Occurrence of Polymelia in a Female Child

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ABSTRACT

We report a rare case of polymelia in a 6-month-old female child who presented with developed lower limbs and an additional underdeveloped left lower limb.

Key words: Anatomy, infant, polymelia, radiology

INTRODUCTION

Polymelia is a congenital anomaly, which is defined as the presence of accessory limbs attached to various body regions and could be classified as cephalomelia (extra-limb attached to the head), notomelia (extra-limb attached to the back bone), thoracomelia (extra-limb attached to the thorax), and pyromelia (extra-limb attached to the pelvis). These anomalies are usually associated with genetic factors including tamogens, chromosomes, and environmental agents. Here, we report a rare case of pyromelia, a form of polymelia.
DISCUSSION

Isolated limb duplication is a rare congenital condition and only a few cases have been documented. Various adverse embryogenic influences are responsible for this kind of anomaly\(^1\)\(^-\)\(^4\).

Limb differentiation occurs roughly between the 4\(^{th}\) and 5\(^{th}\) weeks of embryonic development\(^3\), and it follows a dorsal to ventral and proximal to distal pattern, with many factors involved in the process\(^3\)\(^,\)\(^4\).

Initially, two pairs of limb buds – anterior and posterior – protrude from both sides of the embryo, and comprise cells of ectoderm and mesoderm. Their interaction is responsible for cell positioning and limb differentiation. The covering ectodermal layer of the limb bud is termed the apical ectodermal ridge (AER). The zone of proliferating activity (ZPA), another group of cells is located subjacent to the AER. Both are necessary for limb development. Mesodermal cells in the ZPA stimulate AER formation and the AER maintain the ZPA\(^6\)\(^,\)\(^7\).

The level and manifestation of limb deformity can thus be used to determine the approximate timing of the teratogenic event that occurred during limb development. As the AER grows more distal, the induced mesoderm cells, comprising rudimentary parts of the limb, can continue to grow without any developmental interference even if the AER is transplanted to the adjacent region. This leads to an assumption that duplication of the limb arises from the influence of the AER with abnormal splitting creating two sets of limbs\(^8\)\(^,\)\(^9\).

Other deformities in our patient such as semivertebrae, spina bifida, scoliosis, and hip dislocation were absent. We found only a left club foot.

Several factors lead to this anomaly [Table 1]. Over 80% of children born to mothers who took thalidomide had limb defects. These defects ranged from absence of the limb (amelia) or proximal limb elements (phocomelia) to loss of the thumb or digit tip. Women taking hormone (progestrone etc.) during pregnancy have chances of the structural anomalies like polydactyly, polymelia, and other congenital defects. Antenatal screening with ultrasonography can be a useful tool to diagnose such conditions in utero.

Treatment for such a case like ours requires surgery to detach the soft tissue between false limb and the true pelvic region.

CONCLUSION

Radiologist plays a significant role in patients with polymelia to assess for additional congenital anomalies before surgical intervention.

REFERENCES

1. Griffet J, Bastiani-Griffet F, Jund S, Moreigne M, Zabjek KF. Duplication of the leg – Renal agenesis: Congenital malformation syndrome. J Pediatr Orthop B 2000;9:306-8.
2. La Torre R, Fusaro P, Anceschi MM, Montanino-Obiva M, Modesto S, Cosmi EV. Unusual case of caudal duplication (dipygus). J Clin Ultrasound 1998;26:163-5.

Table 1: Factors leading to polymelia

| Factor                           | Agents                          |
|---------------------------------|---------------------------------|
| Genetic factor                  | Trans-genes, chromosomes        |
| Environmental                   | Infectious agents, toxins       |
| Techniques involved in fertilization |                              |
3. Kher AS, Gahankari DR, Tambwekar SR, Doraiswamy A, Iyer S, Bharucha BA, et al. Supernumerary limbs: A case report of a rare congenital anomaly. Ann Plast Surg 1996;37:549-52.
4. Kojima T, Hirakawa M, Hirase Y, Hwang KH. Complete congenital duplication with incomplete separation of a lower extremity. Plast Reconstr Surg 1993;91:926-9.
5. O’Rahilly R, Muller F. Developmental Stages in Human Embryos. Washington: Carnegie Institution; 1987. Publication No. 637.
6. Roberts DJ, Tabin C. The genetics of human limb development. Am J Hum Genet 1994;55:1-6.
7. Scherz PJ, Harfe BD, McMahon AP, Tabin CJ. The limb bud Shh-Fgf feedback loop is terminated by expansion of former ZPA cells. Science 2004;305:396-9.
8. Packard DS Jr, Levinsohn EM, Hootnick DR. Extent of duplication in lower-limb malformations suggests the time of the teratogenic insult. Pediatrics 1993;91:411-3.
9. Zhao L, Li MQ, Sun XT, Ma ZS, Guo G, Huang YT. Congenital lumbosacral limb duplication: A case report. J Orthop Surg (Hong Kong) 2006;14:187-91.

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