Systemic bone disease in bony thorax

Kiyoharu Nishioka, M.D. and Kyoichi Hiramatsu, M.D.
Department of Diagnostic Radiology, School of Medicine, Keio University, Tokyo

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胸部骨格からみた系統的骨疾患

慶応大学放射線診断部（主任：西岡清春教授）
西岡清春　平松京一

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胸部骨格は各種の骨，すなわち，長管骨，短骨，扁平骨，特殊骨（鎖骨）から成り，そのため系統的骨疾患には何等かの異常所見を胸部X線像に示す．

本論文では spondyloepiphyseal dysplasia, multiple epiphyseal dysplasia, achondroplasia, lipochondrodystrophia, hypophosphatasia, osteopetrosis, multipleexostosis, Osteogenesis imperfecta, dysostosis cleidocranialis, infantile cortical hyperostosi hyperchondroplasia, metaphyseal dysostosis, craniometaphyseal dysplasia, progressive diaphyseal dysplasia, hyperostosis corticalis generalisata,hyperostosis generalisata with pachydermia, melorheostosis, osteopoikilosis, osteopathia striata について比較的 pathognomonic の所見を検討し報告する．

In chest X-ray diagnosis, examination of the pleura, mediastinum, diaphragm, and thoracic wall is important in addition to the lung and heart. Among diseases giving rise to abnormalities in bony thorax, those summarized in Table 1 are known besides so-called bone diseases. Bony thorax consists of various kinds of bones such as long tubular bone (humerus), short-bone (vertebra), flat bone (scapula, ribs and sternum), and a special bone (clavicle). In systemic bone disease, therefore, some abnormalities are usually noted in chest X-ray picture. Conveniently, bony thorax is clearly visualized in routine chest X-ray film, except for the vertebra and sternum. Consequently, chest X-ray films taken for other

Table 1. Diseases with pathological roentgen findings of the bony thorax

1. Anomalous, Developmental defects
2. Systemic bone diseases
3. Endocrine disturbances
4. Metabolic disturbances
5. Collagen diseases
6. Haemopoetic diseases
7. Systemic bone neoplasms
8. Histiocytoses
9. Heart a. Vasculary diseases
10. Others (Nervous dis., Infectious dis., Nephropathies, Paget's dis., Gout, Sarcoidosis, etc.)

Table 2. Systemic bone diseases with pathological roentgen findings of the bony thorax

1. Spondyloepiphyseal dysplasia (Morquio)
2. Multiple epiphyseal dysplasia
3. Achondroplasia, Hypochondroplasia
4. Lipochondrodystrophia (Gargoilism)
5. Hypophosphatasia
6. Osteopetrosis (Marble bone)
7. Multiple exostosis
8. Osteogenesis imperfecta
9. Dysostosis cleidocranialis
10. Infantile cortical hyperostosis
11. Hyperchondroplasia (Marfan)
12. Metaphyseal dysostosis
13. Craniometaphyseal dysplasia (Pyle)
14. Progressive diaphyseal dysplasia (Engelmann)
15. Hyperostosis corticalis generalisata (Van Buchem)
16. Hyperostosis generalisata with pachydermia (Uehlinger)
17. Melorheostosis
18. Osteopoikilosis
19. Osteopathia striata

purpose can be used for the diagnosis.

Chest X-ray picture in systemic bone disease

Table 2 summarized the systemic bone diseases giving rise to abnormalities in chest X-ray film. In the X-ray diagnosis of these diseases, the characteristics of each disease should be clarified at first. As the fundamental items, on the other hand, 1) bone structure, 2) bone age, 3) stage of disease 4) transitional form between diseases 5) changes according to age, should be considered.

The abnormalities mainly in the bony thorax are described in the order of Table 2.

1. Spondyloepiphyseal dysplasia (Morquio)

Ribs assume horizontal positions, occasionally showing curvature with upward convexity. From the posterior to the anterior direction, the width of the rib increases. Intercostal space is narrow, with bone dysplasia at the epiphysis and deformity (Fig. 1).

