Congenital Partial Hemihypertrophy, Low Set Ears, Hypertelorism, and Epicanthi Folds: A Novel Syndromic Association

Aamir Jalal Al Mosawi
Advisor in Pediatrics and Pediatric Psychiatry, Children Teaching Hospital of Baghdad Medical City Head, Iraq Headquarter of Copernicus Scientists International Panel Baghdad, Iraq.

*Corresponding author: Aamir Jalal Al Mosawi, Advisor in Pediatrics and Pediatric Psychiatry, Children Teaching Hospital of Baghdad Medical City Head, Iraq Headquarter of Copernicus Scientists International Panel Baghdad, Iraq.

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Abstract

**Background:** Congenital hemihypertrophy is a very rare condition that can be isolated or occurs in association with other congenital abnormalities and neoplasms, and well-recognized syndromes. Well-known syndromes that are associated with congenital hemihypertrophy include Klippel-Trenaunay-Weber syndrome, Beckwith Wiedemann syndrome, Goldenhar syndrome, Silver-Russell's syndrome. Partial hemihypertrophy affecting a limb is an extremely rare condition, and was probably first reported in 1949 by web, and very few cases have been reported in the literature.

**Results:** The boy was hypotonic with developmental delay and has not been able sit yet. He had low set ears and facial dysmorphism consisting of hypertelorism and epicanthic folds. His right lower limb was obviously larger than the left. Parents were consanguineous, and family history was negative for similar condition. The rest of the examination was normal. Brain CT-scan, echocardiography, abdominal ultrasound, and chromosomal karyotype showed normal finding.

**Conclusion:** Hemihypertrophy has not been reported in Iraq before. A novel syndromic association consisting of congenital partial hemihypertrophy, low set ears, hypertelorism, epicanthi folds, and developmental delay is reported in this paper.

**Key words:** congenital partial hemihypertrophy; low set ears; hypertelorism; epicanthi folds; and developmental delay; new syndrome.

Introduction

Congenital hemihypertrophy is a very rare condition that can be isolated or occurs in association with other congenital abnormalities and neoplasms including hepatoblastoma (Geiser et al; 1970; Rattan et al; 1995); Wilms tumor (Sauer and Wemmer; 1977; Mohanna and Sallam; 2008); embryonal rhabdomyosarcoma (Samuel; Tsokos; and DeBaun; 1999); epithelioid haemangioendothelioma (Miller et al; 1999); congenital mesoblastic nephroma (Absoudah et al; 2008) [1-29]; and well-recognized syndromes. Well-known syndromes that are associated with congenital hemihypertrophy include Klippel-Trenaunay-Weber syndrome; Beckwith Wiedemann syndrome; Goldenhar syndrome; Silver-Russell's syndrome [30-33].

Partial hemihypertrophy affecting a limb is an extremely rare condition; and was probably first reported in 1949 by web [34]; and very few cases have been reported in the literature.

Patients and methods

Ten month old infant with partial hemihypertrophy; delayed development and facial dysmorphism who was observed at the pediatric neuropsychiatry clinic of Baghdad Medical City was studied.

Results

The boy was hypotonic with developmental delay and has not been able sit yet. He had low set ears and facial dysmorphism consisting of hypertelorism and epicanthic folds (Figure-1). His right lower limb was obviously larger than the left (Figure-2). Parents were consanguineous; and family history was negative for similar condition. The rest of the examination was normal. Brain CT-scan; echocardiography; abdominal ultrasound; and chromosomal karyotype showed normal finding.

Discussion

Congenital hemihypertrophy is well known to be associated with multiple congenital anomalies (Table-1). Partial hemihypertrophy affecting a limb is an extremely rare condition; and was probably first reported in 1949 by web [34]; and very few cases have been reported in the literature [12; 35; 36; 37].

Kasantikul et al (1994) reported a rare case of adrenocortical adenoma in a 7-month-old female infant with congenital hemihypertrophy of left leg [35].

Gönül et al (2009) from Turkey report a very rare case of giant melanocytic naevas with lipomatosis; Dandy-Walker malformation occurring in association with hemihypertrophy of the leg [36].
Clinical Research Notes

Mohanna and Sallam (2008) from Yemen reported a case presented with partial hemihypertrophy of the right leg and foot with polydactyly; a right sided abdominal mass caused by an ureteropelvic junction stricture with hydronephrosis; and absent left kidney [12].

Deyrup et al (2011) from USA reported a very rare association of cutaneous angiosarcomas with congenital hemihypertrophy of the contralateral limb [37].

**Author(s)** | **Congenital anomalies**
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Paré and Elhilali (1972) [14] | Hydrometrocolpos and polydactyly
Henry et al (1973) [16] | Abnormalities of the aortic arch; and of the skeletal; cutaneous and ocular systems.
Temtamy and Rogers (1976) [17] | Macrodactyly; and connective tissue nevi
Fischer ; Strand ; and Shapiro (1984) [18] | Abnormalities of the cerebral vasculature : giant aneurysm; capillary hemangioma; and arteriovenous malformation
Hidano and Arai (1987) [19] | Syndactyly; scoliosis; short forth metacarpus; hypoplastic mandible; peroneal exostosis ;multiple faint nevi flammei ; telangiectasis; nevus anemicus ; fibromatos tumors of the tip of the tongue; mitral prolapse; and vascular; cerebral; abnormalities
Giani ; Lapi ; Pezzati (1991) [21] | Nevus depigmentosus
Dawn et al (1995) [22] | Right upper limb triplication; polythelia; ; congenital hip dislocation; facial dysmorphism; congenital heart disease; and scoliosis
Akarsu et al (2005) [27] | Renal dysplasia and benign nephromegaly

**Table-1: Abnormalities associated with congenital hemihypertrophy**

**Conclusion**

Hemihypertrophy has not been reported in Iraq before. A novel syndromic association consisting of congenital partial hemihypertrophy; low set ears; hypertelorism; epicanthi folds; and developmental delay is reported in this paper.
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