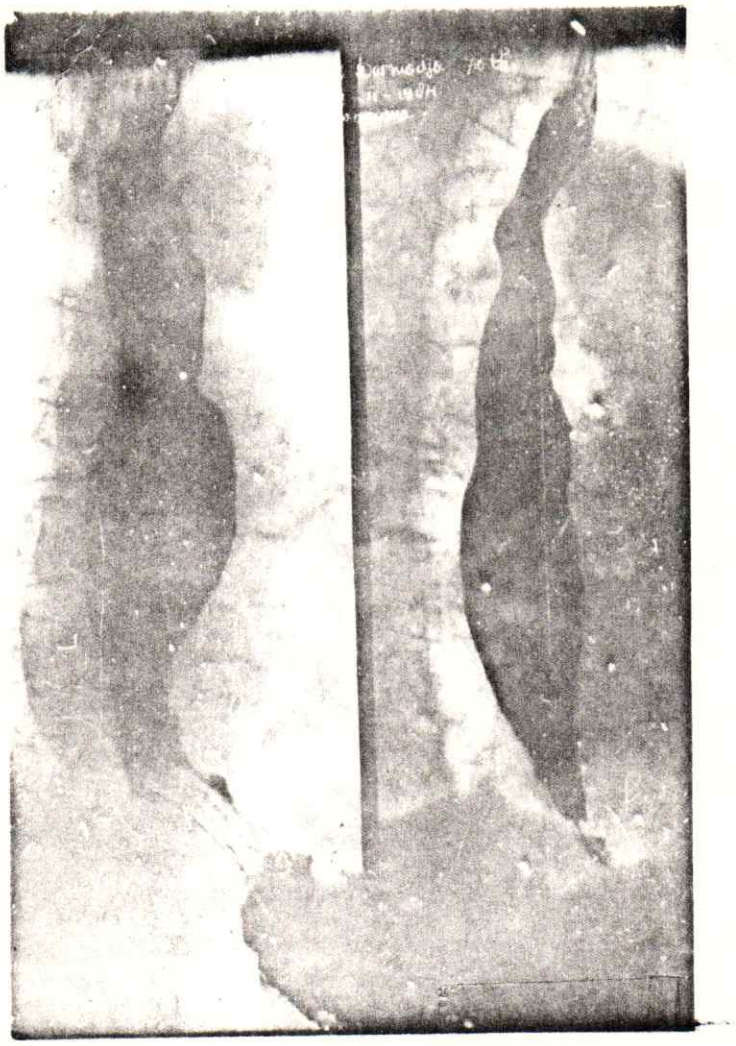
Physical examination : Blood pressure 110/65. Pulse rate : 76/min, regular. There were thickening and tightening of the skin of the face and extremities and narrowing of the aperture of the mouth. Hypopigmentation was visible in some parts of the lesions. The skin over the joints were shiny, thick and wrinkly. Extremities : the fingers were in flexion deformity with stiffness of the joints. Stiffness with limitation of the movement was also found in the wrists, the elbows and the ankles.
Raynaud's phenomenon test was negative.

Laboratory findings : Hb. 13,1 gr %. ESR : 8 - 22 mm. Others were normal. L.E. cells : negative. R.A. test : negative. ANA test : negative. Pulmonary function test was normal. Electrocardiogram was normal.

