

Hypertrophic stenosis of the pylorus with findings of ankyloglossia: case report

Estenose hipertrófica do piloro com achados de anquiloglossia: relato de caso

Estenosis pilórica hipertrófica con hallazgos de anquiloglosia: reporte de caso

RESUMO

A Estenose Pilórica Hipertrófica Infantil ou Estenose Hipertrófica de Piloro é caracterizada por um adensamento incomum dos músculos pilóricos do estômago. Já a anquiloglossia, é uma condição que altera a função da língua devido à presença de freio encurtado. Objetivo: A descrição de um caso de estenose pilórica hipertrófica infantil associado à anquiloglossia, evidenciando os achados relevantes para o diagnóstico e tratamento. Método: A coleta de dados envolveu informações relatadas pela genitora e avaliação completa feita por pediatra devidamente registrada no Conselho de Medicina. A associação das patologias, juntamente com às barreiras no acesso, principalmente organizacional, exibiu uma dificuldade diagnóstica, devido a seus sinais desencadearem uma semelhança sintomática como êmese pós-prandial e deglutição comprometida. Resultado: Após o tratamento cirúrgico evidenciou-se uma melhora considerável no quadro de desnutrição e desenvolvimento do paciente. Conclusão: A abordagem precoce, associada à atuação multiprofissional, é fundamental para o desfecho favorável do caso.

DESCRIÇÕES: Anquiloglossia; Estenose Pilórica Hipertrófica; Píloromiotomia; Refluxo Gastroesofágico; Estenose Pilórica Hipertrófica Infantil

ABSTRACT

Infantile Hypertrophic Pyloric Stenosis or Hypertrophic Pyloric Stenosis is characterized by an unusual thickening of the pyloric muscles of the stomach. Ankyloglossia, on the other hand, is a condition that alters the function of the tongue due to the presence of a shortened brake. Objective: The description of a case of infantile hypertrophic pyloric stenosis associated with ankyloglossia, evidencing the relevant findings for diagnosis and treatment. Method: Data collection involved information reported by the mother and a complete evaluation by a pediatrician duly registered with the Medical Council. The association of pathologies, together with barriers in access, mainly organizational, exhibited a diagnostic difficulty, due to their signs triggering a symptomatic similarity such as postprandial emesis and compromised swallowing. Result: After surgical treatment, there was a considerable improvement in the situation of malnutrition and patient development. Conclusion: The early approach, associated with multi professional action, is fundamental for the favorable outcome of the case.

DESCRIPTORS: Ankyloglossia; Hypertrophic Pyloric Stenosis; Pyloromyotomy; Gastroesophageal Reflux

RESUMEN

La estenosis pilórica hipertrófica infantil o estenosis pilórica hipertrófica se caracteriza por un engrosamiento inusual de los músculos pilóricos del estómago. La anquilosis, por otro lado, es una condición que altera la función de la lengua debido a la presencia de freno acortado. Objetivo: Descripción de un caso de estenosis pilórica hipertrófica infantil asociada a anquilosis, evidenciando los hallazgos relevantes para el diagnóstico y tratamiento. Método: La recolección de datos implicó información reportada por la madre y una evaluación completa por parte de un pediatra debidamente registrado en el Consejo Médico. La asociación de patologías, junto con barreras de acceso, principalmente organizativas, exhibieron una dificultad diagnóstica, debido a que sus signos desencadenan una similitud sintomática como la emesis postprandial y la deglución comprometida. Resultado: Después del tratamiento quirúrgico, hubo una mejora considerable en la situación de desnutrición y desarrollo del paciente. Conclusión: El enfoque temprano, asociado a la acción multiprofesional, es fundamental para el resultado favorable del caso.

DESCRIPTORES: Anquiloglossia; Estenosis Hipertrófica del Píloro; Píloromiotomia; Reflujo Gastroesofágico

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INTRODUCTION

There is a distinction in the definition of vomiting and gastroesophageal reflux (GER), the first being defined as a set of characteristics and signs such as coughing, nausea and effort to eliminate the contents, in contrast to cases of GER, in which the fluid passes spontaneously without any apparent signs, becoming pathological when it affects the quality of life of the neonate, such as growth and development problems, as well as food impairment and possible respiratory disorders and changes in circadian rhythm. ¹

Breastfeeding and gastroesophageal reflux, whether physiological or pathological, are closely linked, and may present classic signs of leakage (regurgitation) of breast milk, or major complications, requiring a careful and effective investigation; one of the main influences of this variant is evidenced in infants with alterations in the lingual frenulum, due to the sucking and swallowing function being in disagreement with the normality in patients with this irregularity, who end up swallowing a greater amount of air during breastfeeding, favoring the appearance of the GER. ²

When the baby has ankyloglossia, a condition that alters the function of the tongue due to the presence of a shortened, inelastic, thickened or positioned frenulum at the tip of the tongue, which exhibits an abnormal fixation of the tongue in the oral cavity, due to a membrane that should have undergone apoptosis in the embryonic period, there may be low production of breast milk by the mother, due to incorrect latching, causing fissures in the breasts, low

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neonatal weight gain, irritability and excessive crying of the baby, as well as choking, tiredness and fatigue of this ally to compensatory movements.

