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RADIOLOGICAL FINDINGS IN PROGRESSIVE SYSTEMIC  
SCLEROSIS (P.S.S.)

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INTRODUCTION

Progressive systemic sclerosis is a generalized connective tissue disease involving the skin, muscles, bones, gastrointestinal tract and other internal organs. It occurs most frequently during fourth, fifth and sixth decade of age.

In P.S.S. the clinical manifestations are not always obvious, and radiological results can usually play a significant role in early diagnosis. It is therefore, of importance that radiologist be aware of all possible lesions.

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This report concerns the roentgenological findings in 20 patients referred to ~~Esomora~~ Medical School university of Tehran with emphasis on the unreported case of mid esophagus narrowing and some uncommon cases of upper lung fibrosis and pseudodiverticula of small intestine.

### RESULTS

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X-rays of 20 patients, 17 women and 3 men were studied. These patients were selected because of positive roentgen findings. The age of the patients, duration of disease prior to admission and clinical symptoms are shown in table I and roentgenological evidence of involvement of gastrointestinal tract, chest, hands and teeth are summarized in table II.

### ROENTGEN FINDINGS

Esophagus:- Esophagus was normal in 6 patients and in the rest it was atonic. In the latter, case 5 showed narrowing of the esophagus, with one case of hiatal hernia (fig I). There was a patient (case I) with intermittent dysphagia for 15 years. On examination she had nail atrophy and fingers' ulceration, and possibility of benign tumour, esophagitis, esophageal web and congenital stricture were suggested. Barium swallow showed spasm of entire esophagus with stricture of mid portion and

Table I

Case No	age	sex	clinical findings	duration
I	37	f	dysphagia, finger atrophy & ulceration	15 years
2	28	f	atrophy and ulceration of finger tips	8y
3	48	m	dysphagia, dyspnea, stiff hands	1y
4	60	"	joint pain and swollen hands	1 mo
5	18	f	stiff hands, finger's ulceration	2y
6	32	"	stiff hands, dysphagia	2 y
7	34	"	raynaud's phenomenon, dysphagia	5 y
8	42	"	raynaud's phenomenon	1 y
9	30	"	raynaud's phenomenon	1y
10	52	"	raynaud's phenomenon	30 y
II	55	"	stiff hands, painful joint, dysphagia	2y
I2	20	"	stiff hands, painful joint, dysphagia	3y
I3	40	"	stiff hands, painful joint	6 mo
I4	24	"	stiff hands, dysphagia	1y
I5	17	m	stiff hands, heart-burn, dysphagia	7y
I6	34	f	stiff hands	1/5y
I7	22	"	painful joint, stiff hands	3y
I8	32	"	painful joint, dysphagia	1y
I9	45	"	heart-burn, dysphagia	1y
20	35	"	stiff hand, painful joint, dysphagia	1y

Table II-Roentgen findings

Case No

I	stricture of mid esophagus, esophageal web, normal chest
2	esophageal dilatation with distal narrowing, normal stomach and small intestine, lung fibrosis with honey-comb shadow of upper lobes, terminal phalangeal resorption with soft tissue calcification
3	atonia and dilatation of esophagus, normal stomach, dilatation of second portion of duodenum with barium retention, hypomotility of small bowel with sacculations of one segment of jejunum. lung fibrosis with honey-comb shadow of lower lobes, normal hands
4	atonia and dilatation of esophagus, normal chest
5	dilatation of esophagus with distal narrowing, normal stomach and small intestine
6	atonia of esophagus
7	atonia of esophagus, normal stomach chest and hands
8	resorption of phalangeal tufts, GI series normal, normal chest
9	atonia and dilatation of esophagus, resorption of phalangeal tufts and styloid process, with diffuse osteoporosis
10	atonia of esophagus, phalangeal tufts resorption
II	dilatation with distal narrowing of the esophagus fibrosis of the lung, phalangeal tufts resorption

	with osteoporosis
I2	dilatation with distal narrowing of the esophagus small bowel dilatation and long transit time, fi- brosis of the lung with honey-comb shadow of lower lobes, pointedness of terminal phalanges with osteoporosis
I3	lung fibrosis, osteoporosis of the hands, normal esophagus
I4	atonia and dilatation of esophagus, delayed empt- ying of stomach normal chest
I5	narrowing of distal esophagus, delayed emptying of stomach, resorption of phalangeal tufts with os- teoporosis, normal chest
I6	phalangeal tufts resorption, normal GI series and chest
I7	phalangeal tufts resorption with osteoporosis, normal GI series and chest
I8	phalangeal tufts resorption, normal GI series and chest
I9	normal esophagus and stomach, duodenal ulcer, heart failure, phalangeal tufts resorption
20	dilatation with distal narrowing of esophagus, hiatal hernia, normal chest and hands

