Idiopathic hemihypertrophy

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ABSTRACT

التضخم في جانب واحد من الجسم هو اضطراب فرط الخلقية المرتبطة بخطر زيادة للأورام الجنينية. تقرير عن 4 حالات لأطفال يمنيين يعيشون في صنعاء حضروا العيادة في مستشفى سام الطبي، اليمن. وذلك خلال 3 سنوات من يناير عام 2009م إلى ديسمبر عام 2011م . الحالتان الأولى والثانية عندهما فقط تضخم في الجانب الأيسر من الجسم، أما الحالة الثالثة عندها جانب التضخم في الجانب الأيسر ورم في البطن، تلك الحالة حُولت إلى مركز السرطان وشخصت (وليمز تيومر) ،الحالة الرابعة عندها فرط في نمو الرجل اليمنى والقدم مع تعدد الأصابع مصحوباً بورم في الجانب الأيمن من البطن (تورم في الكلية اليمنى). تم تشخيص حالته بأنه لديه تضيق حالبي تقاطع مع موه الكلية وغياب الكلية اليسرى.

Idiopathic hemihypertrophy is a congenital overgrowth disorder associated with an augmented risk for embryonal tumors. We present 4 cases of hemihypertrophy in Yemeni children living in Sana'a city. They presented to the outpatient clinics in a private hospital in Sana'a city, Yemen, over a period of 3 years from January 2009 to December 2011. The first 2 cases had no complaints apart from asymmetrical size of one side of their bodies (left side hemihypertrophy). The third case presented with left side hemihypertrophy and an abdominal mass, which upon referral to a cancer center was confirmed to be Wilms' tumor. The fourth case had overgrowth of the right leg and foot with polydactyly, and a right sided abdominal mass. He was diagnosed to have an ureteropelvic junction stricture with hydronephrosis and absent left kidney.

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Hemihyperplasia, previously called hemihypertrophy, is a rare disorder in which one part of the body grows more than the other, causing asymmetry. An asymmetric overgrowth of unknown etiology may involve the total of one side of the body, or it may be restricted in extent to one limb or a side of the face.[1,2] Tumors of the adrenal gland (with or without Cushing syndrome) may be linked with hemihypertrophy, frequently occurring through the first few years of life. These tumors are moreover, associated with Beckwith-Wiedemann syndrome and other congenital defects, chiefly genitourinary tract and CNS abnormalities and hamartomatous defects.[3-5] The tendency for neoplasia in isolated hemihyperplasia is well recognized, but the accurate possibility is not well known. We lay out the diagnostic criteria and tumor screening recommendations for children with isolated hemihyperplasia, based on the most recent information in the literature.[6] Wilms' tumor can be successfully treated, with a general cure rate of more than 85%, by almost uncomplicated therapies. This excellent result has been the effect of combined hard work among surgeons, pediatricians, pathologists, and radiation oncologists.[7] Our objective in presenting these particular cases is to highlight the management of patients with hemihypertrophy.

Case Report. *Patient 1.* A 3-year and 5-month-old girl presented to the outpatient clinic of a private hospital with overgrowth of the whole left side of the body. She is the second child product of a non-consanguineous marriage. On examination, she appeared well, her weight was 13.5 kg, her left side was larger than the right (left hemihypertrophy), and she had no other dysmorphic features. Her pulse, respiratory rate, and blood pressure were normal. Abdominal examination revealed no masses or organomegaly. Investigation including a complete blood count, urine analysis, abdominal ultrasound, and hip joint x-ray were normal. Parents were well informed to examine the child's abdomen weekly, and were also asked to perform abdominal ultrasound scans on her every 3 months.

Patient 2. A 2-year and 2-month-old boy presented to the outpatient clinic of a private hospital with

enlargement of the whole left side of the body. He had 4 siblings and was the youngest; his siblings and parents were normal, and there was no consanguinity between mother and father. On examination, no dysmorphic features were noticed apart from the left sided hemihypertrophy, his weight was 11.5 kg, and the left side of the body was larger than the right side; his pulse, respiratory rate, and blood pressure were normal. Abdominal examination revealed no masses or organomegaly. As in the previous case, the investigations including a complete blood count, urine analysis, abdominal ultrasound, and hip joint x-ray, which were normal. His parents were trained to examine the child's abdomen weekly, and were asked to perform an abdominal ultrasound on him every 3 months (**Figures 1 & 2**).

Patient 3. A 3 and half-year-old boy presented to our hospital with an abdominal mass, it was noted by the mother while bathing and dressing him, and she also noticed that the 2 halves of the child's body were asymmetrical; the left side of the body was larger than the right. His development was normal, he walked when he was 12 months old, and his intelligence seemed to be within the normal range. He was the youngest among 7 siblings; his siblings and parents were normal and there was no consanguinity between parents. On examination: the pulse, respiratory rate, and blood pressure were normal. He had left side hemihypertrophy, the left leg was one centimeter longer than the right, and there was a 6 × 7 cm abdominal mass in the left loin which was hard, but not tender. Abdominal ultrasound revealed a mass in the left kidney and it was proven by histopathological examination of the biopsy to be Wilm's tumor. He was referred to the surgical department in a cancer center for further follow-up.