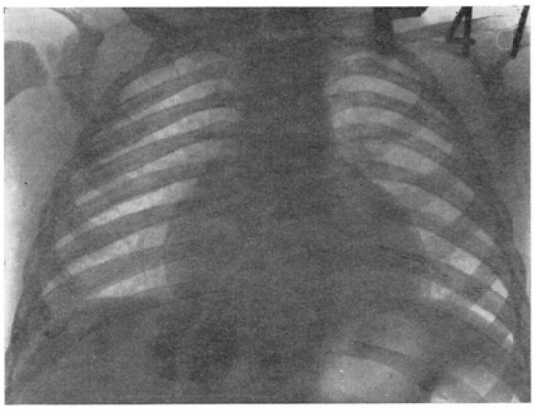

Fig. 1. Spondyloepiphyseal dysplasia (Morguio).

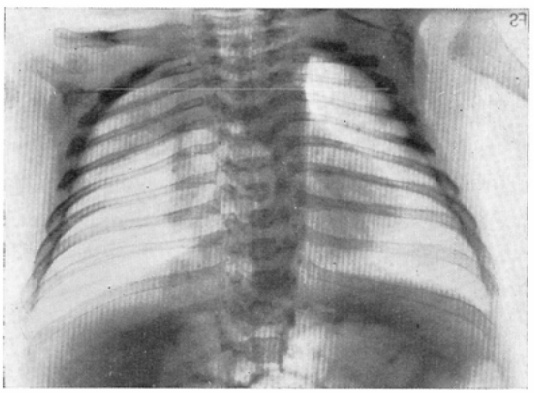

Fig. 2. Multiple epiphyseal dysplasia.

2. Multiple epiphyseal dysplasia

Multiple punctate bone nuclei are found at the sites corresponding to epiphyseal nuclei of the vertebra, rib, sternum, and scapula (Fig. 2).

3. Achondroplasia, hypochondroplasia

Long bones are thick and short, with indentations of the epiphyseal line and delay in the appearance of epiphyseal nuclei. In chest X-ray picture, ribs are short and rib cartilage portion shows a dish-like enalargement (Fig. 3).

4. Lipochondrodystrophia, gargoilism

This is one of the mucopolysaccharidosis simultating morquio disease. In chest X-ray film, narrowing of the intercostal space accompanying vertebral changes is noted. Ribs run rather horizontally with increasing width towards the anterior direction giving an oar-like appearance (Fig. 4).

5. Hypophosphatasia

This is a disease causing bone dysplasia of the primary spongiosa. When it is intense, only the trace of thorax is noted.

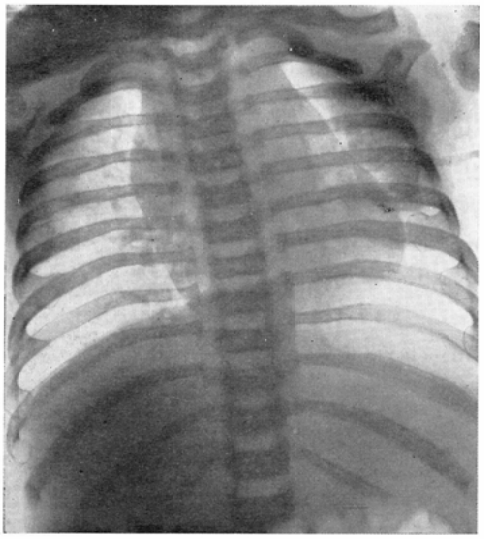

Fig. 3. Achondroplasia.

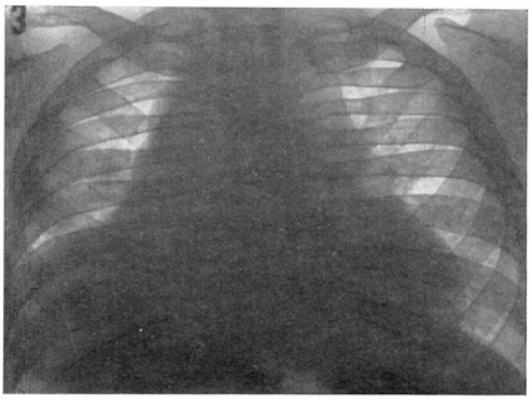

Fig. 4. Lipochondrodystrophia (Gargoilism).

