Abstract
Nail patella syndrome or hereditary osteo-onycho-dysplasia is an autosomal dominant disorder characterized by dystrophic nails, hypoplastic or absent patellae, hypoplastic elbows and presence of pathognomonic iliac horns with or without renal involvement. The incidence is 22 per million inhabitants live birth children. Although most of the series mention posterior dislocation of the radial head but none mention its management, but we manage it as per patients demand and the case is being reported.

Key words: Nail patella syndrome; uncommon; management

Introduction
Nail patella syndrome or hereditary osteo-onycho-dysplasia is an autosomal dominant disorder characterized by dystrophic nails, hypoplastic or absent patellae, hypoplastic elbows and presence of pathognomonic iliac horns with or without renal involvement. Hereditary nature of the condition was first described by Sedgwick (quoted by Little in 1897). Turner described the syndrome in detail in 1933. Autosomal dominancy with 100% penetrance of this syndrome is linked to aberrancy on human chromosome 9's q arm 9q34. The disorder is linked to the ABO blood group locus. The incidence is 22 per million inhabitants live birth children. Although most of the series mention posterior dislocation of the radial head but none mention its management, but we manage it as per patients demand and the case is being reported.

Case report
A 28 yrs female presented in our OPD in March 2010 with pain and restriction of movement of right elbow for last 1 yr. There was no h/o trauma or massage to the right elbow. Family h/o nail changes. On examination we found short stature of the patient, hypoplastic and atrophic finger nails,
Nail patella syndrome was first reported from India by Maini and Mittal (1966) [1]. Palacios proposed the term hereditary osteo-onycho-arthrodysplasia [2]. Clinical tetrad dysplasia of thumbnail, elbow, patellae and iliac horns are main signs [3,4]. Posterior dislocation of radial head, thickening of axillary border of scapula and minor deformities of wrist are the other common findings reported [5]. Bilateral congenital dislocation of hip in absence of iliac horns are also reported [6]. Flexion deformity of hips and hyperextension of interphalangeal joints are also noted [7]. Maini found flexion contracture of distal interphalangeal joints in 7 cases. Scoliosis was also noted in some cases [3]. Renal affection ranging from asymptomatic proteinuria, recurrent urinary tract infection, nephrotic syndrome, chronic glomerulonephritis, hypertension to end stage renal disease have been reported [8,9].

Autosomal dominancy with 100% penetrance of this syndrome is linked to aberrancy on human chromosome 9's q arm 9q34. The disorder is linked to the ABO blood group locus[10]. Both sexes are equally affected and no generation can skip the disease[1,5]. In previously reported cases nail abnormalities are present in more than 90% of cases and nails may be absent, hypoplastic or dysplastic, and toe nails may be normal. Knee and elbow abnormalities are present in 90% of cases. Patellae may be absent or hypoplastic and may be dislocated in 30% of cases. Complications such as arthritis and knee effusion can cause knee pain. Common elbow symptoms are attributed to radial heads that are typically hypoplastic leading to dislocation. The distal ends of humerus are also hypoplastic and posterior processes limits extension, supination and pronation. Iliac horns are observed in 30% to 70% cases and are pathognomonic. Other bone anomalies affecting the ankle, feet, wrist, scapula, skull and spine has been described. Renal symptoms are present in approximately 50% of cases. Most frequent symptoms are proteinuria, haematuria, nephrotic syndrome and hypertension. End stage renal disease develops in approximately 30% of patients.

**Conclusion**

This type of uncommon syndrome may require judicious management depending on patients demand, as in this case patient demands for her dislocated radial head but not for the habitual dislocation of patellae.

Clinical message: The uncommon Nail patella syndrome may be treated as per patients demand..

**References**


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X-ray of Right Elbow

X-ray of Both Knee

X-ray of Pelvis
POST OP X-RAY

Two and half year follow up