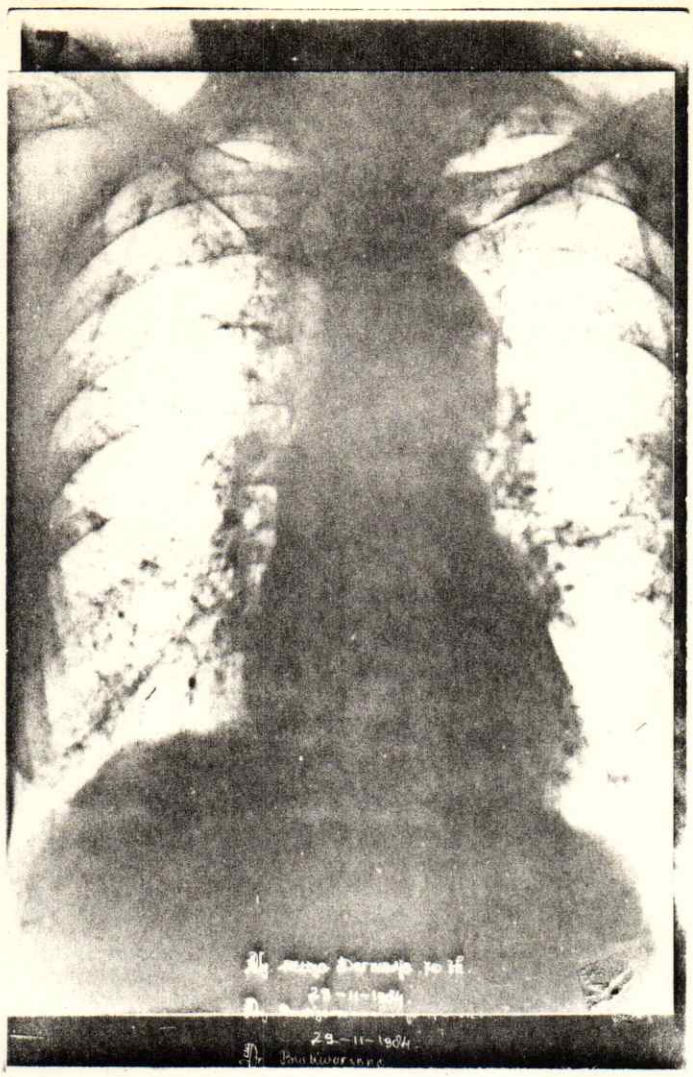
Roentgenogram of the chest revealed bilateral interstitial fibrosis throughout the lower two thirds of the lungs and mild enlargement of the heart with mild congestion of pulmonary vascularity. Roentgenogram of the hands and the feet showed diffuse osteoporosis. Esophagogram revealed dilatation with diminished of the peristalsis of the esophagus and constriction its distal part. There was also delayed of the passage of the barium through the esophagus.

Pathologic examination : Skin biopsy showed generalized hyalinization of the collagenous fibers in the dermis.

Conclusion : Scleroderma .



Roentgenogram of the esophagus of case 2 : Dilatation, hypomotility and constriction of distal part of the esophagus.



Chest roentgenogram of case 2 :
Bilateral interstitial fibrosis throughout the lower two thirds of the lungs.

DISCUSSION :

Scleroderma is only one of the symptoms and we are dealing with a systemic disease neither solely nor primarily involving the skin (Goetz). The generally accepted concept, that (5,8) :

1. PSS is one of the collagen (diffuse connective tissue) diseases.
2. It is a progressive disease leading to prolonged disability and commonly, to death of the patient.
3. The most clear manifestation of the disease : a leathery induration and sclerosis, generally occuring in the skin. The sequences of events is the same in every organ :(a) edema, followed by (b) proliferation of the connective tissue (thickening) and sclerosis of the collagenous bundles (induration) and finally (c) atrophy (vascular changes).

PSS is twice as common in females as in males and most frequently during the fourth and fifth decades (4,5).

Clinical manifestations : (4,5,8)

The initial symptoms usually appear slowly. One third of the patients were preceded by joint aching and stiffness which can proceed in months oryears before clinical diagnosis is apparent. An erroneous diagnosis of rheumatoid arthritis may be made (4,5). Further the skin become firm, thickened and leathery in appearance and tightly bound to the underlying subcutaneous tissue. The skin changes are most pronounced on the hands, usually with subsequent involvement of the face, neck and chest. Areas of hyperpigmentation or hypopigmentation may be present. The face assumes a mask-like appearance. The taut skin over the fingers gradually limits full extension, and fixed flexion contractures develop. These skin changes are present in our cases. Subcutaneous nodules (due to calcinosis) may be present. Raynaud's syndrome is common and this is described in one of our cases (case 1).

