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Chronic Intestinal Volvulus In A Patient Followed For Seckel's Syndrome

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Abstract: *Seckel syndrome belongs to osteodysplasic dwarfism with microcephaly. It is an hereditary autosomic recessive abnormality characterised by the association of a prenatal dwarfism, facial dysmorphism called birds head and a mental retardation.*
We report the case of a 13-year- old female patient who consults for chronic vomiting with weight loss. Clinical examination revealed a generally altered patient with microcephaly, a narrow face on which is implanted a prominent nose curved into a bird's beak, eye catching, fading forehead with clinodactyly .
Radiological investigations were performed the surgical exploration showed a volvulus of the small bowel.
The operative follow-up didn't show any complications
Key words*: Seckel syndrome - volvulus –small bowel*

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I. Introduction

Helmet Seckel described Seckel syndrome as an hereditary autosomic recessive abnormality characterised by a cleft lip, cleft palate, clubfoot, scoliosis, gastrointestinal abnormalities and other deformations of the skeleton. [1-2]

Other abnormalities were described such as microcephaly, cranio-facial dysmorphy and intra-uterine delay of growth.

The pathogenesis of Seckel syndrome is primarily based on marked growth inpaiments. There are chromosomal common fragile sites in Seckel syndrome patients making them prone to malignancies [1-3].

II. Observation

A 14 years old girl was admitted with chronic vomiting, staturo-ponderal delay.
On the physical examination, some malformations were noted such as: microcephaly with a small forehead, a facial dysmorphism (figure 1)with a small face, a prominent beaked triangular nose (figure 1), clinodactyly (figure 3-4)of the thumb and prominent eyes (figure 2).

Figure 1-figure 2: facial dysmorphism

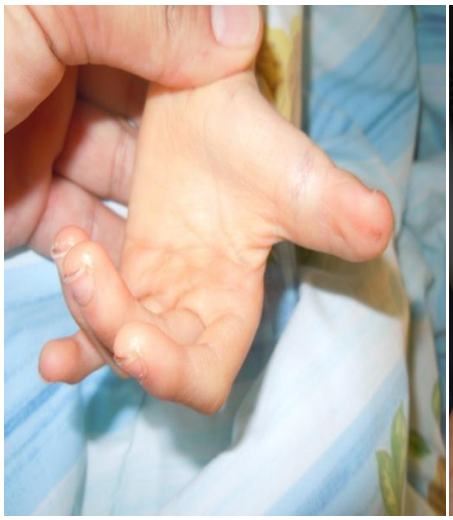

Figure 3

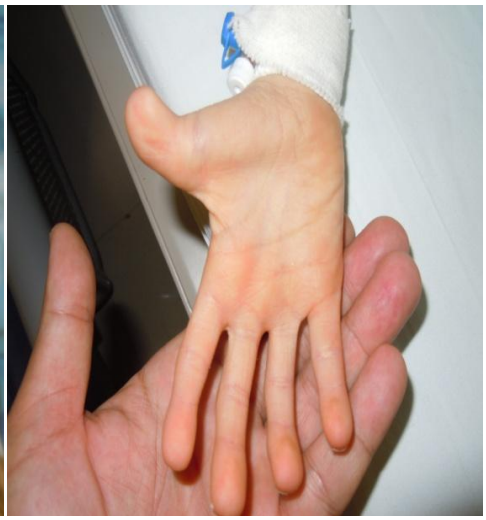

figure 4 :

Figure 3-4 :clinodactily of the thumb

An abdominal X-ray and an upper gastro-intestinal test (figure 5) were performed and showed a dilation of the first portion of the duodenum as well as the bulbar region upstream to a shrinkage of the junction between the first end the second portion of the duodenum.

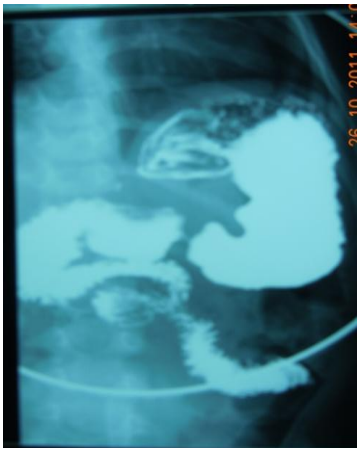

Figure 5 : dilation of the first portion of the duodenum

An upper gastrointestinal endoscopy was performed that showed a dilation in the bulbar and first portion of the duodenum.
Laparotomy showed a volvulus of the small intestine with viable handles after devolvulation.

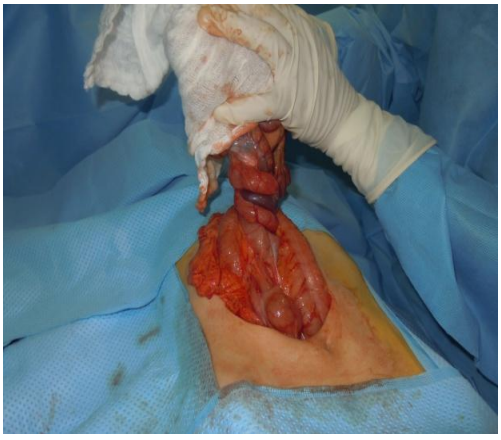

Figure 6: volvulus of the small bowel

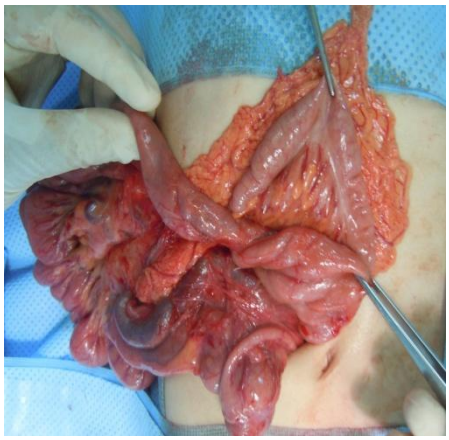

Figure 7: bowels after devolvulation

Post-operative follow up showed no complications. The patient was cleared up four days after the surgery.

III. Discussion

Seckel syndrome is defined by four criterias: intrauterine growth retardation below two standard deviations, microcephaly with cranio-synostosis, and facial dysmorphism with characteristic bird headed appearance prominent beaked triangular nose, variable mental retardation.

The variability of expressivity of these criteria explains the heterogeneity of this syndrome especially as they are often associated with inconsistent clinical signs affecting the central nervous system, axial skeleton, limbs and integuments. [4]

IV. Conclusion :

Seckel syndrome is a genetic disorder with autosomal recessive inheritance. Its enthiopatogeny remains unclear. We wanted to share our case since no cas of bowel malrotation have been reported in association with this syndrome

Références:

[1]. Kjaer I, Hansen N, Becktor KB, Birkebaek N, Balslev T. Craniofacial morphology, dentition, and skeletal maturity in four siblings with Seckel syndrome. Cleft Palate Craniofac J. 2001;38:645-51.
[2]. precocious puberty associated with nonclassical congenital adrenal hyperplasia. J PediatrEndocrinolMetab 2004; 17: 105-10.
[3]. hayani A, Suarez CR, Mclnar Z, LeBeau M, Godwin J. Acute myeloid leukemia in a patient with seckel syndrome. J Med Genet 1994;31:148-9
[4]. P Parent, S Moulin, MR Munck, L de Parscau, D Alix .Le nanisme à tete d'oiseau ou syndrome de Seckel.Difficultésnosomogiques .Arch Pediatrie 1996 ;3 :55-62

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