Case report

Mucocutaneous pigmentation as a diagnostic indicator in Peutz-Jeghers syndrome with progression to early malignant transformation

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Abstract
Introduction. Peutz-Jeghers syndrome is a rare inherited disorder characterized by mucocutaneous pigmentation and gastrointestinal hamartomatous polyps, associated with obstructive and hemorrhagic complications and an increased risk of malignant neoplasms. **Case presentation.** A 19-year-old man with mucocutaneous hyperpigmentation since childhood, recurrent abdominal pain, and chronic dyspeptic symptoms. Physical examination revealed hyperchromic macules on the lips, oral mucosa, palms, and soles, associated with multiple gastrointestinal polyposis, leading to a clinical suspicion of Peutz-Jeghers syndrome. Endoscopy revealed hyperplastic gastric polyps; biopsy showed hamartomatous changes without malignancy. **Treatment.** Conservative management with endoscopic surveillance and genetic counseling was initially instituted. One year later, he presented with acute abdominal pain, leading to jejunal resection. Histopathological examination confirmed well-differentiated (G1) multifocal invasive intestinal adenocarcinoma. He received adjuvant chemotherapy with oxaliplatin and capecitabine for six cycles. **Outcome.** The patient remains under multidisciplinary follow-up, with periodic surgical and endoscopic surveillance, and currently shows no evidence of disease progression. **Conclusion.** This case highlights the potential for early malignant transformation in Peutz-Jeghers syndrome and underscores the importance of close surveillance, timely diagnosis, and multidisciplinary follow-up.

Keywords
Peutz-Jeghers Syndrome, Polyps, Mouth Mucosa, Hamartoma.

Resumen
Introducción. El síndrome de Peutz-Jeghers es una enfermedad hereditaria rara caracterizada por pigmentación mucocutánea y pólipos hamartomatosos gastrointestinales, asociada a complicaciones obstructivas, hemorrágicas y mayor riesgo de neoplasias malignas. **Presentación del caso.** Hombre de 19 años con hiperpigmentación mucocutánea desde la infancia, dolor abdominal recurrente y síntomas dispépticos crónicos. El examen físico evidenció máculas hipercrómicas en labios, mucosa oral, palmas y plantas, asociadas a poliposis gastrointestinal múltiple, lo que orientó a sospecha clínica de síndrome de Peutz-Jeghers. La endoscopía reveló pólipos gástricos hiperplásicos; la biopsia mostró cambios hamartomatosos sin malignidad. **Intervención terapéutica.** Inicialmente se instauró manejo conservador con vigilancia endoscópica y asesoría genética. Un año después, presentó dolor abdominal agudo que motivó resección yeyunal. El estudio histopatológico confirmó adenocarcinoma intestinal invasivo multifocal bien diferenciado (G1). Recibió quimioterapia adyuvante con oxaliplatino y capecitabina durante seis ciclos. **Evolución clínica.** El paciente permanece en seguimiento multidisciplinario, con vigilancia quirúrgica y endoscópica periódica, sin evidencia actual de progresión. **Conclusión.** Este caso resalta el potencial de transformación maligna temprana en el síndrome de Peutz-Jeghers y subraya la importancia de la vigilancia estrecha, el diagnóstico oportuno y el seguimiento multidisciplinario.

Palabras clave
Síndrome de Peutz-Jeghers, Pólipos, Mucosa Bucal, Hamartoma.

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Pigmentación mucocutánea como clave diagnóstica en síndrome de Peutz-Jeghers asociado a transformación maligna temprana

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Introduction

Peutz-Jeghers syndrome (PJS) is a rare, autosomal-dominant inherited genodermatosis associated with germline mutations in the STK11/LKB1 gene, located on chromosome 19p13.3, which encodes a serine/threonine kinase with tumor-suppressor function involved in the regulation of cell growth, polarity, and energy metabolism.[1,2]

The clinical manifestations of PJS usually appear in early childhood and are characterized by the presence of dark brown to black, lenticular, mucocutaneous hyperpigmented macules, located predominantly on the oral mucosa, lips, perioral region, palms, and soles. These pigmented lesions are often the first clinical sign of the disease and may precede the development of gastrointestinal manifestations by several years.[3,4]

