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Kyrina Anastasia et al., J Pediatr Care, Volume 4 DOI: 10.21767/2471-805X-C1-006

DIFFUSE TENDER EDEMA AND INTUSSUSCEPTION: AN UNUSUAL INITIAL PRESENTATION OF HENOCH-SCHONLEIN PURPURA

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An 8-year old boy was admitted with a 72 h history of painful Aswelling of the upper and lower limbs. The oedema had been worsening within the last 24 h and he refused to walk and also reported non-focal abdominal pain that begun 4 days ago. He had no remarkable history apart from a drop off his bicycle 4 days ago after which he was examined in the emergency department and no serious trauma were found. Physical examination revealed painful pitting edema of the limbs along with periorbital oedema, scrotal swelling and a small non-blanching rash on the limbs and on the abdomen that was barely seen. Laboratory assessment showed mild thrombocytosis, increased ESR and decreased serum albumin. He started complaining of severe abdominal ache and subsequently, an abdominal ultrasound was performed and showed small bowel intussusception that was also confirmed by CT. Ninety six hours after the initiation of the edema, palpable purpuric lesions indicative of Henoch-Schonlein purpura (HSP) were appeared on the boy's legs. Therefore, the

patient was administered steroids and showed remarkable clinical improvement. Facial edema in HSP have been previously described mainly in children less than two years old, in this case not only the primary edema was prominent and involved multiple body parts, but also it was spread all over his body. Non-typical, subcutaneous edema involving one or multiple parts of the body represents a severe feature of HSP that may precede the purpuric rash. Thus, pediatricians should be aware of this possibility and should always have a high index of suspicion for HSP.

Biography

Kyrina Anastasia has completed her Medical studies from National and Kapodistrian Athens University. She has worked as a Pediatric Trainee at General Hospital of Kozani in Greece. She has done her neonatal rotation at Chelsea and Westminster Hospital in London. She is now a Clinical Fellow in Pediatrics and Neonates at Countess of Chester Hospital in Liverpool.

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TWO LIBYAN SIBLINGS WITH BETA-KETOTHIOLASE DEFICIENCY: A CASE Report and review of literature

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eta-ketothiolase (mitochondrial acetoacetyl-CoA thiolase, T2) Beta-ketotiliolase (introchonana, accessive disorder characterized by impaired metabolism of ketones and isoleucine. In this study, we report on the first two siblings with T2 deficiency from Libya. Both siblings presented with ketoacidosis, but the severity and outcomes were quite distinctive. T2 deficiency in patient 1, the younger sister, manifested as recurrent severe episodes of ketoacidosis during the first year of life. She unfortunately experienced neurodevelopmental complications and died at 14 months old, after her 5th episode. In contrast, patient 2, the elder brother, experienced only one ketoacidotic episode at the age of 4 years. He recovered uneventfully and has continued to achieve age-appropriate development to date. Upon analysis, the siblings' blood acylcarnitine profiles had shown increased levels of C5:1 and C5-OH carnitine. ACAT1 mutational analysis revealed patient 2 is homozygotic for a novel mutation-c.674C > A (p.Ala225Glu); this mutation was then confirmed by familial analysis. Transient expression analysis of c.674C > A mutant T2 cDNA revealed neither potassium ion-activated acetoacetyl-CoA thiolase activity, which represents T2 activity, nor mutant T2 protein. Therefore, this mutation is truly pathogenic. Interestingly, the incidence of T2 deficiency may be high among the Arab population. This disease should be considered in the differential diagnosis for unexplained ketoacidosis in children. Patients with T2 deficiency could have a favorable outcome if diagnosed and treated early.

Biography

Hanna Alobaidy has completed her MB BCh from Faculty of Medicine, Alexandria University, Egypt in 1980 and Postdoctoral studies in Pediatrics from Karolinska Institute Stockholm Sweden, 1990. She is a Consultant of clinical pediatrics and inborn errors of metabolism. She is currently serving as Consultant Metabolist in outpatient clinic Alkhadra Hospital, Tripoli, Libya. She has committed to a career in Academic Medicine and has over 20 years of experience of undergraduate and postgraduate medical education in University of Tripoli Medical College. She has published papers in national and international peer-reviewed medical journals with over 25 citations. Her top clinical researches are: The 1st study and reference report about the "Pattern of metabolic disorders in Libya, long term experience in tyrosinemia type I with NTBC (17years) and Niemann-Pick C disease follow up".

