

Mietens' Syndrome: A Case Report

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Citation

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Abstract

In 1966, Carl Mietens and Helge Weber published a case series of four children who suffered from a unique cluster of congenital anomalies. These four children were siblings born of consanguineous parents. The constellation of abnormalities included growth failure, dislocation of radial head, bilateral corneal opacities, horizontal and rotational nystagmus, strabismus, small pointed nose with a broad base and mild to moderate mental retardation. A comprehensive search from Pubmed revealed that until now nine cases have been reported, mostly from European countries and Japan. We report what we believe to be the tenth case of Mietens'-Weber syndrome.

INTRODUCTION

In 1966, Mieten and Weber (1) reported a characteristic cluster of anomalies in four out of six sibling born of consanguineous parents in southern Germany. These anomalies included:

1. Mental retardation
2. Growth failure
3. Flexion contracture of the elbows with dislocation of radial head
4. Abnormally short ulna and radius
5. Bilateral corneal opacity
6. Strabismus
7. Horizontal and rotational nystagmus
8. Clinodactyly
9. Small pointed nose with a depressed root.

At that time, this combination of abnormalities were not related to any other known syndromic manifestation.

Again in 1977, Nagano et al (2) reported a case which had remarkable similarities with the ones reported by Mieten and Weber. In the following years, a few more case reports were published in English and non-English literature(3). Until now, to the best of our knowledge, nine cases have been reported. We hereby report a case, which has striking

similarities to the ones reported by Mieten and Weber. This is also the first case of Mietens syndrome reported from the Indian subcontinent.

CASE REPORT

A 19 year old Mr. D was born of non-consanguineous parents through normal vaginal delivery. Birth weight was unknown but hospital records at 4 months of age suggest that he was 5 kg at that time. Boy achieved all developmental milestones normally. Bilateral corneal opacities were noted at 4 months of age for which he was under regular follow-up in the ophthalmology department as a case of Peter's anomaly. The boy was referred to our Out Patients Department for bony prominence of both the elbows. Clinico-radiological examination suggested bilateral dislocation of the radial head. No flexion deformity at the elbow was appreciated. Range of movements at the elbow were normal with no loss of rotatory movements of the forearm. There was no history of trauma. Radiological evidence like dysplastic radial head, short ulna in comparison with radius along with exaggerated posterior bowing were also seen. Other deformities included bilateral corneal opacities, prominent beak shaped nose with a broad base, high arched palate, microphthalmia and scoliosis. An internet search was done to see if this constellation of anomalies belong to any known syndrome. Congenital radial head dislocation is known to be associated with Nail-patella syndrome, Apert's syndrome, Mieten-Weber syndrome among others. Our case closely fits the description of Mietens' syndrome.

Figure 1

Fig.1: Clinical photograph showing posterior prominence of both elbows.



Figure 2

Fig.2(A, B): clinical photograph showing pronation and supination (within normal range).

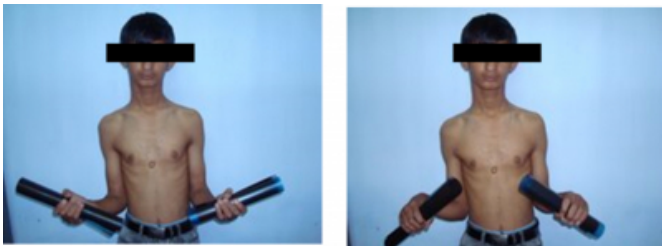


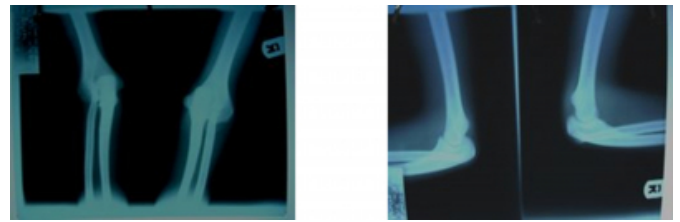
Figure 3

Fig.3(A, B): Clinical photographs showing flexion - extension at the elbow (within normal range).



Figure 4

Fig.4(A,B): Antero-posterior & Lateral; Radiographs depicting bilateral dysplastic radial heads and its dislocation, shorter ulna with exaggerated posterior bowing.



DISCUSSION

Congenital radial head dislocation though rare is one of the commonest anomalies of the elbow joint (4). The dislocation can be anterior, posterior or lateral. Literature is inconclusive as to which type is most common. It has been suggested that (5), while anterior occurs as an isolated anomaly, posterior dislocation is associated with other abnormalities as part of a syndromic manifestation. Some syndromes which include dislocation of radial head include (2):

1. Hemimelia and brachydactylia
2. Diaphyseal achlasia
3. Klinefelter syndrome
4. Cornelia de lange syndrome
5. Ehlers-danlos syndrome
6. Hereditary osteochondritis dissecans
Athrogryphosis
7. Dysplastic epiphysialis multiplex
8. Nail patella syndrome
9. Nivergelt's syndrome
10. Cliedo-cranial dysostosis Silver's syndrome
11. Apert's syndrome
12. Congenital web formation
13. Mieten's syndrome

Amongst the above ,our case best fits with the description of Mietens' syndrome. Pictures reveal striking facial similarities of our case and the previous ones. However , there was no flexion deformity at the elbows and hence no reactive hypertrophy of the trapezius muscle. Mention must

also be made of Cornelia de Lange syndrome(5) ,features of which closely resemble Mietens' syndrome except that it lacks eye features.

Going through the previously published reports and ours, attention must be drawn to the fact that these patients also seem to have some kyphotic deformity of the spine. It needs to be looked further in this regard whether spinal deformity is a constant feature of Mietens' syndrome along with the pros and cons of any surgical procedure in such kind of asymptomatic patients.

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