BILATERAL PATELLAR INSTABILITY IN DIGEORGE SYNDROME

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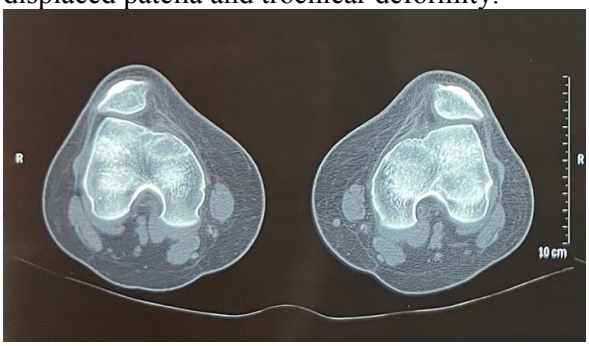
INTRODUCTION:

DiGeorge syndrome is rare genetic condition characterized by cardiac abnormality, abnormal faces, thymic aplasia, cleft palate, and hypocalcemia or hypoparathyroidism.

REPORT:

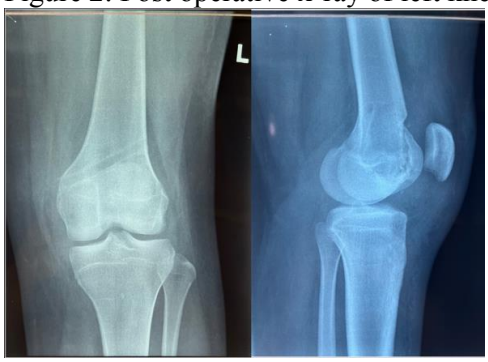
We report a case of 19yo male with underlying DiGeorge Syndrome complaining of recurrent left patella dislocation. Clinical examination showed slightly varus alignment, bilateral calcaneal varus with rocker bottom feet, patella J-sign positive, no effusion, ROM -15° - 140°, lateral patella mobility >2 quadrants with positive apprehension. Beighton score 8. KUJALA score 47. Radiographs showed a laterally subluxated patella, double contour sign with supratrochlear spur. Caton-Deschamps index right 1.06, left 1.23. Computed tomography femoral anterversion was normal right 40°, left 30°. TT:TG 22.2mm, trochlear deformity Dejour D with supratrochlear spur.

Figure 1: Pre op CT knee demonstration lateral displaced patella and trochlear deformity.



We scheduled diagnostic arthroscopy, open trochleoplasty and medial patella-femoral ligament (MPFL) reconstruction using ipsilateral hamstring graft. Intra operatively, maltracking of the patella dislocated at 0°-45°, chondral lesion at central ridge ICRS grade 3. MPFL reconstruction using double strand semi-tendinosus graft anchored on the medial patella ridge with 2 3.5mm Peek CF suture anchors. Femoral origin determined radiographically by Schottle point, technique as described by Schottle et al. fixation using ActivaScrew Interference 7x24mm. Open trochleoplasty, osteochondral flap secured using Fibretape and PEEK PushLock system.

Figure 2: Post operative x-ray of left knee



Post operatively, patella was restrained well and stable throughout range of motion. At 3-6 months follow up, global improvement in IKDC and KUJALA scoring.

CONCLUSION:

Although ligamentous laxity is not characteristic of DiGeorge, a high index of suspicion is required when approaching syndromic cases.

REFERENCES:

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