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SUBCUTANEOUS EMPHYSEMA AND PNEUMOMEDIASTINUM SECONDARY TO H 1N 1 PNEUMONIA IN SAUDI CHILDREN: 2 CASE REPORTS

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Spontaneous Pneumomediastinum (SPM) is an innocuous Condition in children, which usually responds to supportive therapy. The incidence of H1N1 Influenza A infection in children is exceedingly rare and may culminates into spontaneous pneumomediastinum and subcutaneous emphysema. The association of H1N1 viral infection with air leak syndromes such as pneumomediastinum and subcutaneous emphysema has been reported from various centers. We came across with two such cases in our hospital and with a timely diagnosis and appropriate treatment, both were able to recover with an excellent outcome and without any significant complications. **Key words:** Subcutaneous Emphysema Pneumomediastinum H1N1 Pneumonia viral Pneumonia Influenza A

Biography

Ruba Abo Essa has completed her MBBS from King Khalid University 2011. She has Saudi board in pediatrics from Saudi commission for health specialties, Saudi Arabia, Nov 2015 and Arab board in pediatrics from Arab board of health specializations, May 2016. She is currently a pediatric senior registrar and the director of pediatric residency training program, Abha maternity and children's hospital. She has published many papers in reputed journals

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EARLY AND LATE OUTCOMES AFTER SURGICAL MANAGEMENT OF CONGENITAL VASCULAR RINGS

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Persistent respiratory or feeding problems in children may be associated with a congenital vascular ring. Surgical management is fairly standardized, but long-term outcomes are not well described. This study aims to investigate clinical presentation, surgical treatment, and risk factors for early mortality and late outcome. Our database revealed 62 surgically treated vascular ring patients between 1993-2014. Double aortic arch was the most common diagnosis (53%), followed by right aortic arch with aberrant left subclavian artery (39%). A Kommerell's diverticulum was present in 24 patients. Symptoms were mainly respiratory (89%) and feeding problems (32%). Median age at operation was 1 year. Median extubation time and hospital stay were 4 hours (interquartile range (IQR) 2-16 h) and 5 days (IQR 3.8-7.3 days). Mean follow-up was 7.8±5.8 years. Early mortality was 8% and was related to anatomical diagnosis, concomitant anomalies, and need for preoperative intubation. Freedom from residual symptoms at 1 and 6 months was 63% and 82% respectively. Freedom from inhalation therapy at last follow-up was 82% and was influenced by type of vascular ring and preoperative ventilation and dysphagia symptoms always

disappeared. Surgical relief of tracheoesophageal compression is commonly effective in vascular ring anomalies. Respiratory symptoms only persist in a minority of children. Patients with double aortic arch are at increased risk to remain symptomatic, necessitating supportive inhalation therapy lifelong or during intercurrent respiratory infections. Severe repetitive respiratory distress episodes during the first year of life must alert the pediatrician for this clinical entity, as most patients with a vascular ring present early.

Biography

Katrien Francois is a Congenital Cardiac Surgeon at the University Hospital Ghent, Belgium, and currently the Head of the Cardiac Surgery Department at the same hospital. She received her surgical training at the University Hospital Ghent, and congenital cardiac surgery training at Great Ormond Street Hospital for Sick Children, London. She obtained her PhD with a thesis on "Surgical palliation of the functionally univentricular heart". She has published more than 100 papers in peer-reviewed journals.