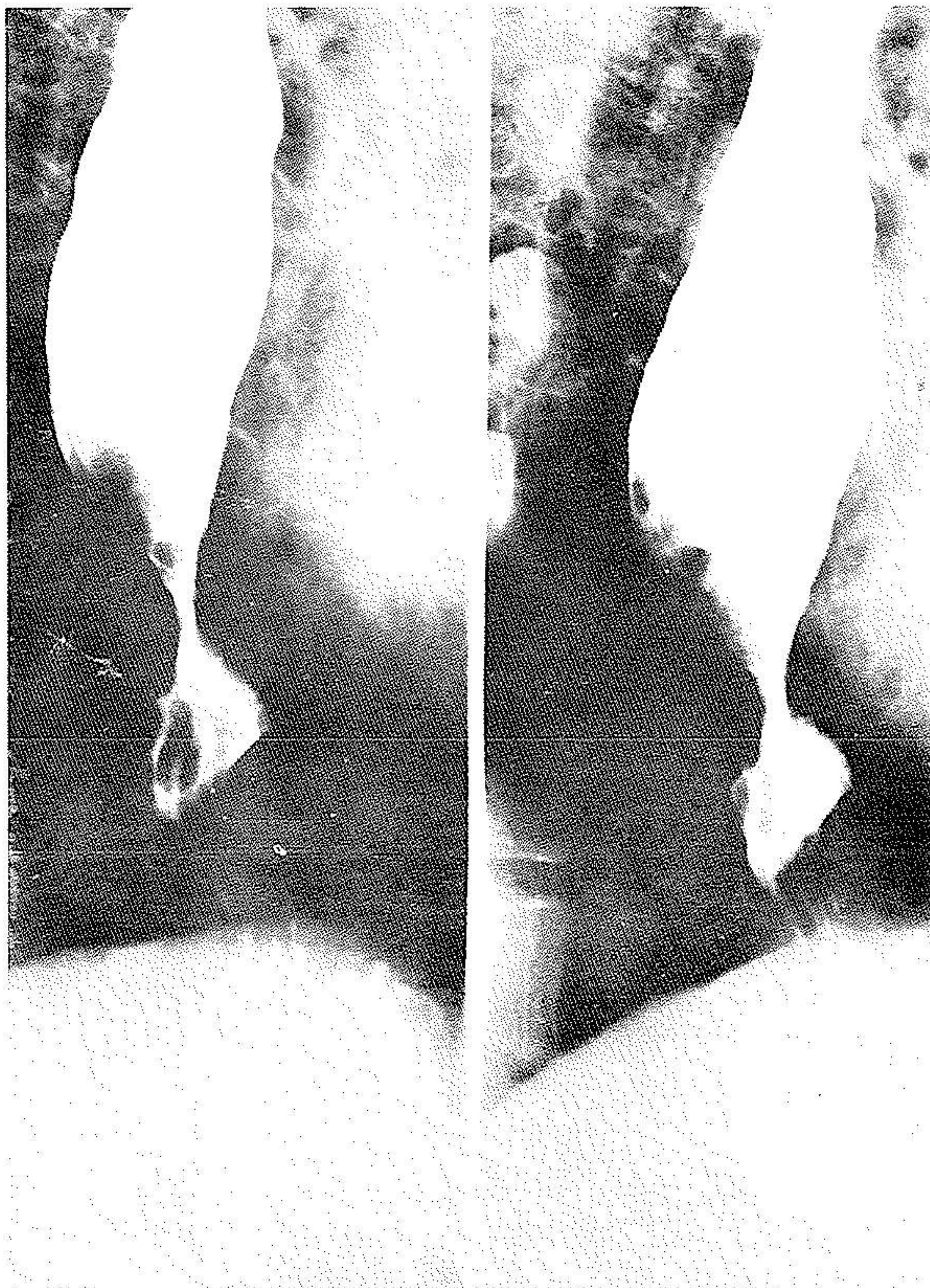


Fig. 1

a web in cricopharyngeal region(fig 2,3).Blood count and hematocrit were within normal limits. Skin biopsy showed evidence of scleroderma.Narrowing of distal portion of the esophagus (4 cases) was similar to that described in other series, but stricture of other portions of the esophagus to our knowledge has not been reported in English literature previously.

Stomach:- Stomach was studied in 13 patients. There were only 2 cases of delayed emptying of stomach.

Small Bowel:- Only 10 patients had roentgenological examinations of small bowel. Observed abnormality in these cases were duodenal dilation (Case 3)(fig 4),coarsening of mucosal folds (case3,12) and sacculatation of a segment of jejunum with barium retention(case3)(fig5). Pseudodiverticula or sacculatation of small intestine is a rare manifestation in scleroderma.

Large Bowel:-We had not any large bowel abnormality in our cases.

Chest:- There were 5 cases of lung fibrosis. Similar to other series fibrosis was more prominent in the lower zones, but in one of the patients in contrast to other reported cases, fibrosis with cystic shadow was evident in the upper zones of the lung (fig6),this patient had symptoms of 8 years duration,and all tests for tuberculosis were negative.

horizontal position is preferred for barium study) (I6). The barium will remain in the esophagus for hours when the patient is in recumbent position, but in erect position the bolus of barium will pass by gravity without delay and this helps in differentiation from achalasia.

The atrophy and atony of the esophagus which result in aperistalsis also lead to dilation which may be manifested on plain roentgenogram of chest as an air esophagogram. The association of air esophagogram without air fluid level, normal fundus and lung changes are pathognomonic of diffuse systemic sclerosis.

As the disease progresses, reflux esophagitis leads to shortening of the esophagus, hiatal hernia and stricture (4,6,10,15). Holmgren (5) reported a case of distal esophagus stricture and hiatal hernia without any inflammatory reaction on biopsy.

One of our patients (case I) as mentioned, had narrowing of middle portion of the esophagus without hiatal hernia, so, we think other than reflux esophagitis, spasm and fibrosis are important factors in esophageal stricture.

There are also reports of diffuse spasm of esophagus in early period of disease (I2).