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Infantile hypertrophic pyloric stenosis (IHPS) is characterized by an unusual thickening of the pyloric muscles of the stomach, causing hypertrophy and hyperplasia of the longitudinal and circular muscle layers of the pylorus, generating occlusion of the gastric outlet. It is more common in males, with a male ratio of 4:1. ¹ Its first exhibition was made in 1888 by the Danish pediatrician Dr. Harald Hirschsprung. ² The condition usually begins in the first month of the child's life (3 to 6 weeks) and, in most cases, before 3 months of age, its presentation shows vomiting in non-bilious, irregular or postprandial "projectiles". Late diagnosis leads to complications such as dehydration and severe malnutrition with disordered serum electrolytes and an acid-base imbalance. IHPS is a rare disease with symptoms similar to other common diseases such as gastroenteritis, gastroesophageal reflux disease and milk allergy, so its evidence as a primary diagnosis becomes

difficult to verify.

The recommended test for the detection of hypertrophic pyloric stenosis is ultrasonography, due to its high sensitivity, specificity and easy execution, it is noteworthy that it is an imaging-dependent technique. Ultrasonography findings show changes when the pyloric canal length is >15mm and the pyloric wall thickness >3mm, may also include absence of gastric emptying.³ The treatment initially consists of rehydration and correction of electrolyte imbalances, according to the severity of the signs, after rehydration, the definitive treatment is performed, which is pyloromyotomy, reported in 1912 by Ramstedt,² in which the pyloric muscle is fractionated up to its submucosal layer, to relieve compression, allowing the stomach contents to flow into the duodenum, the surgery can be open or performed laparoscopically, at the surgeon's discretion. The operation has low morbidity and 4 hours after surgery the patient can resume eating.¹

The use of medications, perinatal factors and modifiable environmental exposures may be associated with the development of IHPS, another notable point is its familial incidence with a 20-fold increased risk among siblings, heritability between twins estimated at 87%, SNP heritability of the whole genome 30%⁴ and the co-occurrence of multiple genetic syndromes demonstrate a hereditary tendency in the disease.² A modifiable exposure factor that demonstrates a major influence on the development of hypertrophic pyloric stenosis is the choice made for early nutrition, and exclusive breastfeeding with breast milk is associated with a lower risk of developing IHPS compared to formula feeding,⁵ presenting a 4.6 times higher risk when compared, a plausible explanation displays the differences in the volume of feeding between breastfeeding and bottle feeding, as well as the rate of ingestion, both of which can affect the distention and emptying of the stomach. Emphasizing the investigative interest regarding hypertrophic pyloric stenosis and its possible factor associated with the development of ankyloglossia, aiming at a better clarification on the approach and

early treatment of this harm.

METHOD

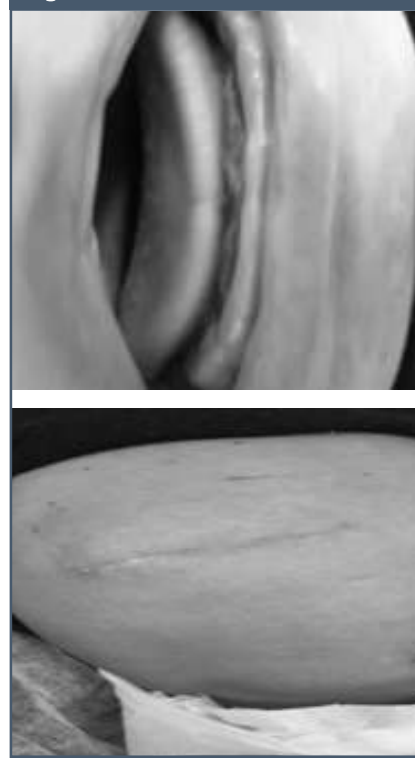
The observational and descriptive study was based on articles published on the platforms SciELO, Pubmed and the "Saúde Coletiva" journal (Barueri) for scientific basis and contextualization; data collection involved 1 sample, non-probabilistic for convenience, making the inclusion and exclusion criteria not applicable to the respective study, using data from medical records and complementary exams that elucidate the portrayed case, following guideline 510/16 as normative for the elaboration of the research.

RESULT

JGM 3 months old, with postpartum clavicular fracture, vaginal delivery, 40 weeks, weight 3,800g, height 50 cm, HC 33, BMI 15.37, mother seeks care center located in Paraná (South, Brazil) to receive childcare guidelines, feeding with breastfeeding and formula, reports bone rupture of the newborn's clavicle, being immobilized with the Velpau method, with improvement. Apgar score 8/9, umbilical stump not apparent, vitamin A+D prescription, awaiting Guthrie test results, weight gain 58 g/day. After 2 and a half months of birth, the child had signs consistent with gastroesophageal reflux disease, the mother reports performing a 30 degree decubitus position on the baby, which does not accept breast milk well, presenting malnutrition, depletion, wrinkled skin, face with dermatitis and changes in the lingual frenulum, demonstrating an inappropriate insertion from the tip of the tongue to the inferior alveolar ridge and limitations in the projection of the tongue (extension and retraction), causing difficulty in swallowing, being hospitalized for 13 days under intravenous hydration, but without any improvement in relation to jet vomiting, favoring the intensification of the condition.