Peutz-Jeghers syndrome is associated with the formation of hamartomatous polyps distributed throughout the gastrointestinal tract, with the highest prevalence in the small intestine, particularly in the jejunum. Although these polyps are histologically benign, patients with PJS have a significantly increased risk of developing both gastrointestinal and extraintestinal malignancies.[5-7]

Diagnosis is based on the integration of clinical criteria, suggestive family history, and characteristic endoscopic findings, and can be confirmed by molecular genetic studies demonstrating mutations in the STK11 gene. Early identification is essential for implementing appropriate surveillance strategies.[1,5]

Major complications include intussusception, gastrointestinal bleeding, and intestinal obstruction secondary to large polyps. The prognosis is variable and depends largely on adherence to follow-up programs, as well as on the timely detection and treatment of associated neoplasms.[6-8]

Peutz-Jeghers syndrome is a rare disease, with an estimated prevalence of between one in 50 000 and one in 200 000 individuals, according to international reports.[3,7] In Europe, it is classified as a rare disease when it affects < 5 per 10 000 inhabitants. The variability in these estimates reflects methodological differences between studies and the low frequency of the disease. It is most common between the first and third decades of life.[7]

Although the diagnosis of PJS is based on clear clinical and histopathological criteria, the absence of a family history[9] and of the classic hamartomatous architecture poses a significant clinical challenge, as it can be confused with conditions such as Laugier-Hunziker syndrome.[3] This underscores the importance of international guidelines.[10,11]

This case report aims to describe the diagnostic approach and clinical course of a young patient with mucocutaneous pigmentation and gastrointestinal polyposis, with subsequent development of intestinal adenocarcinoma, in the context of clinical suspicion of PJS, in the absence of classic histopathological confirmation and family history.

Case Presentation

A 19-year-old male patient with a history of recurrent iron-deficiency anemia was being followed up in the hematology outpatient clinic due to suspected chronic bleeding of unknown origin. During management, he received 500 mg of oral ascorbic acid once daily, prescribed as an adjunct to optimize dietary iron absorption, while the diagnostic protocol was being completed.

The last hematological follow-up was performed in January 2024, with the etiology of the anemia not yet established. However, on February 29, 2024, the patient was referred to the dermatology department of the same hospital for evaluation of mucocutaneous hyperpigmented lesions.

During the targeted medical history, it was documented that the hyperpigmented macules on the lips, oral mucosa, palms, and soles had been present since birth, with a progressive increase in number and intensity during adolescence, particularly starting at age 17.

On physical examination, hyperpigmented (dark brown) macules ranging from 0.2 to 0.3 cm in diameter were observed on the lower lip (Figure 1) and oral mucosa (Figure 2), as well as hyperchromatic (brownish-black) spots on the palms of the hands and soles of the feet, with a higher concentration along the fingers and toes, which had a diameter of approximately 0.3 cm (Figure 3 and Figure 4).

The patient was referred to the gastroenterology department in March of the same year due to suspicion of PJS, based on the association between mucocutaneous pigmented lesions and a clinical presentation of gastritis-like dyspepsia lasting more than three months.

On March 23, 2024, an esophagogastroduodenal endoscopy was performed with biopsy sampling. The endoscopic report revealed the presence of chronic gastritis and hyperplastic polyps located in the pyloric region.

The initial histopathological examination of the samples described a mucosa with elongated foveolae, architectural distortion, and glandular irregularity. At the epithelial level, cells with pseudoglandular differentiation, cystic dilation of glands, and foci of papillary proliferation were observed (Figure 5A and Figure 5B). Additionally, moderate lymphoplasmacytic inflammatory infiltrate (Figure 5C) and edema in the lamina propria (Figure 5D) were reported. No characteristic "tree-like" or "branching" architecture was observed; nor was there evidence of metaplasia, dysplasia, or malignancy. Following

the initial evaluation, the patient did not continue follow-up within the institutional healthcare system, which limited timely clinical and endoscopic surveillance. This situation may be related to barriers to geographic access and continuity of care; one year later, the patient presented with severe abdominal pain, for which he sought care at a private healthcare facility near his home, where he received a diagnosis and surgical treatment after tumor lesions were identified.