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DIAGNOSIS OF TUBERCULOSIS IN CHILDREN: CHALLENGES AND OPPORTUNITIES

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hildhood tuberculosis is an important public health problem in Cresource constrained settings, but continues to be neglected by physicians and policy makers. Diagnosis is particularly challenging in infants and children for many reasons. Children generally do not produce or expectorate sputum, making it difficult to obtain appropriate samples for analysis. Families generally do not collect and transport respiratory specimens properly. The disease is generally paucibacillary and mycobacteria are shed intermittently, reducing the yield (compared to adults). The hallmark radiological signs (such as pulmonary cavity) are rarely seen in childhood tuberculosis. Further physicians often treat children without confirming the diagnosis. The twin burdens of HIV infection and rising resistance among mycobacteria add further challenges to diagnosis. The mainstay of tuberculosis diagnosis rests on demonstration of acid fast bacilli (AFB) in biological samples (induced sputum, gastric aspirate/lavage, nasopharvngeal aspirate, lymph node aspirate, etc.). However staining with conventional methods yields results in only about 30% confirmed cases. Mycobacterial culture yield is also extremely low, but is somewhat improved by using liquid culture media. Clinical scoring systems have poor sensitivity and specificity; with limited diagnostic validity for treatment decisions. Radiological diagnosis rests on demonstration of one of three signs viz. hilar/ paratracheal lymphadenopathy, military shadows and a fibrocavitary lesion, but these findings are rare. Tuberculin test and

serological assays are two frequently misused tests. The former cannot distinguish infection from disease (hence has limited value in endemic settings) and the latter is unreliable and is discouraged by national and international guidelines. Even interferon gamma release assays (IGRA) have no value in endemic settings. Recent molecular diagnostic tests have raised hopes of better diagnostic platforms. The Xpert MTB RIF system (GeneXpert) is the most promising among these. A series of systematic reviews shows that GeneXpert is superior to microscopy, but inferior to culture (sensitivity ~60%, specificity >95%). This is a setback because although a positive test result is helpful to start treatment, a negative test does not rule out tuberculosis. Further, GeneXpert sensitivity is considerably lower in smear microscopy negative cases (compared to smear positive cases). However, a significant advantage is the rapid identification of rifampicin resistance. Other diagnostic techniques undergoing evaluation in children include: loop-mediated isothermal amplification (LAMP), and LED microscopy. However, children are excluded from the majority of global research studies on newer diagnostic platforms. In summary, diagnosis of childhood tuberculosis is difficult, and needs considerable time and effort. A step-wise approach can increase diagnostic confirmation in this difficult public health problem.

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EARLY AMPLITUDE-INTEGRATED EEG MONITORING 6H AFTER BIRTH Predicts long-term neurodevelopment of Asphyxiated late Preterm infants

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The present study aimed to assess the prognostic value of early amplitude-integrated electroencephalogram (aEEG) in late preterm infants who were born at a gestational age between 34 0/7 and 36 6/7 weeks for the prediction of neurobehavioral development. Late preterm infants (n = 170) with normal, mild, and severe asphyxia underwent continuous recording of aEEG for 4–6 h starting 6–8 h after delivery. The recordings were analyzed for background pattern, sleep-wake cycle (SWC), and seizures. Survivors were assessed at 18 months by neurological examination and Bayley Scales of infant development II. The incidence of adverse neurological outcome in the asphyxia group was significantly higher than in the normal group. For late preterm infants in the asphyxia group, abnormal aEEG pattern had a predictive potential of neurological outcomes with sensitivity of 78.57% (specificity 87.80%; positive predictive value [PPV] 68.75%; negative predictive value [NPV] 92.31%; power 85.45%). Non-SWC and intermediate SWC significantly were increased (25.45% and 52.73%, respectively) in the asphyxia group vs. the normal group. SWC pattern had neurological prognosis value in the asphyxia group with sensitivity of 64.29 % (specificity 87.80%; PPV 64.29%; NPV 87.80%; power 81.82%). Early aEEG patterns are important determinants of long-term prognosis of neurodevelopmental outcome in asphyxiated late preterm infants.

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THE ROLE OF THE MOTHER'S AND FATHER'S VOICE AND TOUCH IN IMPROVING NEONATAL TRANSITION AT BIRTH

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Transition from placental respiration to pulmonary respiration at birth is one of the most radical changes in both the circulation and the pulmonary function, changes which have to occur within a few minutes after birth if the baby is to survive intact. It is the moment when the mother can see, touch and speak directly to their newborn for the first time. The neonate can recognize the voice of its mother (and father) and these voices have an immediate calming effect. The maternal voice has been shown to be recognized by the newborn baby having heard it *in-utero* during the months before. Even before birth, processing of the maternal voice is apparent within the temporal cortex of the fetus. After birth, it is no surprise that the mother's voice can quickly calm a crying baby. The father's voice can also be recognized if the

fetus has been exposed sufficiently during pregnancy. Emotional closeness through vocalization and touch are important in the future parent-infant relationship and may have beneficial effects on the recovery of the neonate. Nothing can be more reassuring than the mother's voice. When the neoante is born asphyxiated, ventilation is a priority. This is carried out away from the mother, but motherside resuscitation, as opposed to the traditional room side resuscitation, allows the mother to speak to and touch her newborn baby and aid the recovery. Equipment is now available to allow this approach. With the neoante by the mother the placental circulation can remain intact.