**Stomach and Bulb of Duodenum:**



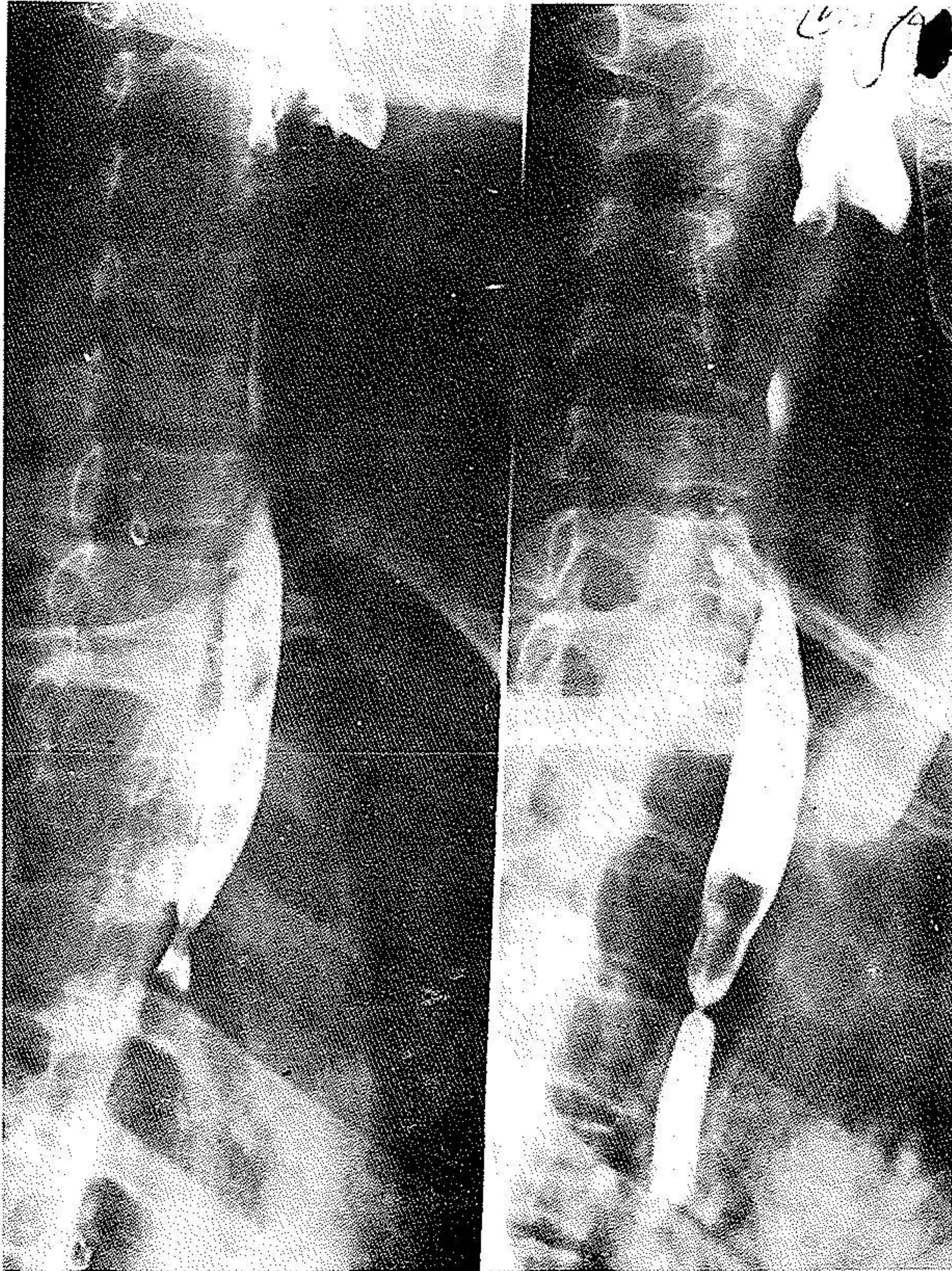


Fig. 2

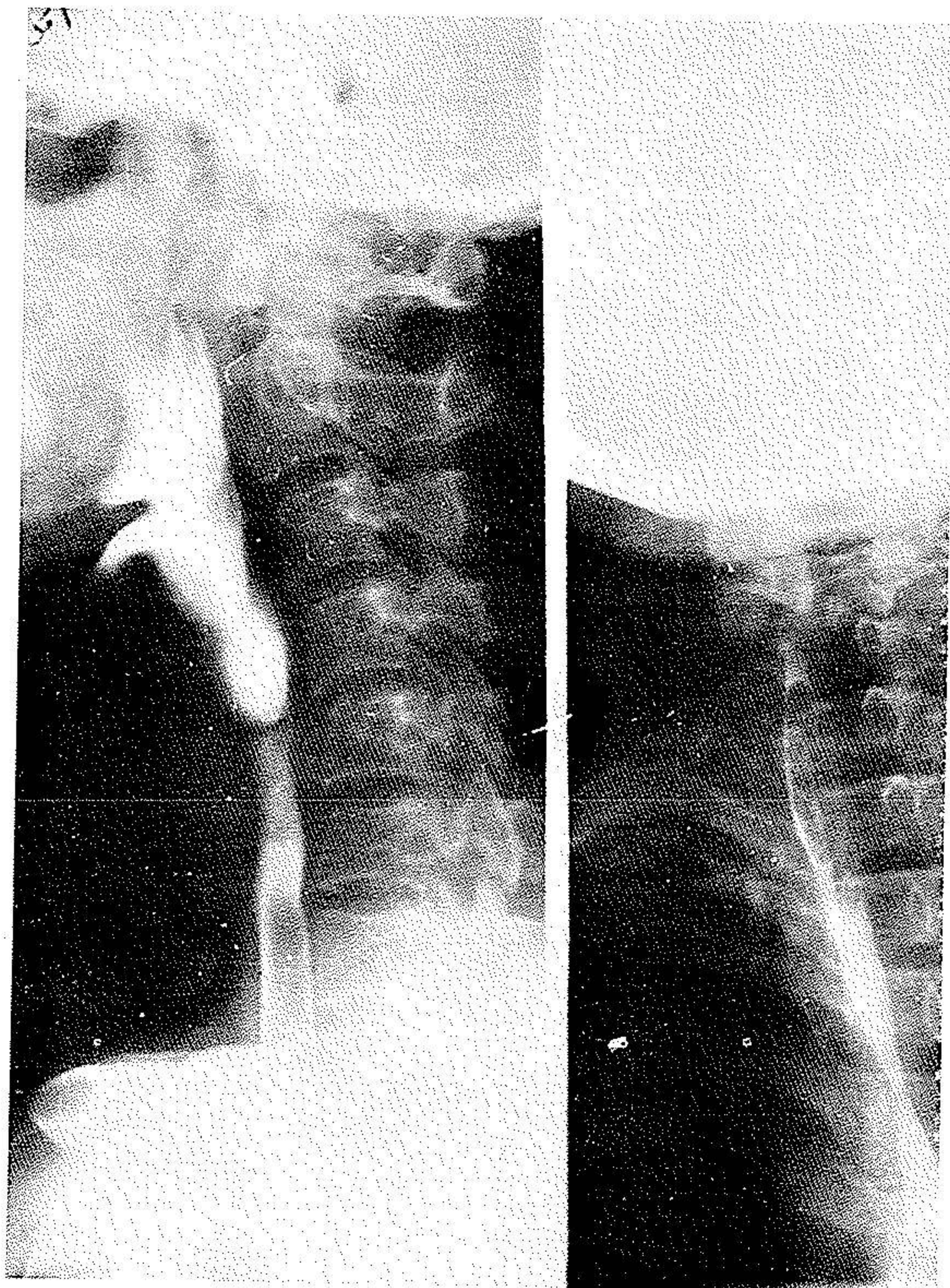


Fig. 3

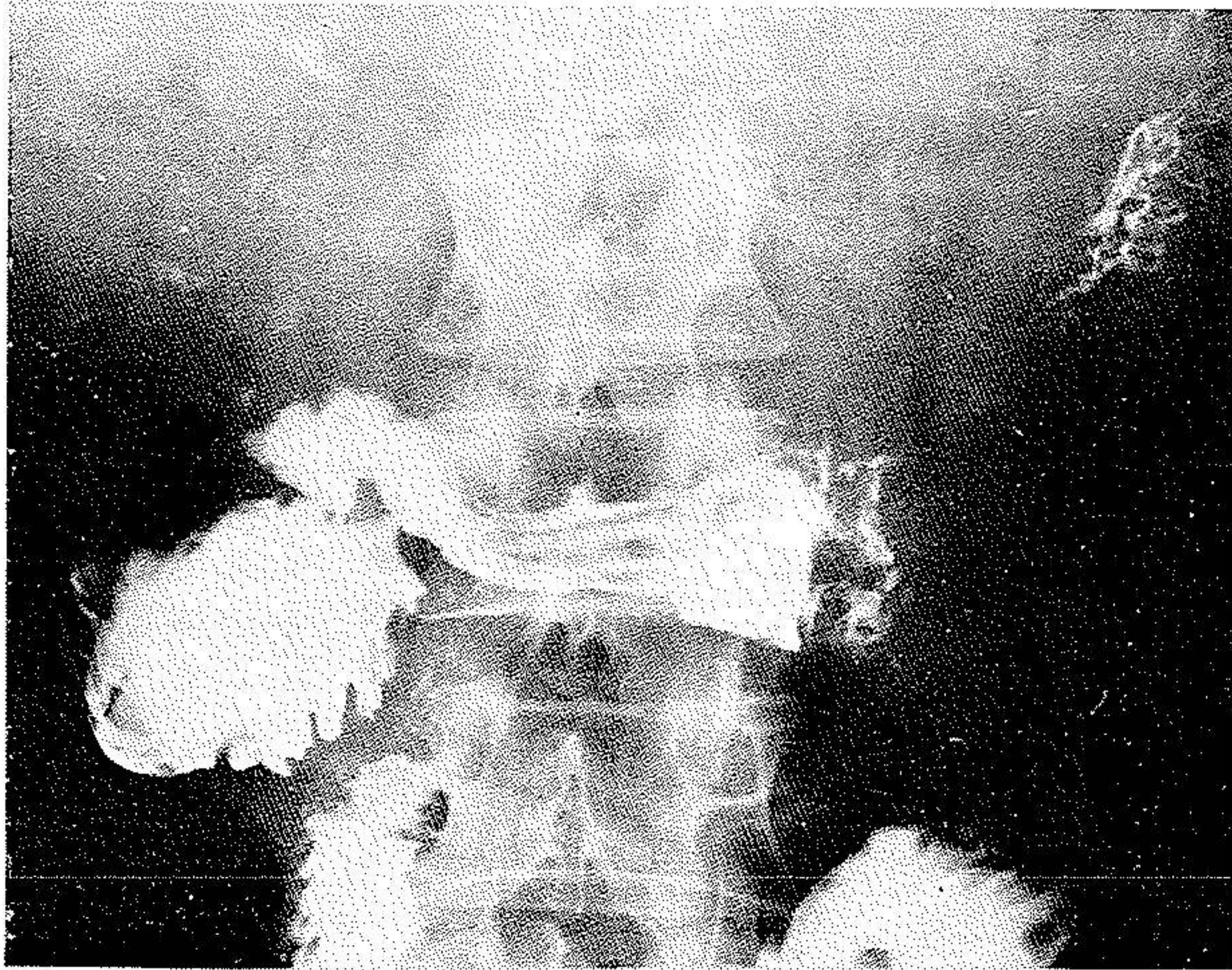


Fig. 4



Fig. 5

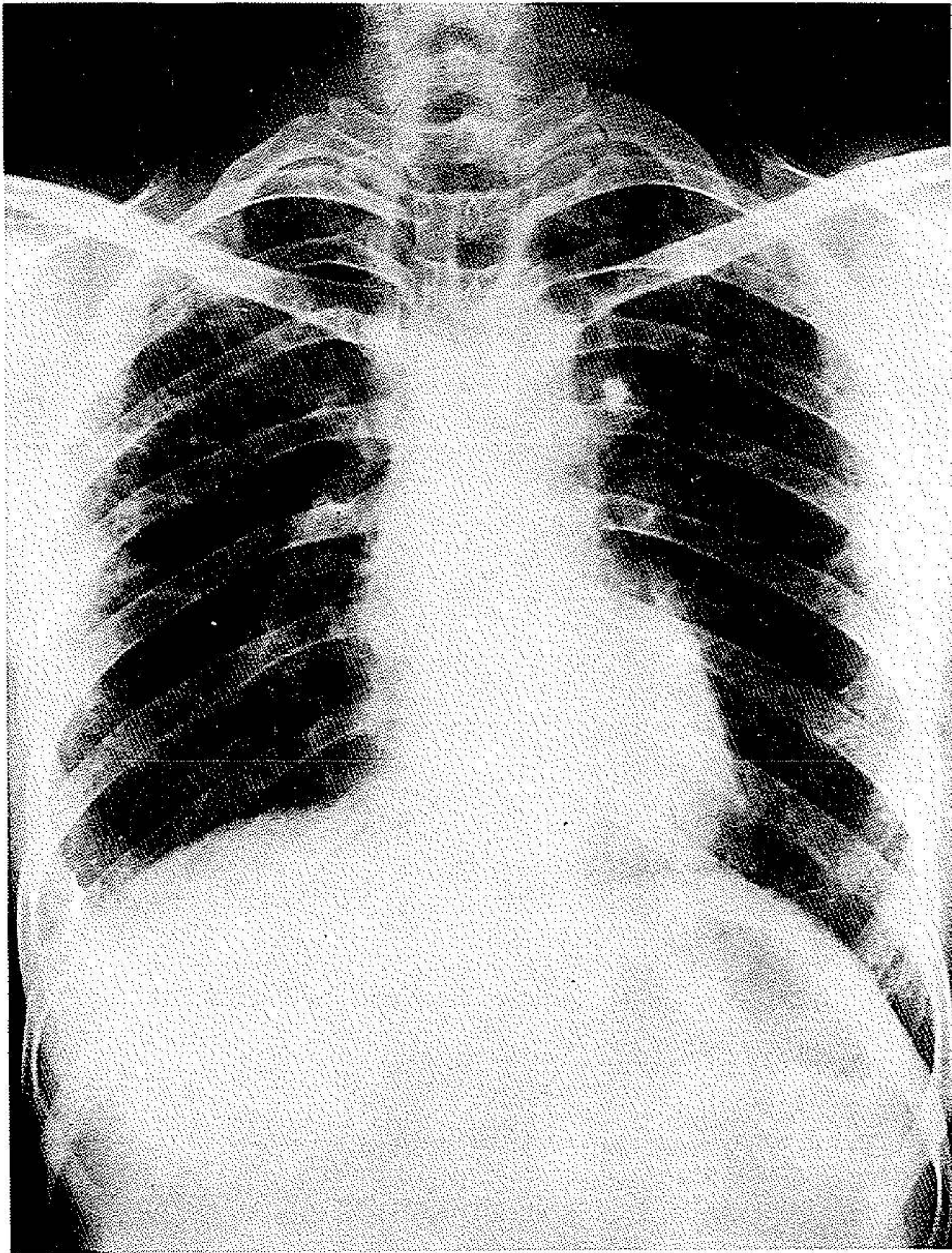


Fig. 6

One patient had cardiomegaly and other signs of congestive heart failure.

Soft Tissue:- There was calcification of finger tips in one patient(fig7).

Bone and Joints:-The most common finding was resorption of terminal phalanges (II cases) (fig8), with osteoporosis (9 cases). There was one case of styloid resorption(fig9).

Teeth:- There was only one case of slightly widened periodontal membrane(fig 10).

Urinary tract had not been evaluated in this series.

#### DISCUSSION

##### Gastrointestinal Tract:

The gastrointestinal tract is one of the most common sites of involvement. Poirier and Rankin reviewed 364 cases of scleroderma in which 44 per cent of the males and 51 per cent of the Females had gastrointestinal symptoms (18).

Esophagus: The esophagus is affected more frequently than other portions of the gastrointestinal tract. In most series the incidence of involvement is 42-75 per cent.

