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Virchow Seckel Syndrome: The First Case in Iraq and the Early Documentation of the Syndrome in the Literature

Al-Mosawi AJ[1,2*]

[1]Department of Pediatrics and Pediatric Psychiatry, Children Teaching Hospital of Baghdad Medical City, Iraq

[2]Head, Iraq Headquarter of Copernicus Scientists International Panel, Iraq

1. Abstract

Background: Virchow Seckel syndrome or bird-headed dwarfism syndrome is a very rare genetic syndrome characterized by intrauterine and postnatal growth retardation with very poor growth of the body and head, narrow bird-like face with a peculiar nose, mental retardation and other congenital abnormalities. An autosomal recessive inheritance and a heterogeneous nature of the condition have been expected. This Virchow Seckel syndrome has not been reported in Iraq.

Patients and methods: Four years and four months old girl was referred to the pediatric neuropsychiatry clinic of the Children Teaching Hospital of Baghdad Medical City because of significant growth and developmental retardation. The child was studied and the relevant medical literature was reviewed with aim of describing the early documentation of her rare condition in the medical literature.

Results: The girl weight at birth was about 1.5 kilograms. She was experiencing very poor growth and her height was 72 cm and her weight 6 kilograms. She had low set ears, small head with narrow face, downward slanting eyebrows and a peculiar nose. She was also mentally retarded with poor language development. Family history was negative for similar cases. Bone age assessment was performed using radiographs of the left and wrist, left elbow, hips and knee and showed delayed bone age of about one year.

Conclusion: During the first century of documentation of this syndrome (1882-1981, about 35 were reported in the literature and in this paper, the first case of this syndrome in Iraq is described.

2. Keywords: Virchow Seckel syndrome; Historic documentation; Iraq

3. Introduction

Virchow Seckel syndrome or bird-headed dwarfism syndrome is a very rare genetic congenital disorder characterized by intrauterine growth retardation and very poor postnatal growth of the body and head, low-set ears, narrow bird-like face with a peculiar nose, down-slanting eyes, receding forehead and mental retardation. An autosomal recessive inheritance and a heterogeneous nature of the condition have been suggested. Various congenital abnormalities have been reported in association with this syndrome [1-14]. This rare disorder has not been reported in Iraq [16-18].

4. Patients and Methods

Four years and four months old girl was referred to

***Corresponding author:** Al-Mosawi AJ, Department of Pediatrics and Pediatric Psychiatry, Children Teaching Hospital of Baghdad Medical City, Iraq, E-mail: almosawiAJ@yahoo.com

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the pediatric neuropsychiatry clinic of the Children Teaching Hospital of Baghdad Medical City because of significant growth and developmental retardation. The child was studied and the relevant medical literature was reviewed with aim of describing the early documentation of her rare condition in the medical literature.

5. Results

F.H was first seen during at the age of 4 years, 4 months and 5 days. She was born to healthy consanguineous parents and her weight at birth was about 1.5 kilograms. She was experiencing very poor growth and her height was 72 cm and her weight 6 kilograms. She had low set ears, small head with narrow face, downward slanting eyebrows and a peculiar nose (Figure 1).

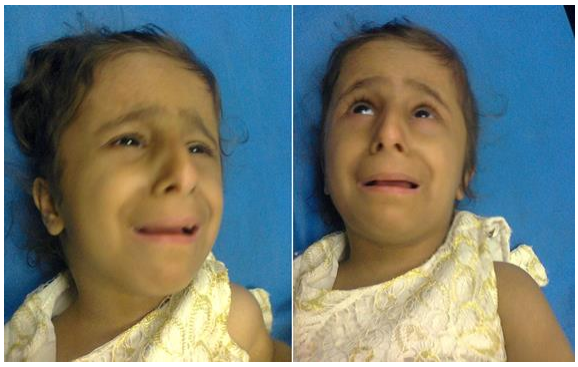

Figure 1: The girl had low set ears, small head with narrow face, downward slanting eyebrows and a peculiar nose.

She was also mentally retarded with poor language development. Family history was negative for similar cases. She had an older brother aged 9 years, his weight was 26 kilograms and his height was 128 cm. She also had two sisters aged eleven and thirteen years respectively, but they were not available for examinations and measurements. Bone age assessment was performed using radiographs of the left and wrist, left elbow, hips and knees and showed delayed bone age of about one year. Radiograph of the left wrist (Figure 2A) showed the appearance of hamate, capitate, epiphysis of radius, triquetrum, epiphysis of metacarpals and phalanges, with absence of the lunatum which appears at four years. Radiographs of the left elbow (Figure 2B) showed the capitellum which appears at two years. Radiographs of the hips showed the great trochanters which appear at four years (Figure 2C). Radiographs of the knees didn't show the heads of fibulae which appear at four years (Figure 2D). Hormonal assessment and chromosomal karyotype showed no abnormalities.

