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Extensive and unusual Mongolian blue spots in a child with GM_1 gangliosidosis type one

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Keywords: Mongolian blue spots; gangliosidosis; child

Case presented to Section of Paediatrics, 27 November 1991

We report a child with GM_1 gangliosidosis type 1 and extensive and unusual mongolian blue spots.

Case report

SA was admitted to St Mary's Hospital at the age of 5 months with difficulty in breathing and failure to thrive.

She was born at 36 weeks gestation by normal vaginal delivery, birth weight of 2.5 kg was on the 50th centile. A large sacral mongolian blue spot was noted at the discharge examination. At the age of 5 months her weight was on the 3rd centile. Her intake of milk was poor and she had not yet started on solids. She had coarse features, was tachypnoeic at rest, and had severe and chronic upper airway obstruction. There was a marked thoraco-lumbar kyphosis, but chest examination was otherwise normal. The liver and spleen were both enlarged. She had multiple areas of hyper-pigmentation on both dorsal and central trunk and elsewhere. In the weeks preceding admission these had increased in both size and number (Figure 1). Her neuro-developmental progress had been poor. She was floppy with poor head control and was not making any attempt to reach out or to support herself.

The parents, both from Pakistan were second cousins. Two of father's cousins had died before the age of 2 years.

As an inpatient she was managed with nasogastric tube feeding, oxygen and physiotherapy and following extensive investigation was found to have GM_1 gangliosidosis type 1. The multiple areas of hyperpigmentation were examined histologically following punch biopsy and were typical of a mongolian blue spot.

Her continuing care was at home where she died peacefully aged 9 months. Father's two cousins also had GM_1 gangliosidosis type 1 and at least one of them had extensive mongolian blue spots.

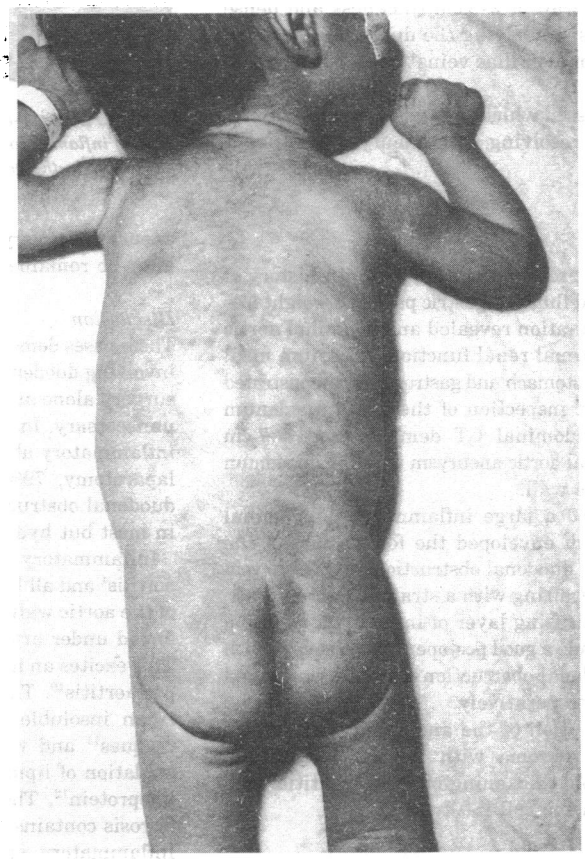

Figure 1. Multiple areas of hyper-pigmentation

Discussion

GM_1 gangliosidosis type 1 is a ganglioside storage disease characterized by its early onset, rapid progression to severe neurological impairment and poor prognosis with death usually occurring by the age of 2 years. It is a rare condition with an incidence of about 1 : 1-200 000. Inheritance is autosomal recessive and antenatal diagnosis is possible. The condition is diagnosed by estimating beta-galactosidase activity in white cells or skin fibroblasts.

Mongolian blue spots are common and are seen in up to 90% of Asian babies, and less commonly in other races. They are benign and have no known associations. Histologically they are characterized by melanocyte proliferation in the mid dermis. They are usually lumbo-sacral and can be single

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or multiple. They can increase in both size and density but usually disappear by the age of 5 years[1]. They can persist into adult life in 3-4% of cases, this was found in a study of healthy Japanese males[2]. Extensive mongolian blue spots do certainly occur in up to 5% of cases although their extent and natural history have not yet been studied.

The mongolian blue spots seen in our child were both very extensive (including the ventral surface of the trunk) and unusual in that they were rapidly increasing at the age of 6 months.

