Scientific Letter | Published: 10 July 2020

Vulval Edema as a Manifestation of Childhood Metastatic Crohn's Disease

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The Indian Journal of Pediatrics **88**, 182(2021)

To the Editor: A 12-y-old female child presented with recurrent aphthous ulcers since the last two years, vulval swelling since the last nine months, abdominal pain since the last one month with significant weight loss. On examination she had multiple painful aphthous ulcers on the lips, failure to thrive, painful asymmetrical bilateral vulval and clitoral edema, and perianal tags. Investigations revealed anemia (Hb = 8.9 g/dl), normal WBC (WBC = 6200/cmm; *N* = 68%, L = 22%), raised platelet counts (Platelets = 4.57 lakhs), raised CRP (67 mg/L), low albumin (1.75 g/dl), and high fecal calprotectin (310 µg/g). Ophthalmology examination was normal. Mantoux and pathergy test were negative. Colonoscopy revealed multiple linear ulcers with skip areas from caecum to sigmoid colon. Colonic biopsy revealed non-caseating granulomas. Though desirable, patient refused vulval biopsy. On the basis of the above clinical, endoscopic and biopsy findings, the diagnosis of "metastatic" vulval and

intestinal Crohn's Disease (CD) was made. She was treated with exclusive enteral nutrition therapy with polymeric formula, azathioprine, and steroids with significant improvement of both vulval edema and intestinal CD within four months of therapy with normalization of inflammatory parameters and fecal calprotectin.

"Metastatic" CD is defined by the presence of skin lesions without contiguity with the gastrointestinal tract. Vulval involvement in CD is extremely rare in children [1, 2]. Labia majora erythema and edema are common manifestations and in few cases these may progress to ulceration [3]. Perianal lesions have been estimated to be between 13.6% and 62% in pediatric CD patients [4]. Studies show that 20%–36% of patients with vulval CD do not exhibit any gastrointestinal symptoms, and vulval CD may be the first manifestation of their underlying disease [5]. Simultaneous presence of intestinal CD on colonic biopsy, perianal tags and disappearance of vulval edema to CD treatment was suggestive that the diagnosis was most likely "metastatic" CD. Recovery of "metastatic" lesions is variable, often refractory, and does not parallel to that of intestinal lesions. Treatment options include corticosteroids (topical, intralesional, systemic), antibiotics, traditional immunosuppressants, biologics and surgical debridement, all of which have shown mixed results.

We report this case as vulval involvement of childhood CD is an under-recognized manifestation.

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Ethics declarations

Conflict of Interest

None.

Additional information

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Cite this article

Kapur, S.V., Oswal, J.S. & Viswanathan, V. Vulval Edema as a Manifestation of Childhood Metastatic Crohn's Disease. *Indian J Pediatr* **88,** 182 (2021). https://doi.org/10.1007/s12098-020-03419-4

- Received01 January 2020
- Accepted15 June 2020
- Published10 July 2020
- Issue DateFebruary 2021
- DOIhttps://doi.org/10.1007/s12098-020-03419-4

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