Urgently requested the Ultrasound (USG). USG report shows liver, gallbladders, pancreas, spleen, adrenals, kidneys,

Figure 1: Ultrasound



Source: author.

retroperitoneum and bladder with normal aspects, highlighting abnormality in the pylorus, with findings of thickened muscle wall, measuring 7 mm, pyloric canal length of 18 mm and pylorus volume of approximately 2.4 ml, demonstrating to be altered, being compatible with the suspicion of pyloric stenosis together with alterations similar to the appearance of ankyloglossia, exposed and mentioned factor due to the difficulty in sucking breast milk, rejection of the breast, and the difficulty in carrying out the extension and retraction of the tongue, favoring the severe malnutrition found in the case.

DISCUSSION

Infantile hypertrophic pyloric stenosis (IHPS) usually presents between the second and fifth week of life, in rare situations after the 12th week, as in the case reported, its origin is still unclear, and multiple distinct etiological factors are suspected. The use of ultrasound (USG) is the common

technique for diagnostic confirmation, findings such as muscle thickness greater than 3 mm, pyloric canal length greater than 15 mm are confirmatory for IHPS. 7 Symptomatic compatibility of IHPS can be evidenced in other comorbidities such as: gastroenteritis, hernia, appendicitis, making the primary diagnosis difficult to establish. 1

Classic signs of IHPS depict recurrent postprandial jet vomiting, difficulty in sucking breast milk and weight loss, in babies with some secondary alterations such as lingual frenulum alteration, even ankyloglossia, and the symptoms and signs can be atypical or directed to other more recurrent diseases, and emesis can be in a lower incidence without projectile presentation, with an increase in appetite and excessive gastric peristalsis. 7

The patient's condition showed a strong coincidence with findings of ankyloglossia, later confirmed, being an anomaly characterized by a short lingual frenulum or with limited movement due to its insertion, which can show the tip of the tongue inserted in the inferior alveolar ridge, being a partial and more commonly usual condition, or the integral fusion of the floor of the mouth constituted by the geniohyoid and mylohyoid muscles, exhibiting an extremely rare and delicate condition, constituting a refusal to breastfeed, difficulty in swallowing and frequent vomiting due to the vacuum that forms between the correct fixation of the infant's mouth and the nipple of the mother's breast, generating air ingestion causing recurrent regurgitation and frequent colic. 8 The use of complementary formulas instead of breastfeeding, as evidenced in the clinical case, has demonstrated a risk factor for the development of pyloric stenosis, emphasizing the conduct of indication of breastfeeding as a protective factor for numerous diseases. 6

The medical treatment is usually done surgically, being submitted to pyloromyotomy and for the abnormality in the frenulum joint frenotomy was performed, the surgeries show a low morbidity rate. Postoperative care indicates the introduction of intravenous fluids until the infant can tolerate feeding, with mild post-surgery

regurgitation being normal, stopping until the 5th day after the procedure. 6

The postoperative approach by the speech therapy service would be ideal, as it would help in the correct diagnosis, assist in immediate breastfeeding (preventing early weaning), rehabilitate suction, assist in healing and prevention of relapses, ensuring optimal guidance to parents. An excellent way to assess the improvement of ankyloglossia after frenotomy would be to assess the way the baby sleeps; being ideal the mouth closed and the tongue on the roof of the mouth, that is, touching the hard palate. It is noteworthy that the frequency with which the patient was taken to health centers and primary care centers helped in the initial diagnosis and continuous early intervention, highlighting the importance of access to children in primary care, especially in the first years of life, where the continuity of the process reaches degrees of progressive evaluation of the attending professional, verifying their physical and mental development, suitable for each stage of life, and it is up to the same to indicate the possible failures that witness in the process of child growth, as well as their conduct and ways to improve the prognosis. 11

CONCLUSION

The case report made it possible to identify some aspects consistent with the diagnosis of infantile hypertrophic pyloric stenosis, highlighting its difficult initial finding, among the most common findings are characteristics such as post-breastfeeding jet vomiting, rejection of the mother's breast or difficulty in sucking milk, leading to severe malnutrition; in accordance with the findings, there was still a need for intervention in the lingual frenulum, which showed irregularity in its fixation, consistent with findings of ankyloglossia, being a major aggravating factor in the general condition. There was a significant improvement after the surgical intervention, showing a progressive and optimized recovery, favoring the child's development according to the growth curves.

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necessary to act in a multiprofessional way with the active participation of the nurse. In addition, it is understood that there needs to be an empathic, longitudinal and integral bond between health professionals

and families. The contributions of the study are to alert other health professionals and parents about the importance of early diagnosis and treatment of childhood hypertrophic pyloric stenosis and findings

of ankyloglossia, emphasizing the need for more research and data comparisons, evidencing an investigative character of the disease, favoring the dissemination of practical knowledge. ■

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