Treatment

The patient underwent surgical resection of the affected jejunal segment. Histopathological examination of the surgical specimen confirmed a well-differentiated (G1) invasive intestinal adenocarcinoma with multifocal localization, identifying two tumor lesions measuring approximately 3 cm × 3 cm and 2.5 cm × 2 cm, respectively, with infiltration extending to the serosal layer but with surgical margins free of neoplasia.

Following oncological resection, he received outpatient adjuvant chemotherapy with a CAPOX (XELOX) regimen based on intravenous oxaliplatin (100 mg) and capecitabine (four 500 mg tablets per day), with two tablets taken 30 minutes after breakfast and two tablets 30 minutes after dinner for 14 days, followed by a rest period in each cycle, completing six cycles every 21 days administered on August 19, September 12, October 3 and 24, November 14 and December 5, 2025. According to the clinical documentation provided, starting with the third cycle (October 3, 2025), the dose was increased to 230 mg of oxaliplatin with the aim of achieving the optimal dose intensity based on the patient's body surface area and given adequate clinical tolerance.

Clinical Diagnosis

Although the histopathological finding of a hypepigmented polyp does not correspond to the classic hamartomatous lesion of PJS, this result does not rule out the diagnostic suspicion. The combination of characteristic mucocutaneous lesions and the presence of multiple gastrointestinal polyps suggests an atypical or incomplete presentation of the syndrome. Furthermore, the subsequent identification of multifocal intestinal adenocarcinoma in the jejunum strengthens this presumptive diagnosis, given the recognized increased risk of gastrointestinal neoplasms associated with PJS, even in the absence of classic histological confirmation during the initial evaluation.

Outcome

The patient was provided with detailed information about his condition, emphasizing its nature, possible complications, and the importance of clinical follow-up. Currently, he remains under follow-up by the Gastroenterology, Internal Medicine, Coloproctology, and Oncology departments for surveillance and comprehensive management. Regarding the cutaneous manifestations, these did not require specific treatment, given their benign nature and the absence of associated symptoms.

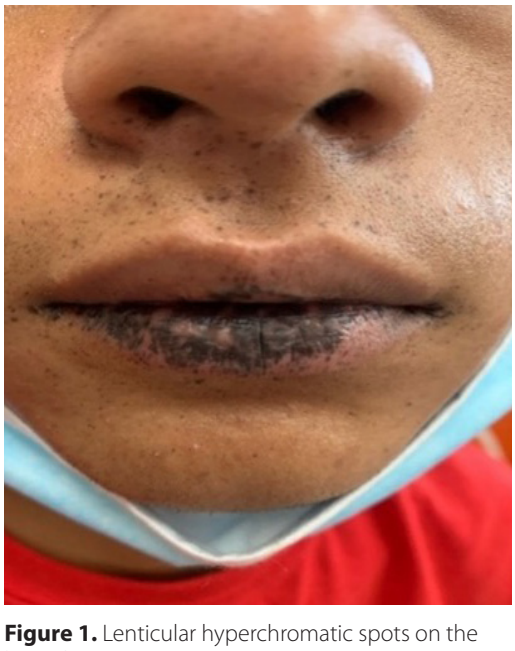

Figure 1. Lenticular hyperchromatic spots on the lower lip.

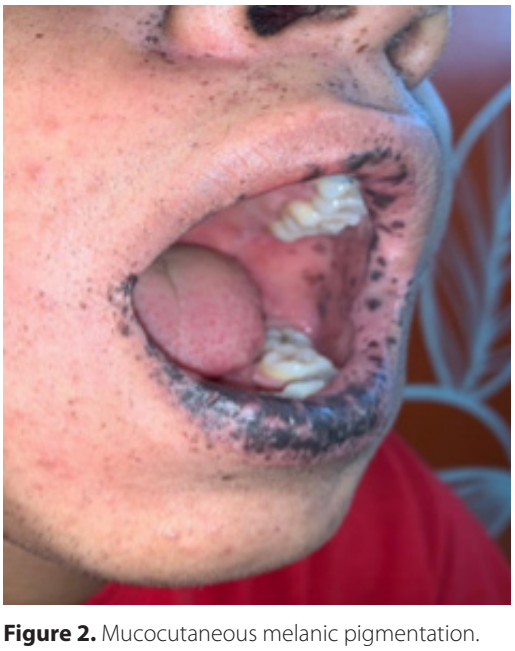

Figure 2. Mucocutaneous melanic pigmentation.