The possible chance association between GM_1 gangliosidosis type 1 and extensive mongolian blue spots was first described in 1981 by Weissbluth *et al.*[3] who reported a 5-month-old child with extensive areas of hyper-pigmented skin and the condition. Skin biopsy from this child showed the hyper-pigmented areas to resemble mongolian blue spots. A further child was reported in 1989[4] and two more in 1990[5]. There are now six reported cases including the four previously reported, our case and her father's cousin. These six children therefore suggest that there might be an association of GM_1 gangliosidosis type 1 with extensive and unusual mongolian blue spots.

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Association between pyoderma gangrenosum and ulcerative colitis

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Keywords: pyoderma gangrenosum; inflammatory bowel disease; ulcerative colitis

Case presented to Section of Dermatology, 20 October 1988

The association between pyoderma gangrenosum and ulcerative colitis is well recognized, the pyoderma usually occurring on the legs[1]. Occasionally however, pyoderma gangrenosum occurs around the site of a stoma, when it is often incorrectly diagnosed.

We report the case of a 56-year-old woman with ulcerative colitis who has had two episodes of peristomal pyoderma gangrenosum. These occurred after the formation and then the resiting of the stoma and were at one time thought to be artefactual in origin.

Case report

A 56-year-old woman developed ulcerative colitis in 1979, and underwent panproctocolectomy with ileostomy formation in 1980. Histology of the gut showed non-specific colitis. There were no features to suggest Crohn's disease. One month postoperatively, she developed ulceration around the stoma site which persisted for about 8 months and then resolved spontaneously.

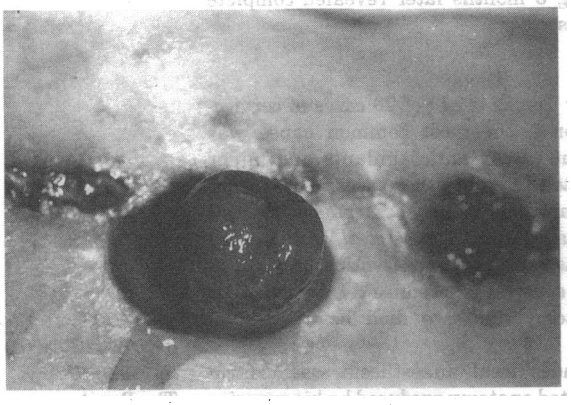

Figure 1. Peristomal ulceration with the typical undermined edge of pyoderma gangrenosum

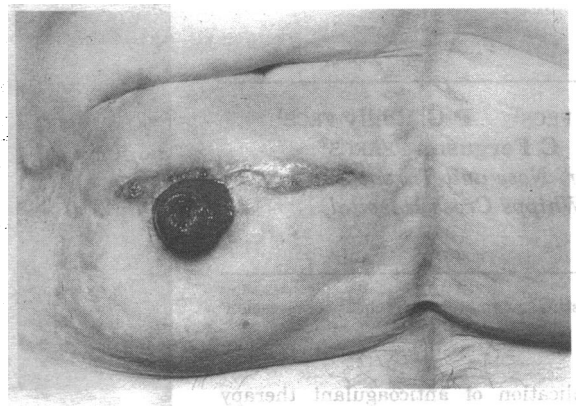

Figure 2. Characteristic atrophic scar

She remained well until 1986 when resiting and refashioning of the ileostomy became necessary due to ileostomy retraction. Six weeks postoperatively, she again developed a persistent peristomal ulceration with an undermined purplish edge (Figure 1). There was no significant bacterial growth from the ulcerated site which continued to extend despite local measures. An artefactual origin was considered and she was referred to the dermatology department for a second opinion. Because of the characteristic clinical appearance and the typical history of ulceration occurring shortly after stoma formation a diagnosis of pyoderma gangrenosum was made and minocycline was prescribed. This resulted in rapid improvement of the ulceration and healing with atrophic scar formation (Figure 2). The ulceration has not recurred since stopping the minocycline.

Discussion

Peristomal pyoderma gangrenosum has only rarely been reported as occurring in ulcerative colitis. In a series of 1132 cases of ulcerative colitis there were 21 cases of pyoderma gangrenosum, only one of which occurred in association with the stoma[2]. A second case was described in detail by Giroux[3]. His patient had two episodes of peristomal pyoderma gangrenosum occurring shortly after formation and then relocation of an ileostomy and at a time when his ulcerative colitis was quiescent.

Other cases of pyoderma gangrenosum occurring after trauma (although not specifically related to stoma formation) have been described[4]. These have occurred in both ulcerative

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