Gastrointestinal tract(4,5,8) : Dysphagia is the most common esophageal symptom, but it may not be present, even with abnormal X ray findings. In our cases, this symptom is absent. Other esophageal symptoms are : nausea, vomitus and substernal distress with burning sensation or actual pain which is more pronounced in the recumbent position.
Clinical syndrome of small intestinal obstruction may be present, when there is involvement of the small intestine. With involvement of the colon, the patient may be asymptomatic, or alternating constipation and diarrhea may be present.

Heart (4,8) : Cardiac involvement usually occurs late in the disease. There may be no symptoms in spite of considerable pathologic abnormality. The most constant features are cardiac enlargement, signs of cardiac decompensation secondary to pulmonary hypertension and electrocardiographic abnormalities.

Lungs (4,8) : Varying degrees of dyspnea can occur, cough may be either dry or quite productive. Pulmonary function tests often reveal impaired diffusing capacity (3,7,8) and at times restriction of vital capacity before there is any clinical or roentgenologic evidence of lung diseases (3).

Kidney (4) : Renal insufficiency may occur with or without hypertension.

Roentgenographic manifestations :

Joint involvement occurs frequently in PSS and is often mistaken for rheumatoid arthritis (5,7,8,9). Roentgenographic findings of the hands and wrists of these two diseases sometimes similar (i.e.: bone ero-

sions, narrowing of the joints, alteration of the ulnar styloid process, subluxation and even ankylosis of the wrist) (5,9). The earliest roentgenographic manifestation of rheumatoid arthritis i.e.: periarticular deossification (1) or juxtaarticular osteoporosis is spesific for differentiating from PSS, but later the osteoporosis can involve the entire bone and is then no longer of diagnostic significance (9).
Typical roentgenographic of the extremities of PSS are (2,5,8,9):

1. flexion contractures of the hands.
2. soft tissue atrophy over the distal phalanges.
3. soft tissue calcification, particularly in the hands.
4. localized bone resorption, the most common site is the tufts of the terminal phalanges in the hands.
5. When the bone erosions are present, they are often bilaterally, asymmetric and were sometimes located dorsally; features not characteristic of rheumatoid arthritis (2). This erosive arthropathy involves proximal interphalangeal, radiocarpal and metacarpophalangeal joints.

Only diffuse osteoporosis was found in the hand radiographs of our patients.

Gastrointestinal tract : Dilatation and decreased peristaltic activity (hypomotility) are the most common abnormalities demonstrated on roentgenologic examination. As a result, the passage of barium through the gastrointestinal tract may be delayed (5,8,10).

1. Esophagus (5,7,8,10,12):
   The esophagus is the common site of manifestation. It is widened and atonic with diminished or absent peristalsis. The findings are more pronounced in the lower portion and can be adequately demonstrated only when the patient is in horizontal position. The barium will remain in the esophagus for hours when patient keeps this position, but in erect position the bolus of barium will pass by gravity without delay. This helps to differentiate PSS from achalasia (5) or cardiospasm (8). Esophageal narrowing can be found associated with regurgitative esophagitis because of atony and dilatation at cardio-esophageal junction (8). According to this, short esophagus type herniation of the stomach through esophageal hiatus may be present (7). One of our cases (case 2), the esophagogram revealed dilatation, hypomotility and constriction of distal part of the esophagus. There is also delayed passage of barium through the esophagus.
2. Small intestine (5,6,8) :
   Dilatation of the small intestine are generally localized in the proximal portion. Involvement of the duodenum is common (5,6,8). This location may be one of the differentiating features from the sprue syndrome (malabsorption), which may present similar roentgenographic ma-

nifestation. The dilatation in the latter condition tends to be localized in the middle part of the intestine (5). Differentiation from small intestine obstruction must be made if there is very markaed dilatation and delayed in transit of the intestine in PSS. The most important differential diagnostic point is generally the clinical one, which the patient with obstruction of the small intestine is acutely ill(5). Pneumatosis intestinalis, which occasionally occurs, is seem as radiolucent cyst or linear streaks within the wall of the small intestine(4).

