ISSN: 2634 - 680X

Journal of Clinical Case Studies, Reviews & Reports

SCIENTIFIC Research and Community

Case Report

Open Access

Idiopathic Megacaecum: about a Rare Case Report

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ABSTRACT

Megacaecum is a non-commonly observed and very difficult to diagnose condition. The pathogenesis is yet to be fully understood as there are very few cases documented in the literature, we report the case of a 23-year-old woman, who was suffering from a sub-occlusive syndrome along with an abdominal and pelvic pain, and in whom the diagnosis was determined by an immediate abdominopelvic CT scan. The aim of this article is to report an additional case to the literature and to highlight the pathogenesis, clinical and radiological findings, along with the potential treatment of this particular diagnosis.

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Received: November 10, 2021; **Accepted:** November 17, 2021; **Published:** November 24, 2021

Introduction

Megacaecum is uncommon and poorly known. It is a rare congenital disorder that can be diagnosed in adults. Its pathogenesis is little known as there are only very few cases reported in the literature: It is possible that it is due to an anomaly of the enteric nervous system where there is an abnormal number of neurons (hyperganglionosis, hypoganglionosis or aganglionosis) or neurons with abnormal biochemical or physical properties [1]. Only nine cases have been documented in the literature making it an exceptional and probably underdiagnosed medical condition [2, 3].

In this article, we report an extra case to the literature and shed light on the pathogenesis, the clinical and radiological findings as well as the potential treatment of this particular diagnosis.

Case report

We report the case of a 23-year-old female patient with no particular medical history who presented to the adult emergency department with diffuse abdominal and pelvic pain, more pronounced in the left iliac fossa and a sub-occlusive syndrome; consisting of obstruction of passage of fecal matter and of gas, without emesis, evolving for less than 24 hours, symptoms seemed to be relieved by supine positions and the patient later reported the spontaneous resumption of the bowel transit.

Physical examination showed a supple, non-distended abdomen and a diffuse abdominal sensibility with an empty rectal ampulla.

Laboratory investigations were unremarkable. An enhanced abdominal CT scan was performed (Figure 1,2,3&4) and showed the appearance of a megacaecum descending in the pelvis, with significant aerial distension and fecal stasis, with no evidence of mechanical or functional obstruction distally.

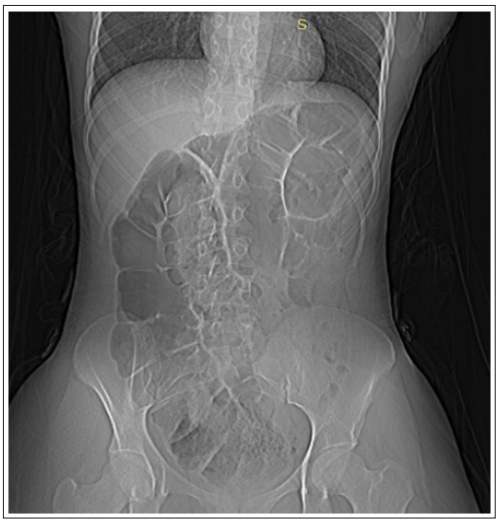

Figure 1: Scout view showing a significant aerial distension of the caecum and the right colon

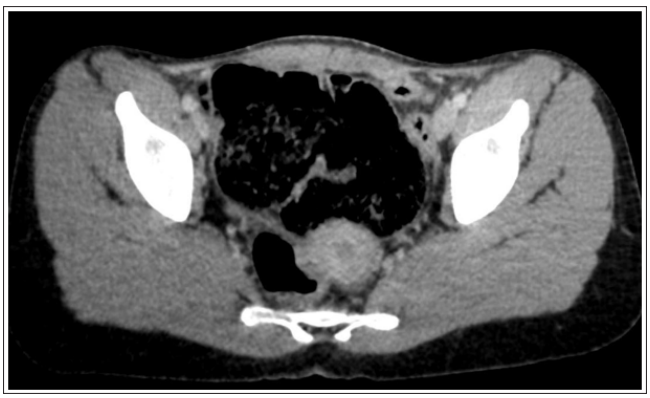

Figure 2: Enhanced abdominal CT scan (axial) showing a megacaecum descending in the pelvis, with significant aerial distension and fecal stasis, with no evidence of mechanical or functional obstruction distally