Patient 4. A 4 and half-month-old boy presented to our hospital with enlargement of the right leg and foot as well as extra digits (seven toes) and also swelling in the right side of the abdomen. He is the first child, and a product of a non-consanguineous marriage. On examination, he appeared alert, and well, his weight was 5.5 kg, pulse, respiratory rate, and blood pressure were normal. He had overgrowth of the right leg and foot with polydactyly and a right-sided abdominal mass. The mass was 3×4 cm, and hard but not tender. Laboratory findings revealed hemoglobin of 9 g/dl, white blood cell count was $10x10^9$/L, blood urea was 80 mg/dl, and serum creatinine was 1.5 mg/dl. Abdominal ultrasound revealed that the right kidney was enlarged with prominent dilatation of the right pelvicalyceal system due to ureteropelvic junction stricture with moderate parenchymal reduction, and the left kidney was not detected (congenitally absent). This patient was referred to a urologist (**Figure 3**).

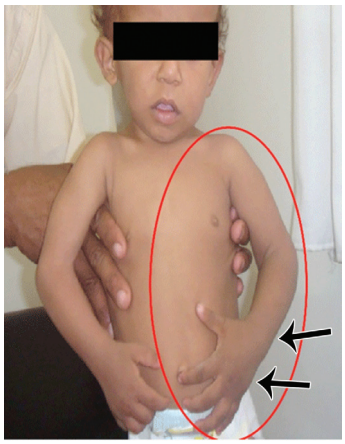

Figure 1 - Enlargement of the left hand and forearm (arrows) in patient 2.

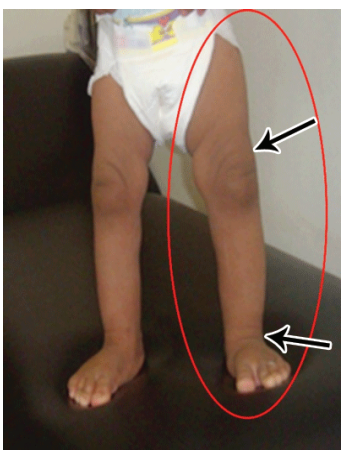

Figure 2 - Enlargement of the left foot, leg, and thigh (arrows) in patient 2.

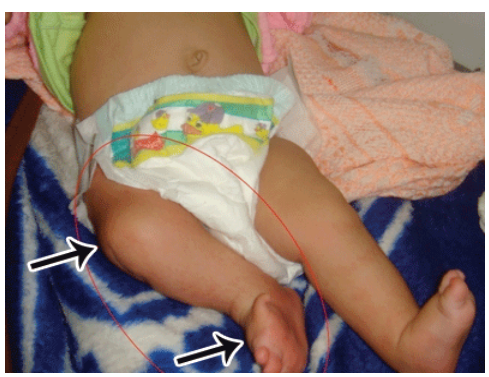

Figure 3 - Overgrowth of the right leg and foot with polydactyly (arrows) in patient 4.

Discussion. Idiopathic hemihypertrophy is an unusual enlargement or overgrowth of half of the body or half of a body part. There may be associated asymmetric hypertrophy of internal organs. The reported incidence of hemihyperplasia is thought to occur in around one in 86,000 live births, yet this number is not constant.[8] Hemihyperplasia may be an isolated finding or it may be associated with other syndromes such as Beckwith-Wiedemann, Klippel-Trenaunay-Weber, or McCune-Albright syndromes. Idiopathic hemihypertrophy is often linked with mild mental retardation, genito-urinary anomalies, and an oncogenic potential (Wilms' tumor).[9]

Wilms' tumor accounts for most renal neoplasms in childhood, and occurs with roughly equal incidence in both genders and all races, with a yearly incidence of 7.8 per million children younger than 15 years.[3] An imperative feature of Wilms' tumor is the association with congenital anomalies, the most common being genitourinary anomalies (4.4%), and hemihypertrophy (29%).[3] The risk of tumor development in isolated hemihyperplasia is approximately one in 20, or around 5%. The best follow-up plan is to follow the patients until the age of 6 years; these children should have abdominal ultrasound scans at 3 monthly intervals. There is currently inadequate indication to screen children above 6 years of age.[10]

Of the 4 reported cases that were seen over a period of 3 years in our hospital, only one had Wilms' tumor, but there might be more cases seen in other hospitals, or may have not been clinically diagnosed, especially those who have only isolated hemihypertrophy.

In conclusion, idiopathic hemihypertrophy occurs in all races, it is often associated with genito-urinary anomalies, and an oncogenic potential. Parents should be trained to examine the child's abdomen weekly, and the child should have abdominal ultrasound scans every 3 months.

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