

Osteopathia Striata with Cranial Sclerosis:

2

1

2

2

(Osteopathia striata with cranial sclerosis)

10

31

(osteopathia striata, OS)
(metaphysis)

(patent ductus arteriosus, PDA)

(longitudinal striation)가
(1).

(cranial sclerosis)

(osteopathia striata

가

with cranial sclerosis,

OS - CS)

OS

(1, 2). OS

가

(Fig. 1A). OS

OS - CS

(1 - 4). OS - CS

(Fig. 1B).

(macrocrania), (frontal bossing),
(hypertelorism), (nasal ridge),

(iliac crest)

(Fig. 1C).

(anterior fontanelle) 가 10

OS가

(Fig.

(skeletal survey)

1D).

(computed tomography, CT)

가

(Fig. 1E).

OS - CS

(palatine bone)

(vomer)

가

가

(cranial foramina)

1

10

가

37

2.9 kg

가

가

2

31

가

(unerupted tooth)

1

2

2006 10 29

2006 12 18

가

(Fig. 2A). (Fig. 2B), CT (Fig. 2C). (Fig. 2D - F).

가 (isolated form) OS (OS - CS) (1, 2). OS가 OS - CS (autosomal dominant) (X - linked dominant inheritance) (1, 4). 1

가 (polydactyly) (symphalangism), (clinodactyly) (3, 9).

가 (trabeculae) (5 - 7). 1

가 (2, 8). 2

가 (transmission) 2

가 4 6

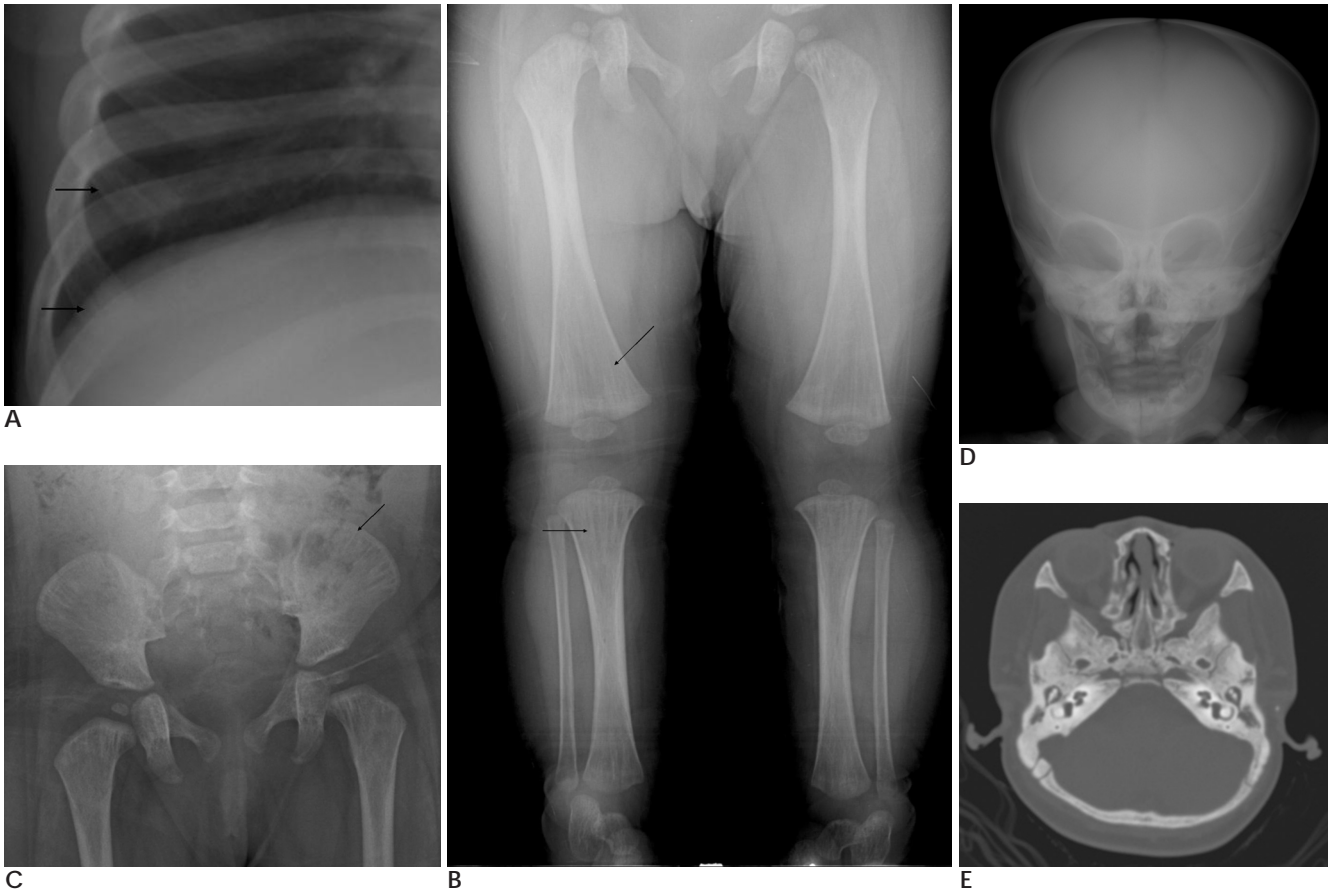


Fig. 1. Patient 1. A 10-month-old girl.

- A. Magnified view of the ribs show sclerotic lines along the axis of ribs(arrows).
- B. Vertically running sclerotic lines are seen at the metaphyseal ends of both femora and tibiae(arrows).
- C. Pelvis shows vertical and fan-shaped sclerotic lines on both iliac crests(arrow).
- D. Skull shows macrocrania and widely opened anterior fontanelle.
Note sclerosis at the orbital roof, nasal bridge, maxilla and mandible.
- E. CT scan with bone setting attenuation shows sclerotic widening of basal skull and vomer of the nasal bone.