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PREVALENCE OF NOCTURNAL ENURESIS AND RELATED FACTORS IN CHILDREN AGED 5-13 IN ISTANBUL

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Background & Aim: Enuresis (nocturnal eneuresis) is a health problem frequently encountered in childhood and refers to involuntary urination into the bed during sleep that occurs at an age at which bladder control would be expected. This study was carried out in two socio-demographically different districts of the province of Istanbul, Turkey, among children ages 5-13, for the purpose of determining the relationship between the incidence of nocturnal enuresis and certain demographic characteristics.

Method: The study design is descriptive and relational. The research was conducted at two health centers in two districts in the province of Istanbul that are thought to exhibit different socio-demographic characteristics. Data was collected by the researchers by means of a questionnaire.

Findings: Enuresis was a complaint expressed by 16.2% of the

cases in the study group; 8.3% reported intermittent bedwetting. The data collected in the two districts pointed to a significant difference in terms of the frequency of enuresis in favor of the district where socio-demographic features were inferior (p<0.005). When family histories were explored in cases of children with enuresis, it was found that the mothers of 76.2% had the problem of enuresis while 14.9% had enuretic fathers. Thus statistically, the presence of enuresis in the family history was seen to have had a markedly significant impact on the occurrence of enuresis in the child (p<0.00).

Conclusion: It was concluded that familial predisposition to the condition constituted a more significant risk factor for enuresis compared to socio-demographic or economic characteristics.

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TUBERCULOSIS IN CHILDREN IN A HOSPITAL IN THE SUBURBS OF DAKAR, SENEGAL

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Introduction: Tuberculosis remains a major public health problem in Senegal despite all the means used to deal with this disease. In an attempt to strengthen tuberculosis control in children, it was envisaged to undertake this study, the main objective of which was to describe the epidemiological, clinical and evolutionary profile of tuberculosis cases, all types taken together, in children monitored at the Centre hospitalier Roi Baudouin de Guédiawaye.

Materials & Methods: A descriptive retrospective study was carried out from 1 January 2010 to 31 December 2014 in the Centre hospitalier Roi Baudouin de Guédiawaye located in the Dakar suburbs. Tuberculosis cases diagnosed in children during the reporting period were compiled from patient records specially designed for this purpose. The data was entered on Excel and the analysis was done using Stata IC 12 software.

Results: A total of 121 new cases of tuberculosis were reported in children during the study period. The average age of the children was 10.84 years. The study population was predominantly female (52.89%). Pulmonary location was more frequent with 70.25% of cases. Extra pulmonary locations were mainly represented by lymph node (30.55%), osteoarticular (16.67%) and pleural (13.89%) forms. The bacilloscopy was positive in 47.93% of the cases. Tuberculosis was associated with HIV in 14.05% of cases. The evolution was favorable in the majority of cases and the hospital mortality was 4.96%.

Conclusion: Given that childhood tuberculosis is still common and mortality high, there is need for further studies to identify the factors contributing to the fact that this epidemiological situation remains unchanged.

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LIFE HABITS IN CHILDREN WITH CEREBRAL PALSY

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Objective: Since according to the International Classification of Functioning, Disability and Health (ICF) participation is the ultimate outcome for individuals with disabling conditions, children with cerebral palsy are at risk of reduced participation in everyday activities. Purpose of this study is review of life habits according ICF in children with cerebral palsy.

Materials & Methods: In this cross sectional study 53 children with cerebral palsy (5-13 years old) from rehabilitation centers were selected via the non-probability convenience sampling. The quality of accomplishment of life habits and the overall satisfaction with performance were assessed via Life-H Questionnaire through interviews with parents. Life habits divided into 12 categories. The categories are nutrition, fitness, personal care, communication, housing and mobility refers to daily activities; the categories responsibilities, interpersonal relationships, community life, education, employment and recreation address social roles.

