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1, 2 가 가 (Fig. 1).

(myositis ossificans progressiva)'
 (fibrodysplasia
 ossificans progressiva)'
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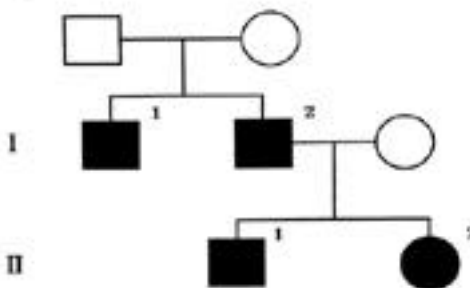


Fig. 1. Pedigree chart

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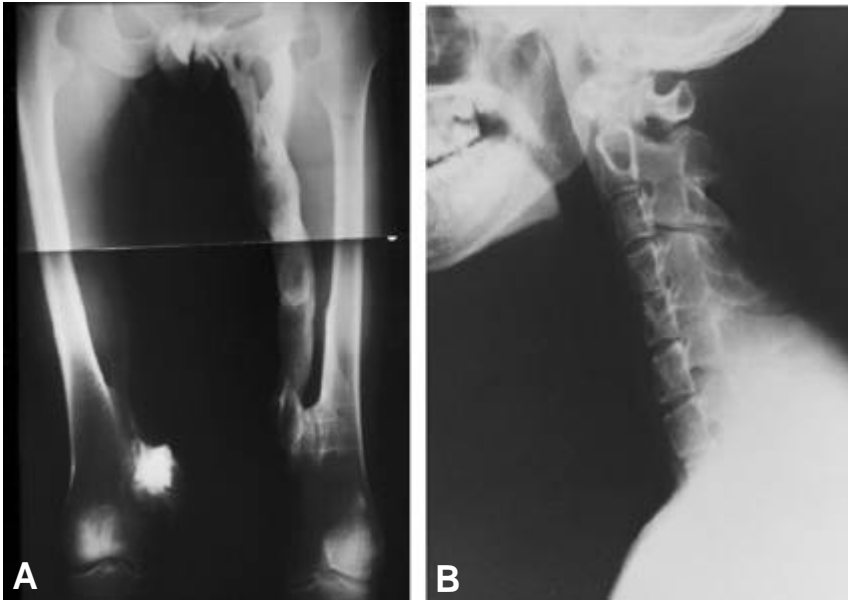
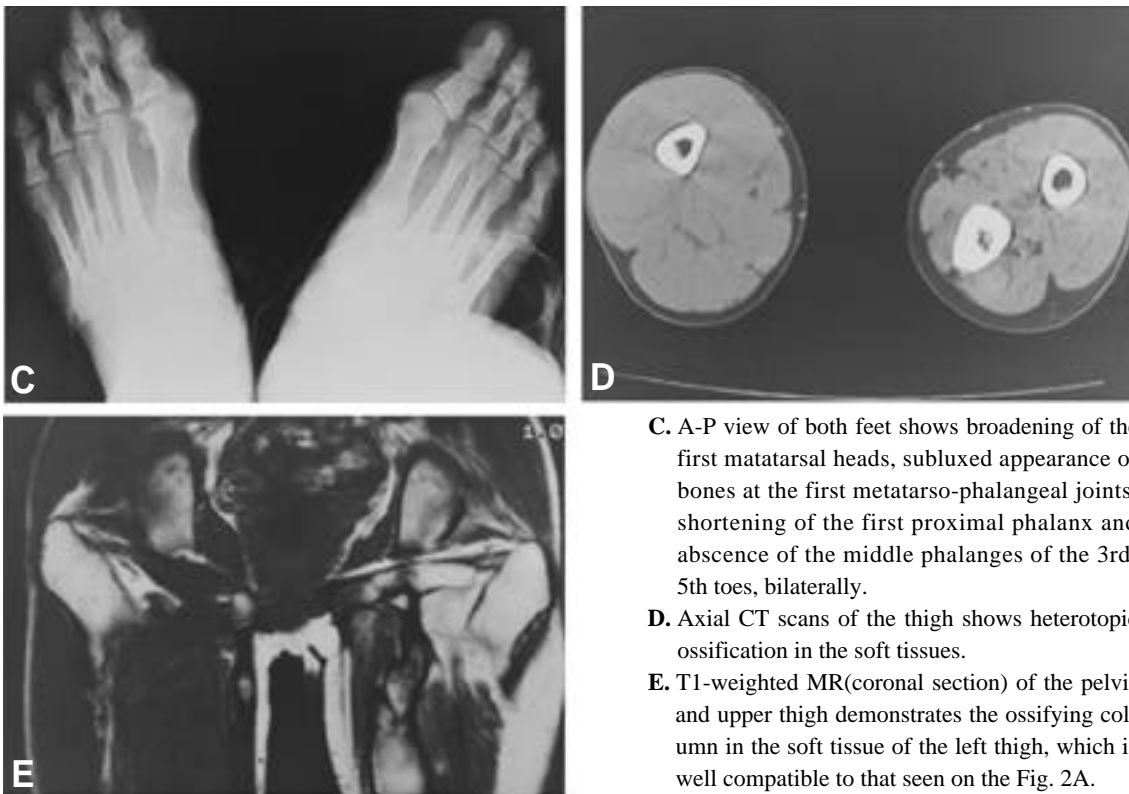