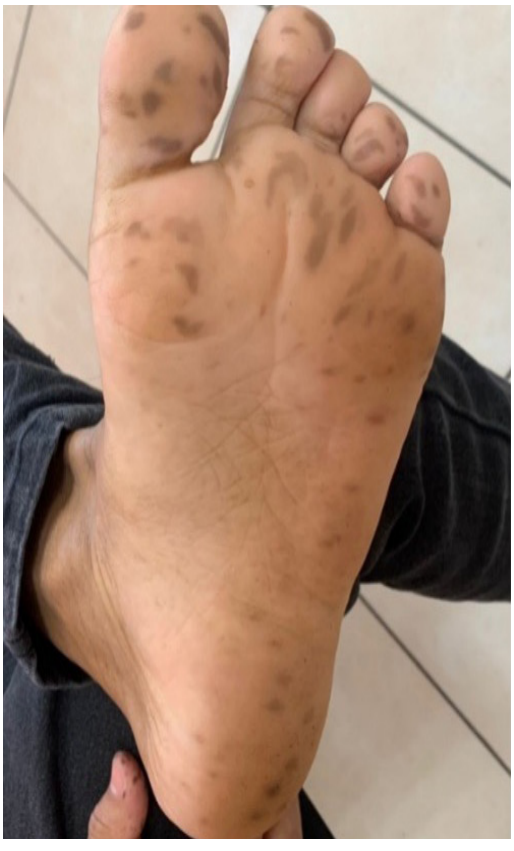

Figure 3. Melanin pigmentation on the soles of the feet.

Figure 4. Melanin pigmentation in both palms of the hands.

Discussion

Collectively, the clinical, endoscopic, histopathological, and follow-up data supported a diagnosis of PJS in the patient.

In this case, other differential diagnoses, such as juvenile polyposis syndrome, were ruled out, as it is not characterized by mucocutaneous pigmentation, although it presents with hamartomatous gastric polyps with cystic glands and inflammatory infiltrate[3,12] (Table 1).

Laugier-Hunziker syndrome constitutes a relevant differential diagnosis of PJS, due to the presence of mucocutaneous pigmentation; however, the absence of longitudinal melanonychia, a distinctive feature of Laugier-Hunziker syndrome, contributes to its diagnostic exclusion.[3,14]

Hereditary mixed polyposis syndrome was also ruled out, as this condition does not present with pigmentary abnormalities.[13] Furthermore, Cronkhite-Canada syndrome is an acquired condition characterized by inflammatory, sessile hamartomatous polyps in the stomach and colon. It is characterized by diffuse hyperpigmentation of the face, palms, and soles, accompanied by alopecia, nail dystrophy, and diarrhea, with a moderate risk of colorectal cancer (Table 1).

Peutz-Jeghers syndrome is an autosomal dominant condition that is rare in El Salvador, with limited information available. The available information comes primarily from isolated case reports at referral hospitals such as the Rosales National Hospital and the Benjamín Bloom National Children's Hospital.

In 2021, the guidelines from the International Society for Hereditary Gastrointestinal Tumors (inSiTGHT) and the European Hereditary Tumor Group (EHTG) were based on criteria such as the presence of two or more confirmed hamartomatous polyps, along with a positive family history and mucocutaneous pigmentation to establish the diagnosis.[10]

In 2024, the World Health Organization updated the diagnostic criteria for the syndrome, establishing that the diagnosis can be considered in the presence of three or more histologically confirmed PJS-typical polyps, any number of polyps in association with a family history, characteristic mucocutaneous pigmentation along with a family history, or the coexistence of polyps with mucocutaneous pigmentation.[3,10,15]