Atrophy and fibrosis of smooth muscle cause impairment of peristalsis and dilation, especially in the distal two thirds of the esophagus (in many cases the motility disturbance is not evident in the erect position, so the



Fig. 7

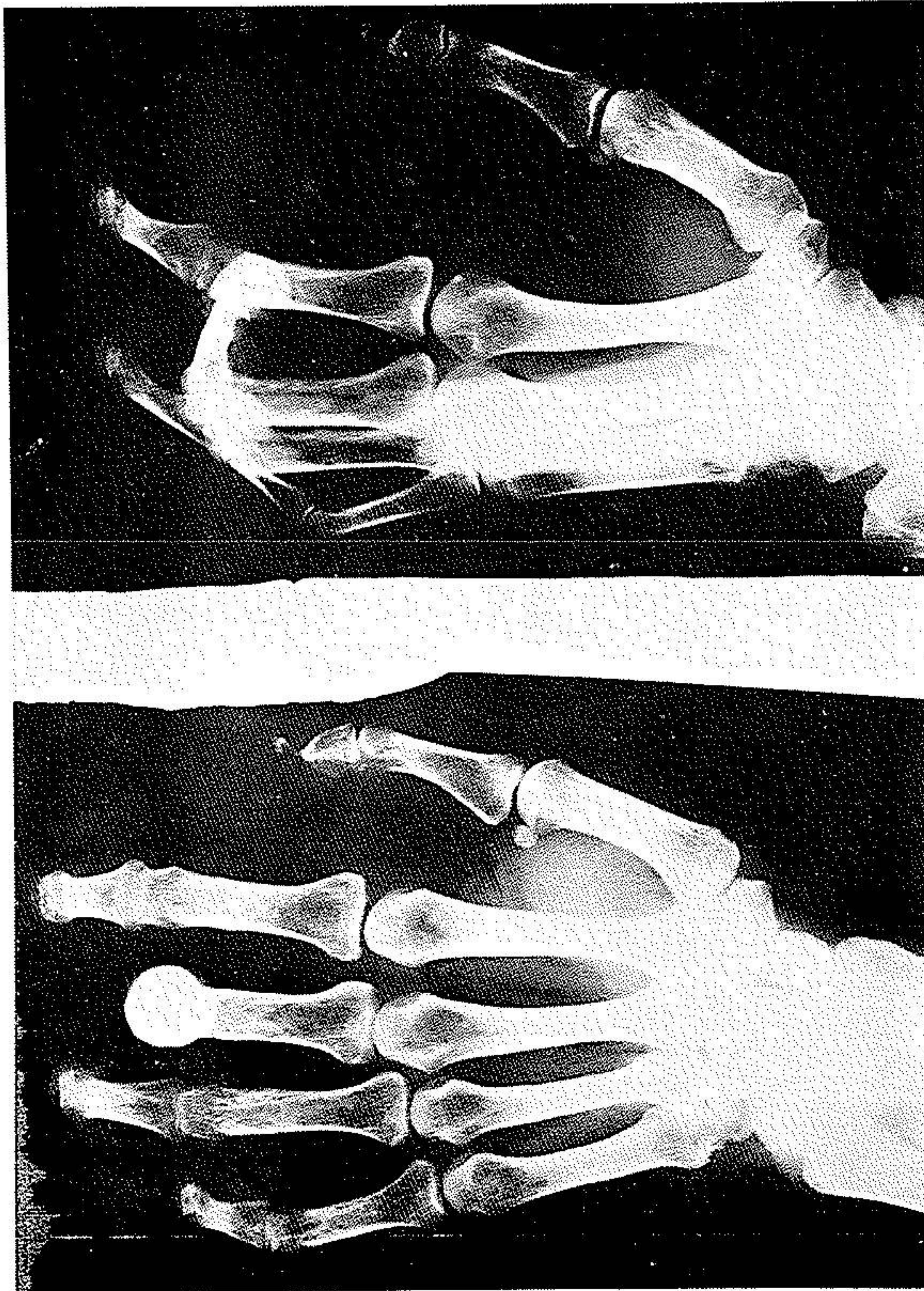


Fig. 8



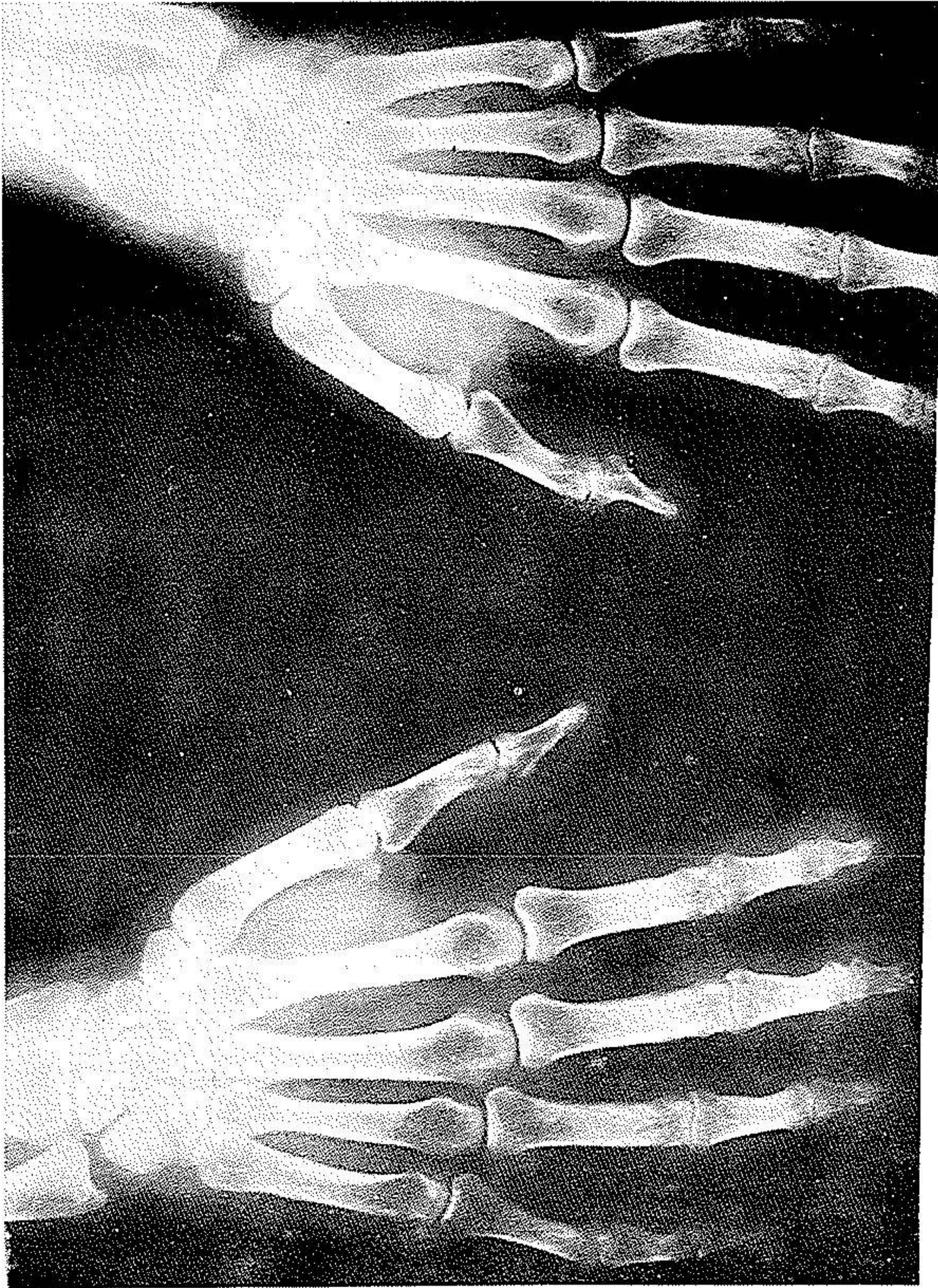


Fig. 9

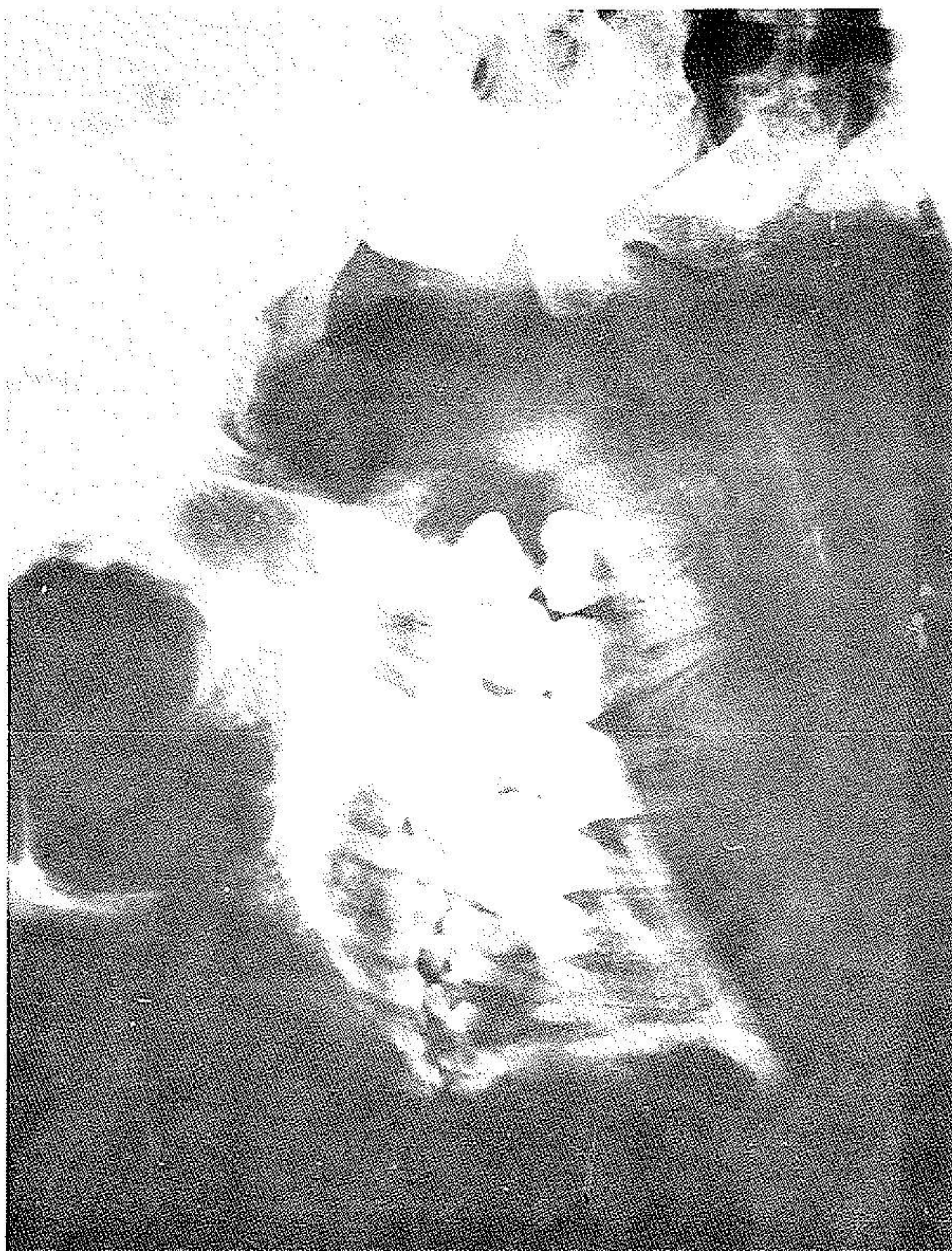


Fig. 10

The stomach and bulb of duodenum usually are involved in terminal phases. Kemp Harper reported degrees of dilatation in stomach in 18 per cent of his patients. In some cases the duodenal bulb is dilated, and barium is retained as long as 24 hours after ingestion. Incidence of duodenal ulcer during the course of disease is 4.2 per cent (10,19,22).

