



Name of Journal: *World Journal of Gastroenterology*

Manuscript NO: 45100

Manuscript Type: **CASE REPORT**

**Crohn's-like acute severe colitis associated with Hermansky-Pudlak syndrome: A case report**

Girot P *et al.* HPS and acute severe colitis

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**Abstract**

**BACKGROUND**

**4** Hermansky-Pudlak syndrome (HPS) is a rare autosomal recessive disorder characterized by oculocutaneous albinism, platelet storage pool deficiency and systemic complications associated with ceroid deposition in the reticuloendothelial system. HPS types 1 and 4 are associated with Crohn's disease (CD)-like gastrointestinal disorders, such as granulomatous enterocolitis or perianal disease. Cases of colitis can be particularly severe and, before the use of anti-tumor necrosis factor alpha (TNFα) therapy had become common, were reported as showing poor responsiveness to medical treatment.

**CASE SUMMARY**

We present the case of a 51-year-old albino woman who presented with acute

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**Similar** to the patient in **Case** 1, two of their patients underwent colectomy, one of which was due to **severe acute** rectal hemorrhage.

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## Complicated Crohn's-like colitis, associated with Hermansky-Pudlak



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**Hermansky-Pudlak syndrome** (HPS) is a rare autosomal recessive inherited disorder ... This **colitis** can be **severe** and has been reported to be poorly responsive to medical ... We **report** a patient with HPS which was complicated by inflammatory bowel ... An analysis of most reported **cases**, since Schinella et al. first reported ...

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## Intestinal Disease in Hermansky-Pudlak Syndrome: Occurrence of ...

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Background & Aims: **Hermansky-Pudlak syndrome** (HPS), a rare autosomal recessive .... A granulomatous **colitis associated** with HPS was first described in ... Despite several **case reports** describing gastrointestinal involvement in HPS, the ..... in susceptibility to idiopathic IBD such as **Crohn's** disease or **ulcerative colitis**, ...

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## Medical Treatment of Colitis in Patients With Hermansky-Pudlak ...

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This study will determine if medical treatment of colitis (inflammation of the colon resulting in loose bowel movements, rectal bleeding, and belly pain) that is used for other colitis conditions, such as Crohn's disease and ulcerative colitis, is safe and effective for treating colitis in patients with Hermansky- ...

## Intestinal Disease in Hermansky-Pudlak Syndrome: Occurrence of ...