Although the patient did not strictly meet the classic histopathological criteria and denied a family history, it should be noted that PJS exhibits autosomal dominant inheritance with important nuances.[1,3]

Furthermore, the finding of multifocal invasive intestinal adenocarcinoma located in the jejunum provides additional evidence that supports the presumptive diagnosis of PJS, consistent with these patients' known predisposition to developing gastrointestinal neoplasms. Patients with PJS have an increased risk of extraintestinal neoplasms, particularly pancreatic, breast, and gynecological cancers (ovarian and cervical), as well as other less common sites, which justifies a comprehensive, multidisciplinary follow-up strategy throughout their lives.[5,6,10,13] In the

gastrointestinal tract, malignant transformation in PJS is predominantly associated with the development of adenocarcinoma, described as the most common histological type, generally through a dysplasia-to-carcinoma progression sequence. Other intestinal neoplasms have been reported, but less frequently.[1-5,6,13] Although malignant transformation typically manifests more frequently in later stages of life, its occurrence in young adults, as in this case, has been documented and highlights the importance of implementing early detection strategies and close interdisciplinary follow-up, particularly in the face of atypical phenotypes or incomplete clinical presentations.[5,6]

The literature reports that between 10 % and 20 % of patients affected by a de novo mutation or variable penetrance of the STK11 gene develop PJS.[3]

Consequently, in the absence of a positive family history, this case is classified under the suspicion of a spontaneous mutation or minimal clinical expression in the parents. It has been documented that manifestations may be subtle or absent in the ascending lineage.[3,10] Even in atypical or incomplete presentations, these patients remain at increased risk for neoplasms, thus requiring periodic endoscopic surveillance and specialized genetic evaluation.[1,3,10]