#### Small Intestine:

Small intestine is usually involved in later stage, and the incidence of involvement in various reports are 25 per cent. The lesion begins from duodenum and jejunum. Dilatation of the duodenal loop has been observed by many authors, this is similar to superior mesenteric syndrome, but can be differentiated roentgenographically from that, because, in scleroderma the duodenal loop is hypomotile and unaltered in appearance by placing the patient in prone position, but in superior mesenteric artery syndrome the loop is hypermotile and empties in the prone position (3,7,11,13,21).

Dilatation of small bowel may be suggested in some cases as paralytic ileus or even organic obstruction. In the usual cases of dilatation the mucosal folds tend to be separated, this is true in mechanical obstruction, paralytic ileus, or sprue. But scleroderma produces tightly packed folds" accordion like" in spite of dilation.

Horowitz found this sign in over 60 per cent of the cases, therefore it is a valuable sign in diagnosis of scleroderma when small bowel involvement occurs prior to other signs. Explanation for this event is replacement of muscularis propria by fibrous tissue (8).

Peristalsis is diminished, and barium may remain in the small bowel for many hours. The effect of gravity aid in the progress of barium, therefore it is preferable that the patient remains recumbant during the study. Segmentation of the barium column is occasionally observed, but flocculation is usually not present (12,13,15). Horowitz reported cases of intussusception of the small bowel in scleroderma.

Other less frequent complications in scleroderma are, pneumatosis cystoid intestinalis, and sacculation with pseudodiverticula formation (1,8,13).

#### Large Bowel:

The roentgenological alteration of large bowel is more specific and help in definitive diagnosis, but is rare. Pseudodiverticula (sacculation) is the most typical change. they are most frequently in the transverse and descending colon and more obviously seen in postevacuation films. Sometimes there are alternating areas of narrowing between sacculations. In rare cases there is a huge dilation of colon resembling hirshprung disea-

se (10,12,13,16). other less common findings in P.S.S. are loss of colonic haustration and slight to moderate generalized dilatation. In a recent report, an increase in colonic length was felt to be characteristic of scleroderma (14).

#### Soft Tissue and Bony Changes:

Soft tissue calcification and terminal phalangeal absorption in scleroderma are classic. Either manifestation may predominate or occur alone. Absorptions of the distal phalanges are more common than soft tissue calcification (80 per cent), it usually begins distally and progresses proximally. In severe cases absorption with a resulting pointedness may be observed in the middle and proximal phalanges. Sometimes destruction may occur primarily in the middle third of the phalanx, resulting in separation of distal and proximal portions of the bone. Erosion occurs at first on the palmar aspect of the tips of the terminal phalanges, thus it is important to have lateral view of fingers to detect early resorption (4,6,10,12,16, 23). Rarely absorption of the distal end of the radius and ulna may occur. There are some reports about the absorption of the distal end of acromion and clavicle also (4). Keat reported a patient with classic scleroderma who demonstrated erosion of the superior aspects of the pos-

terior ribs, and indicated that this sign is not specific for rheumatoid arthritis as suspected earlier, but rather a manifestation of collagen disease (9).

As a result of loss of the mobility secondary osteoporosis may occur in the hands (10).

Joint narrowing and ankylosis of hand similar to rheumatoid arthritis have been reported but are rare (17).

Bjersand (2) described one case which in addition to usual boney changes of scleroderma, had marked periosteal new bone formation at the diaphyses of both humeri and femora, which may be the result of peripheral arterial insufficiency as in polyarteritis nodosa. Edeiken (1929) reported intraosseous calcification in scleroderma, this also may be the result of vascular insufficiency (10).

Soft tissue calcification is not as common as bone resorption (58 per cent), and occurs usually many years after the onset of disease, the more common sites of involvement are extremities which are most often subjected to pressure especially the hands, periarticular soft tissue, trunk and even face (10,16,22).

#### Lungs:

The incidence of lung involvement in Kemp Harper series was 22 per cent (10), There are various manifestations in the chest X-ray:

In the early stages there is a fine reticular pattern, as the disease progresses the reticulation tends to become coarser and possess a reticulonodular pattern. There is a tendency for predominant involvement of the lower lung zones, with small cystic lesions as a honeycomb shadow. Seldom, and in severe cases, middle and upper zones of lung may be affected (4,6,10,16).

- Sometimes there is a large pneumatocele.
- pneumothorax as a result of cyst rupture.
- Recurrent pneumonia.
- Pulmonary hypertension.
- Pleural involvement in contrast to other collagen disease is rare.
- Calcification is very uncommon.
- The association of scleroderma and carcinoma is well recognized. Most of reported cases were bronchioalveolar carcinoma and some cases of bronchogenic carcinoma (4, 10,21).

#### Heart:

The reasons of cardiomegaly in scleroderma are:

- Myocardial fibrosis.-Pericardial effusion - Right ventricular enlargement secondary to pulmonary hypertention (10).

#### Teeth:

Stafne and Austin (1944) first described widening of periodontal membrane in 7 per cent with scleroderma. The pre-molar and molar were affected more commonly than the canine and incisors. This widening is usually uniform (3,10,16,20).

#### SUMMARY:

Radiological findings of 20 patients with scleroderma have been described. We had common and uncommon roentgen signs of scleroderma similar to other series and one case with mid esophagus narrowing which we think have not been reported previously.

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