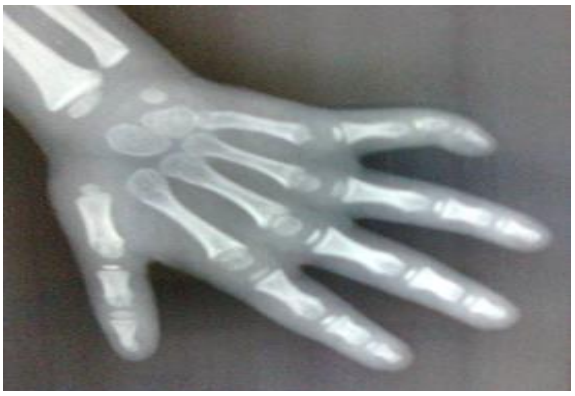

Figure 2A: Radiograph of the left wrist showing the appearance of hamate, capitate, epiphysis of radius, triquetrum, epiphysis of metacarpals and phalanges, with absence of the lunatum which appears at four years.

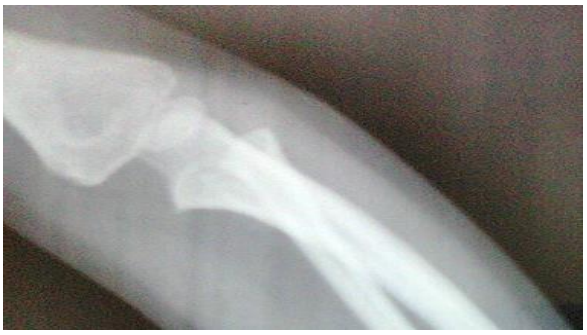

Figure 2B: Radiographs of the left elbow showing the capitellum which appears at two years.

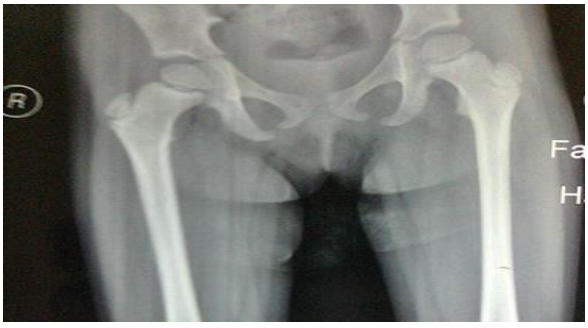

Figure 2C: Radiographs of the hips showing the great trochanters which appear at four years.

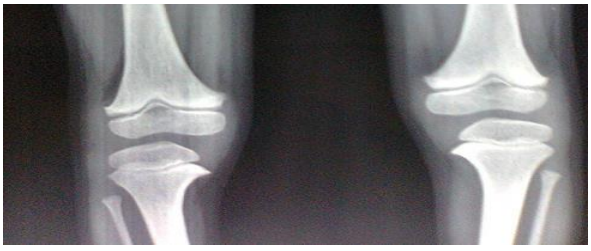

Figure 2D: Radiographs of the knees didn't show the heads of fibulae which appear at four years.

6. Discussion

Rudolf Ludwig Carl Virchow (Figure 3) was most probably the first to describe the clinical association of proportionately small head and body with mental retardation and a peculiar appearance which he described as a bird-headed dwarfism.

Figure 3: A sketch of Rudolf Ludwig Carl Virchow (October, 13, 1821-September, 5, 1902), a German physician.

Figure 4: Helmut Paul George Seckel (1900-1960), an American physician.

Thereafter, an American physician Helmut Paul George Seckel (Figure 4) provided in 1960 a more detailed description of the occurrence of the condition based on his two patients and previously reported cases in the literature. In his paper, Seckel discussed the case of Caroline Crachami, whose skeleton remains in the Hunterian Museum of the Royal College of Surgeons, London. Her height was 49.5 cm when she died in 1824 at the age of nine years.

In 1967 McKusick et al. reported the occurrence of the syndrome in 3 of 11 siblings and suggested the possibility of autosomal recessive inheritance [7].

In 1981, Mitzkat and Dietz reported two cases and were able to count only 20 cases previously. However, with our sophisticated review we found at least 35 cases had been reported over about one century (1882-1981) before Mitzkat and Dietz reported their cases in 1981 [1-14,19-25].

7. Conclusion

During the first century of documentation of this syndrome (1882-1981, about 35 were reported in the literature and in this paper, the first case of this syndrome in Iraq is described.

8. Acknowledgement

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