6. Osteopetrosis, Marble bone

This disease is characterized by generalized osteosclerosis, through deposition of calcium in the primary spongiosa and not by osteosclerosis of the secondary spongiosa. Ribs, scapula, sternum and clavicle show homogeneous osteosclerosis, with trace of fracture and deformity. Vertebra shows sandwitch-like apearance (Fig. 5).

7. Multiple exostoses

The frequent sites of occurrence of exostoses are knees, feet, arms, and metaphysis of shoulder joint. In chest X-ray, there are frequently seen in the scapula and ribs, and rarely in the vertebra and clavicle, presenting as hill-like protrusion with broad base (Fig. 6).

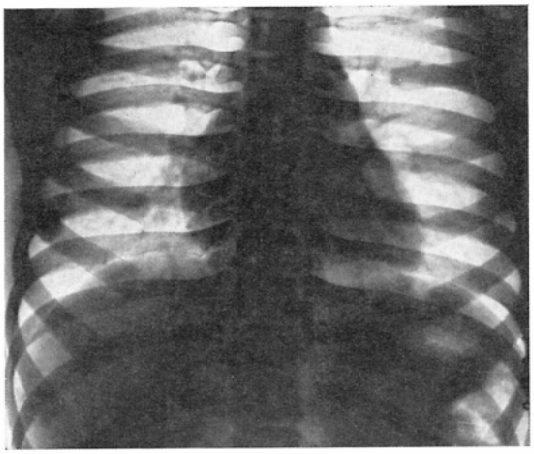

Fig. 5. Osteopetrosis, marble bone.

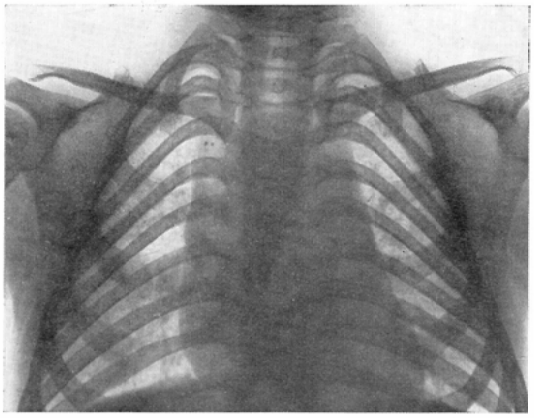

Fig. 6. Multiple exostosis.

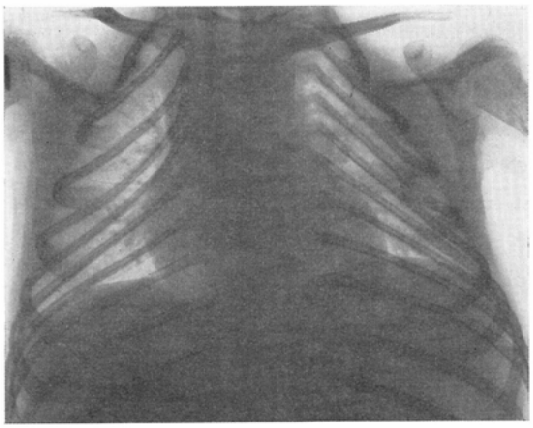

Fig. 7. Osteogenesis imperfecta.

8. Osteogenesis imperfecta

The length and width of the bone decrease, causing bone atrophy. Fracture and bending frequently occur. In the chest X-ray film, the running of the ribs becomes steep, width of the rib extremely narrowed, with fracture and deformity (Fig. 7).

9. Dysostosis cleidocranialis, dysostosis generalisata

In this disease, dysostosis occurs in the clavicle, skull, vertebra, public bone and proximal end of the femur along with all the bones. In chest X-ray film, bilateral splitting or defect of clavicle is generally characteristic. The defect is on the acromion side and the sternal side remains. Dysplasia is also seen in the ribs, frequently accompanied by scoliosis and anomalous rib (Fig. 8).