3. Large bowel (3,5,7,8) :

The colon may be atonic and dilated. Sacculation or "wide mouth diverticula" (3,7) or "wide necked diverticula" (6) is the most typical changes. This saccules are most frequently in the transverse and descending colon. An important feature of this abnormality is the asymmetric location of the saccules in their relationship to the haustrations. These saccules are visualized well on post evacuation film (8).

Lungs (3,4,5,7,8,11) : The characteristic finding is a bilateral diffuse interstitial fibrosis (linear pattern) especially in the lower two thirds of the lungs. Small cystic (pseudocystic) areas are also the characteristic manifestation and occasionally a large pneumatocele is present. These changes must be differentiated with sarcoidosis, pneumoconiosis, lymphatic spread of carcinoma, pulmonary fibrosis of Hamman Rich, histiocytosis, extensive bronchiectasis and other types of fibrosis (5). We found these changes in the chest roentgenogram of one of our cases (case 2).

Heart (5,8) : The heart may or may not be enlarged. When the enlargement occurs, it generally has a triangular shape which is similar to myxedema heart disease and pericardial effusion. The pulmonary artery segment may be prominent in patients who also have pulmonary hypertension.

Miscellaneous : Mandibular angle erosions with considerable bone destruction may be present (2). Teeth : Widening of the periodontal membrane was described by Stafne and Austin in 1944 and posterior teeth were involved more often than the anterior (2,5,8).

Recently some authors found that some patients with overlap clinical and radiological features of some collagen diseases, which could not found at postmortem examination (3), could be diagnosed as Overlap-PSS syndromes or Mixed Connective Tissue Disease (MCTD). MCTD appears to be an important subgroup of the collagen vascular disease in that there is a lower incidence of serious renal involvement and a more predictable response to corticosteroids therapy (11,14). There are no distinctive roentgenographic features to differentiate MCTD from clinical overlap syndrome of scleroderma and SLE. When features of both coexist in a single

patient, the radiologist may first to suggest the correct diagnosis of MCTD, after that serologic testing will confirmative (13).

Pathology of the skin (4,8) :

The early changes in the skin are those of edema and minimal perivascular infiltration (chiefly lymphocytes and plasma cells). The edema and infiltrarion diminish, and marked induration secondary to dense collagen changes becomes apparent. The collagen fibrils are marked thickened. Later there is atrophy of the epidermis. There is marked proliferation and fibrinoid degeneration of smaller vessels with partially or completely occluded. Dermal appendages are atrophied and rete pegs are lost.

Department of internal medicine diagnosed case 1 as probable PSS and case 2 as definite PSS according to Medsger criteria (table 1).

SUMMARY :

1. We have reported two cases of Progressive Systemic Sclerosis (PSS), one with minimal roentgenographic manifestation (i.e.: osteoporotic appearance of the bones of the hands and wrists only) and the other with some visceral roentgenographic manifestations (i.e.: esophagus, lungs and heart alterations).
2. Clinical and roentgenographic manifestations of the organs that can be involved in PSS, have been discussed.
3. Attention must be payed for clinical and radiological similarity of joints and bones manifestations of PSS, Rheumatoid arthritis and Mixed Connective Tissue Disease (MCTD)/overlap syndrome.
4. Knowledge of all possible manifestations of PSS is important to the radiologist, who may be the first to suggest the correct diagnosis, especially in those cases in which the skin changes are preceded by involvement of the internal organs.

Tabel 1.

Minimal Criteria for Diagnosis of Systemic Sclerosis in Hospital Survey

DEFINITE CASE :

Acrosclerosis and GI involvement.

Acrosclerosis and involvement of two internal organs, except GI (for example: heart, lung, or kidney).

PROBABLE CASE :

Acrosclerosis and one internal organ except GI.

Acrosclerosis with skin involvement of trunk.

Sclerodactyly with RP and GI involvement.

Sclerodactyly with RP and one internal organ, except GI, plus one other involvement (for example: muscle or joints).

No skin or RP but GI and one other internal organ involvement.

POSSIBLE CASE :

Acrosclerosis.

Sclerodactyly with RP and one internal organ involvement except GI.

No skin or RP but two involvement, including one internal organ (for example: lung plus joints).

GI = Castrointestinal; RP = Raynaud's phenomenon.

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