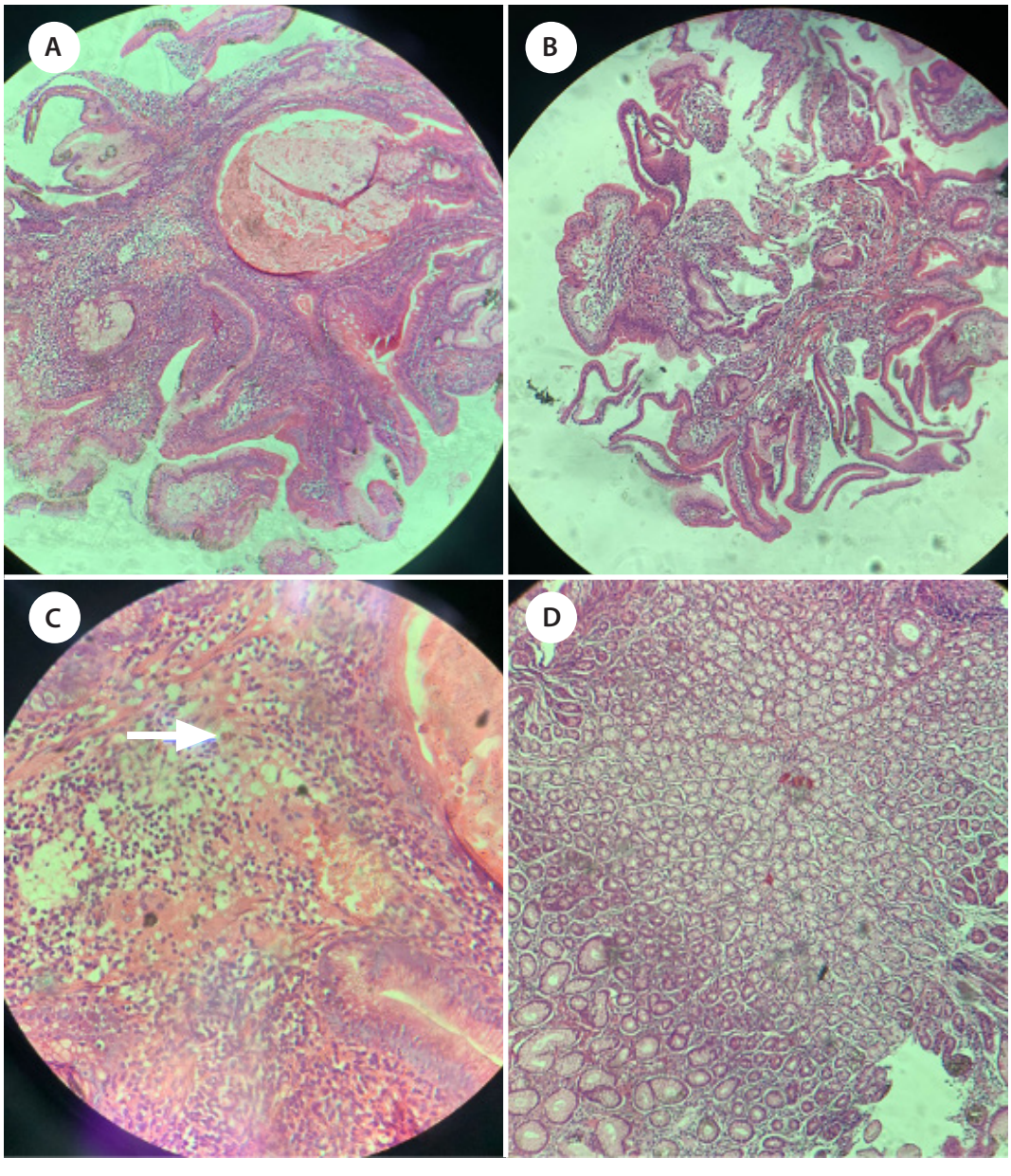

Figure 5. Gastric polyps, stained with Hematoxylin and Eosin (H&E). A. Ovoid-shaped polyp composed of dilated, cystic, irregular, and elongated glands lined by parietal, chief, and foveolar cells (4x light microscopy). B. Large-lumen dilated glands with elongations and foveolar distortion (10x). C. Inflammatory polyp formed by granulation tissue, presence of polymorphonuclear leukocytes (white arrow) in the lamina propria along with edema (40x). D. Glandular hyperplasia and inflammatory infiltrate of lymphocytes in the lamina propria (40x).

Table 1. Differential diagnosis of polyposis and mucocutaneous pigmentation.

Feature	Peutz-Jeghers syndrome (PJS) [1,13,14]	Juvenile Polyposis[3,12,13]	Laugier-Hunziker syndrome[3,14,15]	Cronkhite-Canada syndrome[10,12,13]
Types of polyps	Hamartomatous (with arborization of smooth muscle)	Hamartomatous (expanded lamina propria, without smooth muscle)	Absent (no associated polyposis)	Inflammatory and sessile hamartomatous
Location	Small intestine (most common), colon, and stomach	Predominantly in the colon and rectum	Not applicable	Stomach and colon (generally sparing the small intestine)
Pigmentation	Dark spots on lips, oral mucosa, and fingers	Generally absent	Macules on lips and mouth, and longitudinal bands on nails	Diffuse hyperpigmentation (face, palms and soles)
Hereditary pattern	Autosomal dominant (STK11 gene)	Autosomal dominant (SMAD4 or BMPR1A genes)	Sporadic (not hereditary)	Non-hereditary (acquired)
Cancer risk	Very high (gastrointestinal, breast, pancreas, gynecological)	Increased (Colorectal and gastric)	No increased risk (benign)	Moderate (Colorectal)
Other findings	Recurrent intestinal intussusceptions	Anemia, hypoproteinemia	Benign: diagnosis by exclusion	Alopecia, nail dystrophy, and diarrhea

Patients with PJS may present with other types of polyps, such as hyperplastic, adenomatous, or even inflammatory polyps.[3] Therefore, even if hyperplastic gastric polyps are described, this does not rule out PJS, since the pathognomonic finding is melanotic macules, and this is sufficient to raise suspicion, even in the absence of a characteristic polyp in the first biopsy.[3,10,14] Additionally, it has been reported that these polyps can coexist with neoplastic transformation processes, including small intestinal adenocarcinoma, which highlights the histological heterogeneity of the syndrome.[1,6]

As of 2026, no staging classification has been developed for PJS. Current guidelines are based on clinical, histological, and molecular criteria for diagnosis and management, not for establishing a disease stage.[10,11] These guidelines are followed in Central America and El Salvador.