10. Infantile cortical hyperostosis

In this disease, periosteal ossifications of various forms are noted. In chest X-ray, lamellar ossification is seen apart from and outside of the pre-existing bone cortex in the ribs, clavicle, and scapula. An onion-peal appearance is occasionally noted.

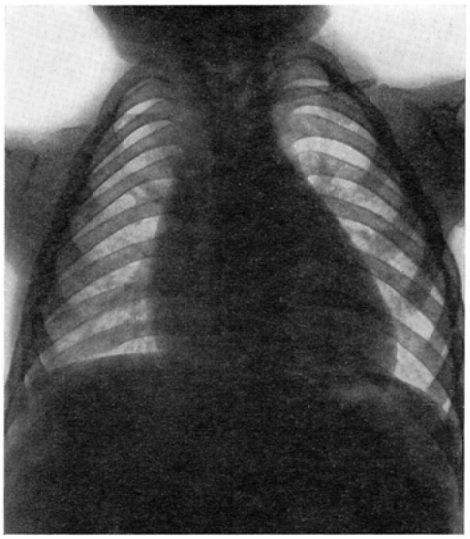

Fig. 8. Dysostosis cleidocranialis.

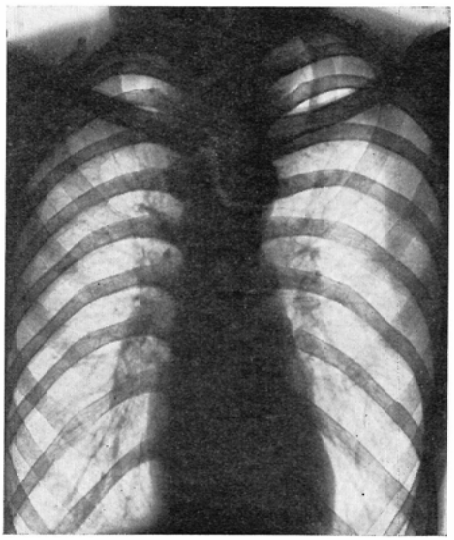

Fig. 9. Hyperchondroplasia (Marfan).

11. Hyperchondroplasia, marfan's syndrome

While the abnormality of the bone substance is absent, the length of the bone throughout the body abnormally increases. In chest X-ray film, ribs become extremely long and the thorax expands superiorly and inferiorly. The sternum is depressed, giving a funnel chest appearance (Fig. 9).

12. Metaphyseal dysostosis

Dysostosis of the metaphysis of all the bones is noted. Deformity and sclerosis of the ribs, clavicle and scapula and narrowing of the thorax due to short ribs are noted.

13. Craniometaphyseal dysplasia (Pyle)

The metaphysis of the long bones is dilated, cortex is thin and translucent. On the contrary, diaphysis becomes sclerotic Due to the disturbance in modeling, the clavicle becomes triangle in shape and osteosclerotic along with the ribs. Intercostal spaces are narrow.

14. Progressive diaphyseal dysplasia

Thickening and deformity of the clavicle and ribs are noted.

15. Hyperostosis corticalis generalisata (Van Buchem)

Osteosclerosis of the ribs is noted especially in the upper edge.

16. Hyperostosis generalisata with pachydermia (Uehlinger)

Thickening due to periosteal ossification and osteosclerosis are noted in the ribs scapula, resembling findings in Paget' disease.

17. Melorheostosis

From the scapula to the upper extremity, a picture resembling downstream of wax along the bone is noted. Rib involvment is rare.

18. Osteopoikilosis

In the spongiosa of the bone, multiple, small, round or oval osteosclerotic patches are noted. Scapula, ribs and sternum are rarely involved.

19. Osteopathia striata (Voorhoeve)

In the metaphysis of the bone, numerous lesions of linear sclerosis corresponding to the long axis of the bone are noted. In the chest X-ray film, mild changes are occasionally seen in the clavicle and ribs.

Conclusion

Since most of these findings are rather pathognomonic, diagnosis can be made even with the chest X-ray film taken for other purpose. Such concept may be applicable to X-ray pictures of hand, pelvis head and other portion in addition to chest X-ray films, and this is quite important for diagnostic radiologist.

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