Fig. 2. A. Radiograph of both femur shows osseous protruded mass on the medial condyle of right femur and ectopic ossification of left medial thigh along adductor muscles.
B. Radiograph of lateral view of the cervical spine shows ossification of posterior column of the entire cervical spine with flattening of curvature.



C. A-P view of both feet shows broadening of the first metatarsal heads, subluxed appearance of bones at the first metatarso-phalangeal joints, shortening of the first proximal phalanx and absence of the middle phalanges of the 3rd, 5th toes, bilaterally.
D. Axial CT scans of the thigh shows heterotopic ossification in the soft tissues.
E. T1-weighted MR(coronal section) of the pelvis and upper thigh demonstrates the ossifying column in the soft tissue of the left thigh, which is well compatible to that seen on the Fig. 2A.

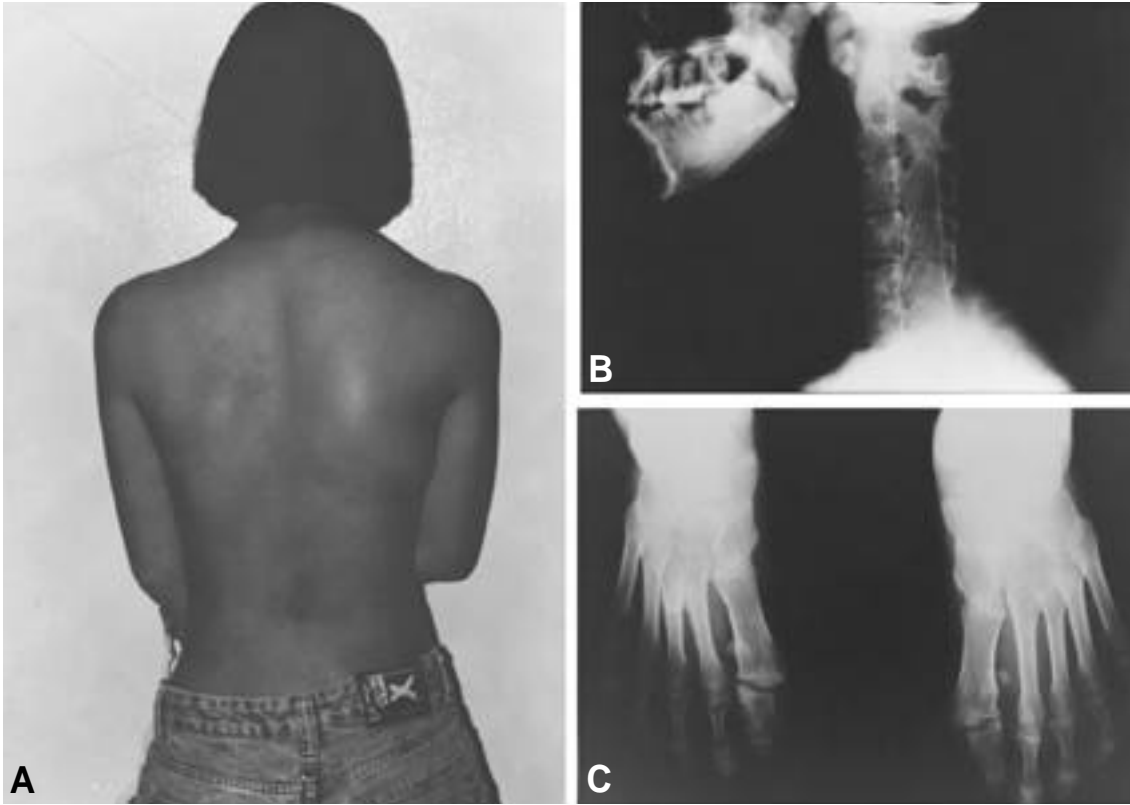


Fig. 3. A. Photograph of the dorsal aspect of the trunk shows diffuse moderate swelling of the right scapular region.
 B. Radiograph shows ossification of the posterior column of the entire cervical spine with flattening of curvature.
 C. A-P view of the foot in the patient's sister shows findings as like as that of the Fig. 2C.

1 20 (Fig. 1 CT MRI
 -1), , (Fig. 2D, 2E).
 , , 2 16 1 (Fig. 1
 . -2), (Fig. 3A),
 8 . 10
 HLA B-27 , , (Fig. 3B),
 , 1
 가 (Fig. 2A). (Fig. 3C).
 가 (Fig. 1 -2)
 (Fig. 2B). 1 , 2
 (Fig. 2C). . (Fig. 1 -1)

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1692 Guy Patin
 2), 1868 von Dusch
 myositis ossificans progressiva
 1932 Mair
 fibrodysplasia ossificans
 progressiva
 7), 1964 5)

Vitamin D
 ACTH cortisone
 가
 (calcium chelating agent)
 diphosphonate disodium ethane-1-hydroxy-1,
 1-diphosphonate(EDTA)
 가
 (ectopic bone)
 9),

BMP4(bone morphogenetic pro-
 tein 4)
 , Isotretinoin
 가 10)

4:1 6)

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8)

가

10

4,9)

1)

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REFERENCES

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- 1) Chung IH, Han DY and Choi DY: Myositis ossificans progressiva. *J Korean Orthop Assoc*, 8:56-58, 1973.
- 2) Cohen RB, Hanh GV, Tabas JA, et al.: The natural history of heterotopic ossification in patients who have fibrodysplasia ossificans progressiva. *J Bone Joint Surg*, 75-A:215-219, 1993.