The rationale for surveillance via colonoscopy and endoscopy[13] (beginning at age eight and continuing with biannual follow-ups in adulthood) is to prevent intestinal intussusception, which is the most common acute complication of PJS caused by polyps larger than 15 mm.[1,8,10] In this context, this case takes on special relevance, given that it was not possible to determine whether the finding of adenocarcinoma corresponded to accelerated malignant transformation in the context of PJS or to a neoplastic lesion undetected during the initial evaluation. Far from undermining the interpretation of the case, this diagnostic uncertainty underscores the importance of systematic and timely follow-up, not only to prevent mechanical complications but also to facilitate early detection of potentially malignant lesions and optimize clinical management.

In adults, the incorporation of enteric resonance imaging is critical, as it allows for evaluation of the small intestine, the preferred site for hamartomas and a blind spot for conventional endoscopy.[10,11] This preventive approach aims to transform the management of PJS from emergency medicine (laparotomies for obstruction) to a strategy of intestinal preservation and early detection of extraintestinal neoplasms (breast, pancreas, and gonads).[1,3,10,13]

In cases of clinical suspicion of PJS, where histopathological findings are inconclusive, and the family history is negative, genetic counseling plays a fundamental diagnostic and preventive role, since the absence of affected relatives does not rule out the condition but rather necessitates a thorough evaluation of parents and siblings to identify carriers with variable penetrance or minimal clinical manifestations;[3,10,13] in order to provide individuals and families with information about the nature, modes of inheritance, and implications of the condition to assist them in making medical decisions.[11]

In turn, management of the syndrome must be comprehensive and individualized, focusing on the early detection of polyps and cancers, as well as on providing psychological support and genetic counseling for patients and their families.[11]

Diagnostic limitations in El Salvador are significant; therefore, the approach has been adapted to local resources, prioritizing clinical diagnosis based on international criteria (EHTG/InSiGHT) and histological confirmation, in addition to periodic endoscopic surveillance in tertiary care hospitals, in order to prevent intussusception and bleeding.[10]

Given the high risk of oncological and mechanical complications associated with PJS, a therapeutic approach was adopted in line with the recommended management principles for this condition, optimized according to available resources and aimed at preventing adverse events through close clinical follow-up. In this context, the development of multifocal jejunal adenocarcinoma in a young patient, such as the one presented, constitutes a rare but clinically relevant manifestation that highlights the need for more rigorous surveillance strategies tailored to individual risk, even in scenarios where classic diagnostic criteria are not fully met.

In the context of small bowel adenocarcinoma, treatment is primarily based on surgical resection with clear margins, which is considered the treatment of choice. In patients with risk factors or locally advanced disease, the use of adjuvant chemotherapy based on regimens including oxaliplatin and fluoropyrimidines, such as CAPOX or FOLFOX, is recommended.

Management should be dynamically adapted to incorporate new evidence and innovative strategies in the prevention and treatment of its complications. Likewise, the development of registry and follow-up initiatives adapted to the Salvadoran context will allow for estimating the true burden of the disease, identifying asymptomatic familial cases, and evaluating the impact of follow-up programs on reducing cancer and other associated complications. Documentation by healthcare professionals is essential, as national protocols inspired by guidelines such as those of the European Society of Gastroenterology, Pediatric Gastroenterology and Nutrition (ESPGHAN), EHTG[10], or the American Gastroenterological Association (AGA), adapted to the country's diagnostic and therapeutic capabilities, could improve the quality and timeliness of care. This, combined with the continuous training of healthcare personnel, can contribute to earlier diagnosis and a reduction in associated morbidity.

Ethical Considerations

This case report was prepared in accordance with the ethical principles set forth in the Declaration of Helsinki and applicable international guidelines for research involving human subjects. Written informed consent was obtained from the patient for the publication of the case and associated clinical information. Confidentiality, anonymity, and the protection of personal data were ensured throughout the entire process of preparing and reporting the case.

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