

가

:

1

(Diffuse esophageal leiomyomatosis)

syndrome)

(Alport

5

가

(diffuse esophageal leiomyomatosis)

(trachea)

(Alport syndrome)

(1 - 3).

5

5 가 2

2

(Fig. 1).
tomography, CT)

(computed

(Fig. 2A, B).

(Fig. 2C, D).
(lobulated)

(Fig. 3).

. CT (Fig. 2A)

(Fig. 3A)

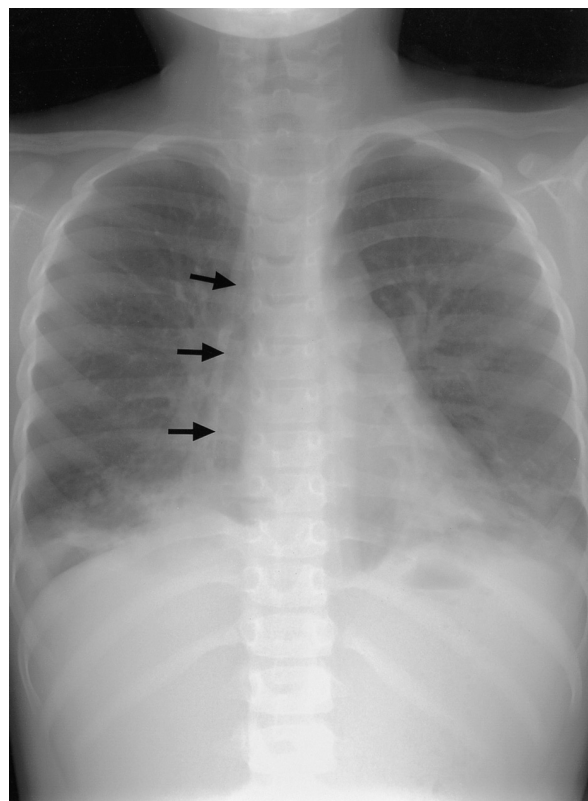


Fig. 1. Plain chest radiograph shows pneumonia in both basal lungs and elongated air density (arrows) along the right mediastinal border due to esophageal dilatation.

: 가

(foot
process) (diffuse effacement)
(glomerular basement membrane)

, CT MRI

(), (ante -
rior lenticonus), (macular flecks)
X

(1, 2).

(diffuse leiomyomatosis)
40 가 (sporadic)

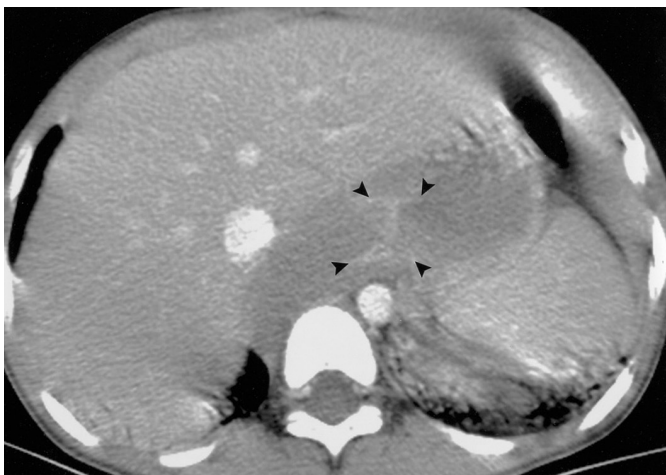
가
(3).



A



B



C



D

Fig. 2. Chest CT scans reveal severe esophageal wall thickening involving the entire esophagus and cardiac portion of the stomach. Esophageal wall thickening is asymmetrically prominent on the left side and most severe at the distal esophagus. The posterior wall of the distal trachea is compressed by the lesion (A, arrowheads) and aspiration pneumonia in both lower lungs is also noted (B). Preservation of mucosal enhancement (C, arrowheads) indicates the submucosal lesion and the involved cardiac portion of the stomach (D, arrows) protrudes into the gastric lumen.

(retrosternal pain) (corkscrew)
 가
 (4, 5), CT
 .
 X
 가 IV 1, 4, 5 (2).
 (2, 6). (coding)
 , COL4A3 COL4A4 2
 (achalasia) , COL4A5
 가 5 cm (head - to - head) COL4A6 X
 CT (2). COL4A5 X
 가 (85%) , COL4A5 COL4A6
 (7 - 가 (deletion) , 가 (2).
 9). (subserosal adhesion)
 가 (9, 10). ,
 (idiopathic muscular hypertrophy of esophagus)
 , (nonneo -
 plastic expansion)
 ,
 (2). 가



Fig. 3. Esophagography shows esophageal dilatation with lobulated margin without mucosal abnormality. The trachea is diffusely compressed by the adjacent esophageal wall thickening (A, arrowheads). Luminal narrowing is most prominent in the distal esophagus (B, arrows) but there is no difficulty in passing the barium into the stomach.

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Diffuse Esophageal Leiomyomatosis in a Child with Alport Syndrome: Case Report¹

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Diffuse esophageal leiomyomatosis is an exceedingly rare, benign, neoplastic condition occurring predominantly in children and young adults. This condition may occur as an isolated finding, or it may be associated with Alport syndrome. We report a case of diffuse esophageal leiomyomatosis with Alport syndrome in a 5-year-old girl who had presented with recurrent pneumonia, and present a review of the literature. We suspected Alport syndrome in the patient because she had a clinical history of congenital cataracts and hematuria, as well as imaging findings of diffuse esophageal leiomyomatosis. Alport syndrome was subsequently confirmed by electron microscopy of the kidney.

Index words : Leiomyomatosis
Alport syndrome
Esophagus, neoplasms

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