Results: The main effect of gender was not statistically significant in any of the variables of life habits. Significant differences were found between gross motor function classification system (GMFCS) and global score of series of life habits and most of life habits' variables except work and community life. There were significant differences between global score manual ability classification system (MACS) and most of variables of the Life-H test except interpersonal relationships, community life, and work. The relevance between satisfaction with performance and accomplishment of life habits was significant.

Conclusion: There was not significant relevance between gender and accomplishment of life habits but GMFCS and MACS levels had significant relevance with accomplishment of life habits.

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BREASTFEEDING AND PHYSICIAN MOTHERS -RESULTS OF AN INTERNATIONAL SURVEY

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We report data from 581 physician mothers contacted through the Academy of Breastfeeding Medicine, using an online questionnaire. Mean exclusive breastfeeding duration was 4.2 months (standard deviation of 2.3, range 0-12) and mean breastfeeding duration was 12.3 months (SD 7.3, range 0-54). The participants intended to breastfeed 68% of their 947 children for at least 12 months. While 98% of infants were breastfed at birth, only 58% continued to receive breast milk at 12 months. Variables that had a statistically significant association with breastfeeding duration after controlling for other covariates were maternal goal for breastfeeding duration, length of maternity leave, and maternal perception of availability of time to express milk at work after return to employment postpartum. Duration of exclusive breastfeeding was negatively associated with maternal age at the time of the study and positively with perception of availability

of time to express milk at work after return to employment postpartum. Being in practice at the time of childbirth was associated with longer duration of exclusive breastfeeding, compared to being in training. Personal exclusive breastfeeding duration had a statistically significant association with selfreporting breastfeeding promotion among female housestaff (residents and/or fellows), colleagues, staff, or students as well as frequency of breastfeeding discussion with pregnant patients or new mothers. Our findings suggest that maternal infantfeeding intentions and work-related factors play an important role in infant-feeding behavior of physician mothers. Longer maternity leave as well as protected time at work for breastfeeding mothers might result in significant improvement in their breastfeeding duration.

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NICE OR NOT SO NICE? AN AUDIT OF EARLY ONSET NEONATAL SEPSIS

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Introduction: NICE guidelines for the management of early onset neonatal sepsis were published in 2012. Aneurin Bevan University Health Board revised local guidelines to incorporate NICE recommendations in February 2016. Our aims were to compare our practice pre and post new guideline.

Method: The audit was undertaken over three months during 2016 in Royal Gwent (RGH) and Nevill Hall (NHH) Hospitals. All babies on the postnatal ward commenced on antibiotics during the first 72 hours were included.

Results: Eighty eight babies received antibiotics; 68 at RGH and 20 at NHH. In 59 cases (67%), the indication for antibiotics was maternal sepsis. In RGH 37 babies (54%) had >1 CRP <5 and were discharged day 2. In NHH 12 babies (60%) had >1 C-reactive protein (CRP) <5 and 2 (10%) were discharged day 2. Eighteen

babies (20%) had at least 1 CRP >30 and had a locked plating of which 14 (78%) were successful and none were positive. All blood cultures were negative. In RGH the mean duration of hospital stay was 3.4 days with a mode of 2.0 days and median of 2.0 days. In NHH the same figures were 4.0, 3.0 and 3.0 days respectively. 22.7 babies per month received antibiotics; this pre NICE figure was 10.4.

Conclusion: We have shown an increase in babies receiving antibiotics following implementation of NICE recommendations. In NHH hospital stay is longer and a contributing factor is off site blood culture processing.

Recommendation: With the current NICE guideline the burden of postnatal ward antibiotic babies remains high.