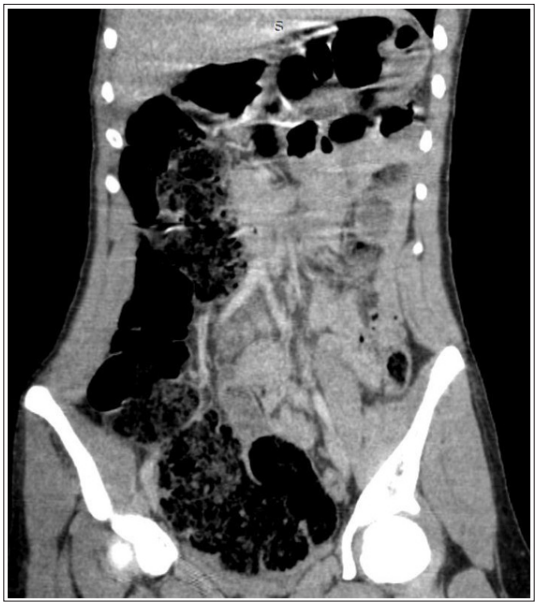

Figure 3: Enhanced abdominal CT scan (coronal) showing a megacaecum descending in the pelvis, with significant aerial distension and fecal stasis, with no evidence of mechanical or functional obstruction distally

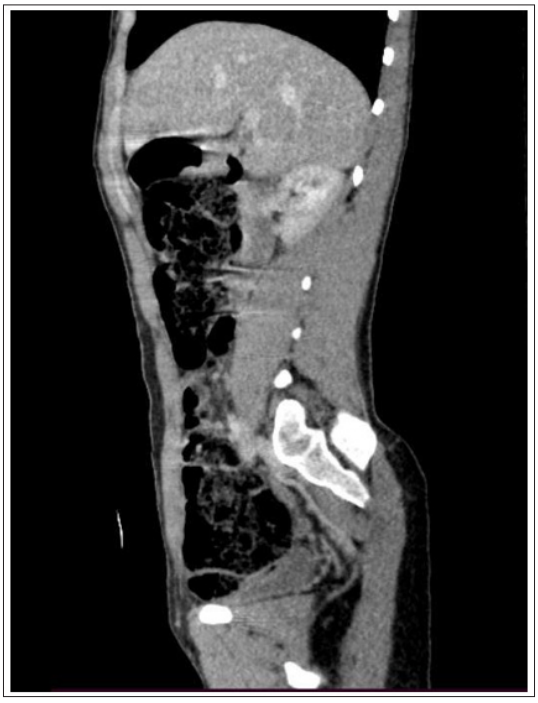

Figure 4: Enhanced abdominal CT scan (sagittal) showing a megacaecum descending in the pelvis, with significant aerial distension and fecal stasis, with no evidence of mechanical or functional obstruction distally

Discussion

Megacaecum is an extremely rare congenital condition that is very difficult to diagnose and manage, our case represents the tenth case to be reported in the literature. This condition is most often encountered in children. The revelation was delayed in our patient. The sub-occlusive syndrome that was the subject of this observation could have been the consequence of an intermittent volvulus of a dilated or mobile cecum [4]. As for abdominal and pelvic pain, it may be explained by distension of the cecum due to significant fecal stasis; When the pressure and volume of the materials becomes too excessive, the cecum tends to plunge into the pelvis. This leads in turn to a stretching of the mesentery connected to the cecum and causes pain. Intensification of the symptomatology could be attributed to an acceleration of the transit by the visceral reflex of the cecum, which is loaded but has trouble evacuating.

The actual pathophysiology is not fully understood. It may relate to an abnormality of the enteric nervous system involving an abnormal number of neurons (hyperganglionosis, hypoganglionosis, or aganglionosis) or neurons with abnormal biochemical or physical properties. In either situation, the clinical manifestations are very similar with varying degrees of colon obstruction based on the extent of the affected colon tract [5, 6].

Several types of mutations were identified as being involved in megacaecum that were distinct from those found in Hirschsprung's disease. Certain authors have mentioned the possible association of megacaecum in patients taking folliculin with certain hormonal disturbances found in certain neuropsychiatric conditions [7]. It has also been observed in association with other neurological malformations such as Spina Bifida [8].

The diagnosis of megacaecum is morphological. The entero-MRI is the exploration of preference, but in our case an abdominal CT scan was performed urgently in view of the clinical presentation of the patient and in view of the unavailability of MRI in the emergency room, Entero-MRI highlights the whole colon and allows to identify easily the megacaecum plunging into the pelvis. This exploration also allows to exclude other potential etiologies for chronic abdominal pain such as abdominal tumors, internal hernias and inflammatory diseases.

To radiologically affirm that a megacaecum is idiopathic, a distal obstruction must be ruled out. Colonoscopy can be used to rule out stenosis and anorectal manometry to rule out Hirschsprung's disease, which would result in distension of the entire colon including the cecum.

The most commonly performed treatment method documented in the literature is a laparoscopic right hemicolectomy [9, 10].

Conclusion

Only few cases have been documented in the literature, the physiopathology is not yet well understood, the clinical symptomatology is often silent and atypical, delaying the diagnosis to a stage of complication.

Imaging plays an essential role in confirming the diagnosis and in excluding differential diagnoses, and the treatment is exclusively surgical.

Disclosure of interest: The authors declare that they have no conflicts of interest concerning this article.

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