(optic foramen) 가
(4, 9),
(hydrocephalus),
(cortical atrophy), (corpus callosum)
(vascular entrapment) (2, agnesis) (2 - 4).
6). 1 OS - CS
(2, 3, 10). 1
(2, 5, 8). 1
(macrognathia) OS - CS
(2, 3, 5, 7). CT

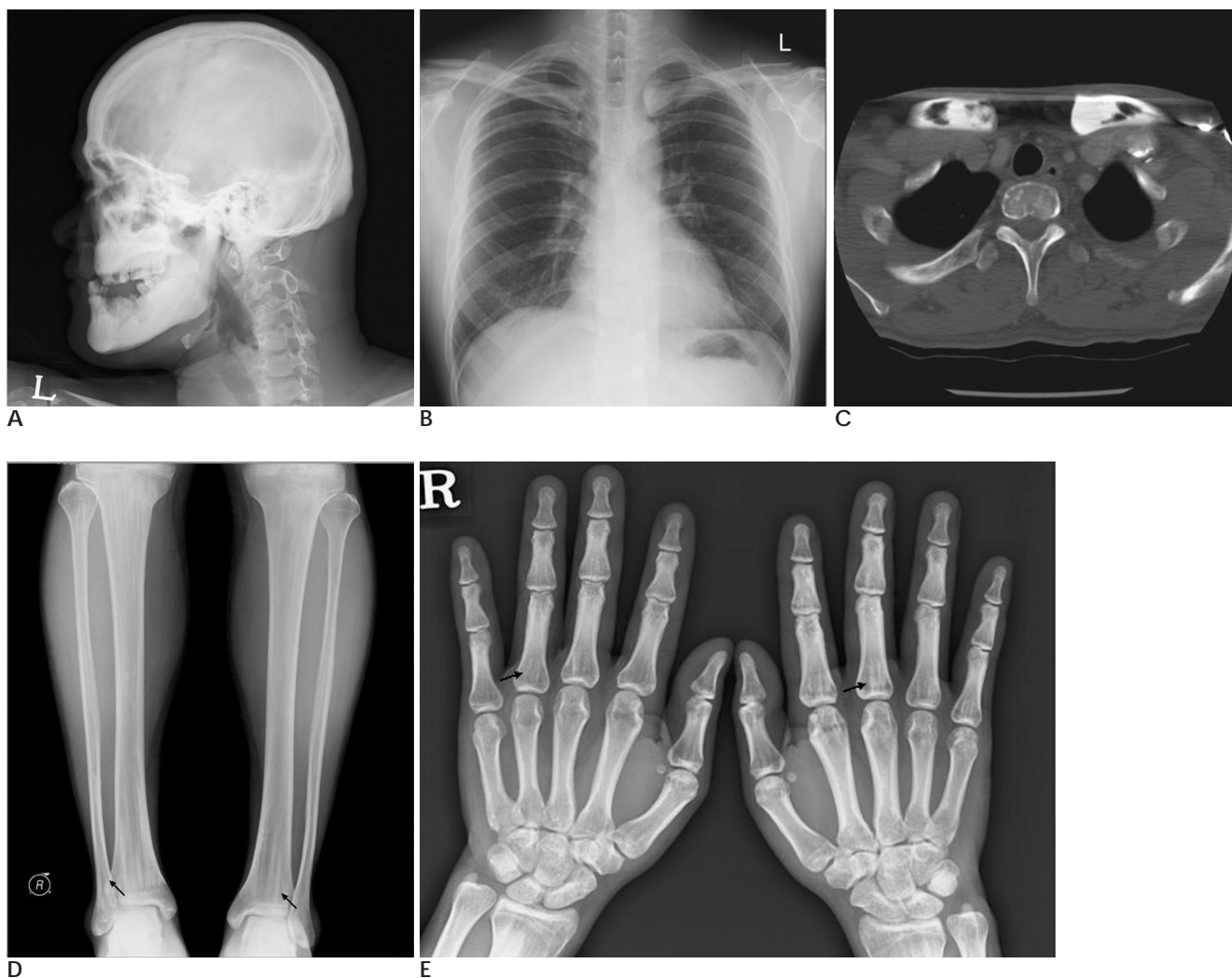


Fig. 2. Patient 2. A 31-year-old male.

A. Skull lateral view shows diffuse sclerosis of the calvaria, particularly at the occipital protuberance. Maxilla and mandible are remarkably thickened and sclerotic.

B. Chest radiograph shows mild sclerosis with widening of medial ends in both clavicles.

C. Chest CT confirms sclerotic widening of the clavicular ends.

D. Note linear sclerotic lines along the metaphyseal ends of both tibiae and fibulae(arrows).

E. Thickened sclerotic lines are seen at both distal radii, ulnae and phalanges(arrows).

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Osteopathia Striata with Cranial Sclerosis: Report of Two Cases¹

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Osteopathia striata with cranial sclerosis is a very rare sclerosing bone dysplasia that is characterized by the radiological appearance of longitudinal striations at the metaphysis of the long bones associated with cranial sclerosis. Here, we report a 10-month-old female infant and a 31-year-old male, who were diagnosed incidentally by showing longitudinal sclerotic lines symmetrically in the long bone ends and basal sclerosis of the cranium. The diagnostic linear striation was found not only in the long bones but also in the ribs and phalanges, which are very rare sites for striation. We discuss the radiology findings of osteopathia striata with the possible complication of cranial sclerosis.

Index words : Bones, dysplasia
Bones, sclerosis
Infants, skeletal system
Bones, abnormalities

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