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SCIENTIFIC LETTER

Perianal disease as the first manifestation in paediatric Crohn's disease☆

Enfermedad perianal como primera manifestación en la enfermedad de Crohn pediátrica

Paediatric Crohn's disease (CD) has a higher incidence of complications than Crohn's disease in adults. Perianal disease (PAD) is also a predictive factor of poor prognosis, justifying a more aggressive treatment.[1]

Three cases of paediatric patients diagnosed with CD and PAD with a single manifestation at onset are reported, with the objective of describing their presentation, phenotypic findings and outcome.

Case 1: 12-year-old male with iron-deficiency anaemia. Presenting with deep anal fissure and associated rectal bleeding of 3 months' evolution, with no improvement with topical treatments. Mother was affected by CD, associated with weight loss in recent months. Diarrhoea and fever were presented in the last week. The examination revealed perianal area with mucosal folds, two anal fissures, one with fistulous tracts and suppuration. Laboratory test results: iron deficiency anaemia, CRP 4.3 mg/dl, albumin normal, faecal calprotectin: 82 μg/g, infection screening negative. Upper gastrointestinal (GI) endoscopy: normal and ileocolonoscopy (IC) with biopsies of multiple segments, compatible with ileocolonic CD (A1b,L3,P,G-0 Paris classification). The pelvic MRI scan revealed a simple fistula and an abscess in the perianal region. wPCDAI: 60. Induction was started with adalimumab at a dose of 160, 80 and 40 mg/every 14 days of maintenance, and azathioprine and antibiotic therapy with metronidazole for 3 weeks, along with progressive clinical improvement. Patient readmitted after 1 month with erythema nodosum and fever without focal point and kidney failure during admission, suspending the azathioprine with resolution of the symptoms. Patient is currently undergoing treatment with adalimumab in monotherapy with good control.

Case 2: 8-year-old male attended for haematochezia associated with anal fissure with normal bowel movements of two years' evolution. Since then stagnant weight-to-height ratio. On physical examination: deep anal fissure and sentinel polyp in anal margin. Laboratory tests revealed iron deficiency and hypogammaglobulinaemia, infection screening negative, faecal calprotectin 2758 μg/g. Upper GI endoscopy and IC with normal macroscopic appearance, the staggered biopsies confirming the presence of non-necrotising granulomatous inflammation suggesting the diagnosis of intestinal bowel disease (A1a,L3,P,G-1 Paris classification). An MRI scan was performed: fistulas and abscesses were ruled out. Bone mineral density (BMD) was determined, presenting a *Z*-score of −2.5 in total body. wPCDAI: 25. Treatment was started with infliximab at a 5 mg/kg/day/every 8 weeks and azathioprine up to 2.5 mg/kg/day. At 7 months, the patient presented perianal abscess that required surgical draining and antibiotic therapy for 21 days. The patient developed anti-infliximab antibodies with undetectable levels of the drug. It was therefore decided to change treatment to adalimumab 40 mg/every 14 days. Subsequently, scarring of the abscess and currently on intensified adalimumab every 7 days with good progress.

Case 3: 11-year-old male. Referred from paediatric surgical consultation for recurrent anal abscesses of 3 years' evolution that had required surgical drainage. Patient is currently asymptomatic and with no perianal lesions. Laboratory tests revealed iron deficiency, ESR and CRP and infection screening negative. Faecal calprotectin: 1144 μg/g. Upper GI endoscopy and IC that revealed endoscopic lesions compatible with ileocolonic CD (A1b,L3,B1,G-0 Paris classification). Study completed with pelvic MRI which revealed left perianal fibrous tract, with no signs of acute activity. BMD was normal. Exclusive enteral nutrition (EEN) and maintenance treatment with azathioprine at 2 mg/kg/day were initiated. Induction was completed at 8 weeks with EEN. At 1 year of treatment, monotherapy with azathioprine was maintained at 2.5 mg/kg/day and with no new perianal lesions.

Perianal involvement is inflammation close to the anus and includes polyps, fissures, abscesses and/or stenosis.[2] Information is limited in the paediatric population with an incidence of PAD of between 13.6% and 62% and at diagnosis of CD, 15% had perianal lesions. Young age and severe intestinal involvement have been associated with its onset.[3]

The clinical symptoms are diverse and the patient may even be asymptomatic. PAD may be the only manifestation of the CD and precede the intestinal symptomatology even by years.[3] Fistulising symptoms have been associated with a worse prognosis.[1,4]

The objective of the treatment is to achieve the healing of the abscesses and total closure of the fistula. The arrival of anti-TNF drugs has meant a change in the treatment of these patients, biological therapy being recommended in

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induction and maintenance of the fistulising paediatric PAD, combined with surgery if necessary. Antibiotics are appropriate to support closure of the fistula.[5]

In cases without fistulising involvement, it is not clear whether it would be advisable to start early biological therapy, but due to its association with fistulising disease it could be beneficial.[1]

In children with recurrent perianal disease, CD should be ruled out as it is often the only manifestation. This would help the provision of early treatment and prevent complications.

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Pancreatic perivascular epithelial cell tumour (PECOma). Case report and literature review☆

Tumor de células epiteliales perivasculares (PECOma) de origen pancreático. Descripción de un caso y revisión de la literatura

Perivascular epithelioid cell tumors (PEComas) are mesenchymal neoplasms characterized histologically by the proliferation of perivascular epithelioid cells (PEC) and immunohistochemically by expressing melanocytic markers (HBM-45 and Melan-A) and muscular markers (actin and desmin).[1] The cells in PEComas are arranged around the blood vessels and appear to form part of its wall, often infiltrating the smooth muscle of the small and medium-sized vessels. Furthermore, in histology the cells have small, round or oval nuclei, occasionally with focal nuclear atypia and eosinophilic cytoplasm. The group known as PEComas includes angiomyolipomas, lymphangioleiomyomatosis, clear cell ''sugar'' tumors of the lung as well as other rarer types of tumor, among which are falciform ligament clear cell tumors and abdominopelvic sarcoma of perivascular epithelial cells typically associated with tuberous sclerosis, with which it shares genetic anomalies.[2] The term PEComa is used to refer to all lesions of this type that are not angiomyolipomas, lymphangioleiomyomatosis or clear cell ''sugar'' tumors of the lung.

Their behavioral spectrum ranges from benign to clearly malignant, with proposed histological criteria to assess the potential malignancy (mitotic index, vascular proliferation, necrosis). The differential diagnosis may include carcinomas, smooth muscle tumors, other clear cell neoplasms and adipose tissue tumors. PEComas constitute a genetically diverse group that includes neoplasms that harbor TFE3 gene rearrangements with TSC2 mutations, indicating alternative tumorigenic pathways. Recent advances in the treatment of malignant PEComas are related to a greater understanding of the specific genetic changes and their effects on metabolic pathways which are susceptible to specific interventions.

We present the case of a patient with a solid lesion in the body of the pancreas with puncture by endoscopic ultrasound (EUS) with atypia on whom a corporocaudal pancreatectomy with laparoscopic splenectomy was performed. Accordingly, we performed a review of the literature with cases of pancreatic PEComa published to date.

A 50-year-old female with no medical-surgical history of interest was presented. She was referred for a surgi-

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