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BRONCHIOLITIS GUIDELINE CHANGES AND EFFECTS IN A PEDIATRIC Emergency department

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In 2014 the American Academy of Pediatrics revised prior 2006 guidelines on bronchiolitis treatment. This study aimed to compare outcomes in the emergency department (ED) in a major academic medical center pediatric emergency department after implementing new recommendations, specifically length of emergency department stay and time to decision to admit. Secondarily the use of treatment and diagnostics were compared pre and post implementation of guidelines. A retrospective review of infants under 12 months of age that presented to our pediatric tertiary center's academic ED from 2012-2016 bronchiolitis seasons was performed. Winter of 2015 is when the ED and pediatric department made a combined effort to strictly follow the 2014 AAP guidelines. Those with a diagnosis of bronchiolitis were included. Previous reactive airway disease and wheezing patients were not excluded for this study. Data collected included time of placement in a room in the ED to time of discharge or time to pediatrics consult (for admission). Other data collected for each patient included who received a chest x-ray (CXR), nebulizer treatment, RSV PCR, or any form of steroid. There was no significant difference in the ED length of stay, nor in the time-todecide for admission of patients after the 2014 AAP bronchiolitis guidelines were applied. There was a significant reduction in the overall number of treatment and diagnostic modalities used postimplementation suggesting a potential cost-effective strategy of care.

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ENTEROCUTANEOUS FISTULA IN NEONATES AND CHILDREN: THE ROLE OF OCTREOTIDE AND SOMATOSTATIN IN THE CONSERVATIVE MANAGEMENT

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Enterocutaneous fistula in neonates and children is a very challenging condition, with a low incidence but a significant morbidity and mortality. Conservative management is the cornerstone of treatment, although the role of octreotide and somatostatin in the closure of the fistula still remains controversial. We present an extensive literature review together with a case vignette to illustrate the different aspects of the treatment with these drugs. The great diversity among published treatment regimens makes outcomes difficult to compare. Nevertheless,

given the results in the cases reported in the literature and in our patient, we suggest a possible beneficial effect of octreotide and somatostatin on closure of enterocutaneous fistula. It is necessary to perform a multicenter, double-blind, randomized, placebo-controlled trial to determine the efficacy of octreotide and somatostatin and the most suitable treatment regimen to optimize conservative management of this complex condition.

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PEUTZ-JEGHERS SYNDROME: AN UNUSUAL CAUSE OF RECURRENT Intussusception in a 7-year-old boy

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Introduction & Aim: Peutz-Jeghers syndrome (PJS) is an autosomal dominant inherited disorder characterized by intestinal hamartomatous polyps in association with mucocutaneous pigmentations. Although, intussusception has been reported as a well-known complication of PJS, recurrent intussusception as an alarming finding in a patient with normal gastrointestinal endoscopy is uncommon. A 7-year-old boy who had recurrent intussusception episodes and diagnosed as PJS with histopathologically after surgical excision of involved bowel segments is presented to discuss the clinical features and treatment options of recurrent intussusception as a presenting finding of PJS. We aimed to present this case to emphasize that patients with PJS may present with intussusception and hamartomatous polyps might be precancerous, in this syndrome.

Case Report: A 7-year-old boy was admitted to emergency with complaints of abdominal pain and vomiting lasting three days. He had a history of recurrent colicky abdominal pain for approximately six months. There was no history of hematemesis or melena. He had undergone upper gastrointestinal endoscopy and colonoscopy because of multiple hyperpigmented macules over only the lips and it is revealed normal endoscopic findings. His physical examination showed slight distension with generalized tenderness. According to physical examination findings and bowel obstruction, he underwent explorative laparotomy. A jejunojejunal intussusception was found 5 cm distal of the ligament of Treitz during surgical exploration. Intussusception could not reduce manually and a polypoid mass in the jejunal lumen was the leading point. The intussuscepting segment including the polyp was excised by small bowel resection, and end-to-end anastomosis was performed. Histopathological examination of the specimen revealed hamartomatous polyp and Peutz-Jeghers syndrome was confirmed histologically with low-grade dysplasia.

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Conclusion: Peutz-Jeghers syndrome is a hereditary disease characterized with gastrointestinal polyposis and skin or mucosal pigmentation. Polypoid lesions are almost hamartomatous and are not usually precancerous. Our patient underwent upper and lower endoscopy with presumptive diagnosis of PJS before the clinical findings of intussusception. Endoscopic evaluation revealed normal findings without any polypoid lesion. Although the association of PJS and intussusception is well-known, this complication is rarely presenting finding for PJS. Patients with hyperpigmentous lip lesions with recurrent intussusception episodes may require surgical explorations not only relieving small bowel obstruction but also diagnose the underlying polypoid lesions. In conclusion, patients with PJS should be regularly and closely monitored in order to reduce the number of laparotomies and due to the increased risk of cancer.

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