REFERENCES

1 Daicoff GR, Rhodes ML: Surgical repair of ventricular septal rupture and ventricular aneurysm. JAMA 203:115, 1968


Bone Resorption of the Ribs and Pulmonary Function in Progressive Systemic Sclerosis*

James C. Steigerwald, M.D.;** Martin H. Seifert, M.B.;†
May M. Cliff, M.D.;‡ and Thomas A. Neff, M.D., F.C.C.P.§

We report a case of progressive systemic sclerosis which showed the unusual feature of bone resorption of the ribs associated with pulmonary function abnormalities. This case, to our knowledge, is the first reported in which this bone resorption of the ribs has occurred.

Progressive systemic sclerosis (PSS) is a multisystem disease which frequently causes pulmonary function and chest roentgenogram abnormalities.1 Bone resorption is another common finding in PSS, with the distal phalanges, radius, ulna, clavicle, and jaws most often involved.2,3 We have recently observed a young woman with PSS who, during the course of her disease, developed marked bone resorption of the ribs along with pulmonary function abnormalities.

CASE REPORT

A 25-year-old black woman was first seen at the University of Colorado Medical Center in May 1966 with the chief complaint of increasing tightness of the skin. She had been entirely well until March of 1965 when she first noted arthralgia and arthritis involving the metacarpal phalangeal, and proximal interphalangeal joints of both hands. The results of evaluation at that time had been normal, except for a

*From the Department of Internal Medicine, University of Colorado Medical Center and Denver General Hospital, Denver.

**Assistant Professor of Medicine, Division of Rheumatic Diseases.

† Former Fellow, Division of Rheumatic Diseases.

‡ Associate Professor of Radiology.

§ Associate Professor of Medicine, Division of Pulmonary Disease.

Reprint requests: Dr. Steigerwald, Denver General Hospital, Denver 80204

Figure 2. Left ventriculogram in left anterior oblique position demonstrating ventricular septal defect (arrow) and filling of right ventricle (RV). S, Intraventricular septum; LV, left ventricle.

Figure 3. Ventricular septal defect (arrow).
Abnormalities of the chest roentgenogram are common in PSS, with diffuse mottling in the bases of the lungs representing the earliest change in the lung parenchyma. From this change, there may be progression to diffuse pulmonary fibrosis, most marked at the bases. Abnormalities in pulmonary function most often antedate the onset of roentgenologic changes, with an impaired Dss the earliest change, as was seen in our patient. As the pulmonary fibrosis progresses, the VC decreases, and an obstructive defect may also develop. In patients who have abnormalities of the lung parenchyma on roentgenogram, there are significant measurable decreases in lung function.

Heretofore, however, there have not been, to our knowledge, any reports of bone resorption of the ribs in PSS. This severe deformity and disability made it impossible to do detailed pulmonary function studies in the last three years of the patient's life. We did, however,
measure her total lung capacity (TLC) using a planimetric method as described by Harris et al, which demonstrated a loss in TLC from 4.4 L to 2.6 L in the period from 1966 to 1972. During this period, it is of interest that the appearance of the lung parenchyma on the chest roentgenogram remained normal. Arterial blood gas determinations done at rest and mild to moderate exercise in 1966 and 1970 gave normal results. These studies strongly suggest that the patient's abnormal pulmonary-function test results were due to musculoskeletal chest deformities rather than the more typical parenchymal fibrosis.

In summary, this unusual manifestation of a decrease in TLC and VC associated with marked bone resorption of the ribs seen on chest roentgenogram must be added to the possible pulmonary complications of PSS.

REFERENCES


Airway Obstruction due to Spontaneous Retropharyngeal Hemorrhage*

Michael G. Genovesi, M.D.** and Daniel H. Simmons, M.D., F.C.C.P.†

A patient is described with polycythemia vera who was taking anticoagulants and developed a spontaneous retropharyngeal hemorrhage after a violent sneezing episode. This progressed to tracheal compression with stridor after he had taken some aspirin for relief of the neck pain. An emergency tracheostomy was life saving.

*From the Department of Medicine, UCLA School of Medicine, Los Angeles.
Supported by U.S. Public Health Service Training Grant No. HL 05917.
**Fellow in Pulmonary Disease.
†Professor of Medicine and Physiology; Chief, Division of Pulmonary Disease.
Reprint requests: Dr. Simmons, Department of Medicine, UCLA School of Medicine, Los Angeles 90024

Figure 3. Chest x-ray film showing fractures and dislocations of all ribs with pencilling of distal end of clavicle on right (November 4, 1970).

Figure 4. Chest x-ray film demonstrating further destruction of bone in ribs with complete resorption of distal end of clavicle on right (July 4, 1972).

840 GENOVESI, SIMMONS

CHEST, 68: 6, DECEMBER, 1975