- 3) Kaplan FS, Strear CM and Zasloff MA: Radiographic and scintigraphic features of modeling in the heterotopic skeleton of patients who have

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- fibrodysplasia ossificans progressiva. *Clin Orthop*, 304:238-247, 1994.
- 4) **Kim JY, Jeon JB, Hwang WS, Chung CY, Kim YJ**: Myositis ossificans progressiva. *J Korean Orthop Assoc*, 7:228-234, 1972.
 - 5) **Kim YJ**: Myositis ossificans progressiva. *Korean Med Doctor's News*, No.192, 1964.
 - 6) **Lucotte G, Semonin O and Lutz P**: A de novo heterozygous deletion of 42 base-pairs in the noggin gene of fibrodysplasia ossificans progressiva patient. *Clin Genet*, 56:469-470, 1999.
 - 7) **Mair WF**: Myositis ossificans progressiva. *Edinburgh Med J*, 39:13-36, 69-92, 1932.
 - 8) **Smith R, Athanasou NA and Vipond SE**: Fibrodysplasia(myositis) ossificans progressiva: clinicopathological features and natural history. *QJM*, 89:445-456, 1996.
 - 9) **Smith R, Russell RGG and Woods CG**: Myositis ossificans progressiva: Clinical features of eight patients and their response to treatment. *J Bone Joint Surg*, 58-B:48-57, 1976.
 - 10) **Zasloff MA, Rocke DM, Crofford LJ, et al.**: Treatment of patients who have fibrodysplasia ossificans progressiva with isotretinoin. *Clin Orthop*, 346:121, 1998.

Abstract

Myositis Ossificans Progressiva - Two Cases Report -

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Myositis ossificans progressiva, also known as ' fibrodysplasia ossificans progressiva ' is a rare disorder, most probably inherited as a mendelian dominant trait with irregular penetrance.

It is characterized by congenital malformations of the great toes and progressive edema, calcification and ossification of the fasciae, aponeurosis, ligaments, tendons, and connective tissue in interstitial tissues of skeletal muscle.

The basic defect is in the connective tissue, whereas the skeletal muscle remains fundamentally normal.

We report two cases of a brother and sister whose the disorder is involved in a same family.

Key Words : Myositis ossificans progressiva, Fibrodysplasia ossificans progressiva

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