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BLUE RUBBER BLEB NEVUS SYNDROME: REPORT OF A CASE*

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A case of blue rubber bleb nevus syndrome (BRBNS) in a 17-year old boy with multiple cutaneous and gastrointestinal hemangiomas is presented. The patient had iron deficiency anemia and pica. In addition, we describe, for the first time, left renal artery, right hepatic artery, and ureteral abnormalities in a patient with BRBNS. *Key words: blue rubber bleb nevus syndrome, gastrointestinal involvement, vascular abnormalities.*

The blue rubber bleb nevus syndrome (BRBNS) was first described by Bean[1] in 1958, and is named after the soft, bluish, rubber-like hemangiomas mainly of the skin and the gastrointestinal tract. These lesions may also be found in other locations such as in the peritoneal cavity, the heart, lungs, pleura, and central nervous system[2-4]. To date, over 40 cases have been reported.

Our patient, besides illustrating the characteristics of this syndrome, namely, the nature and distribution of the lesions and iron deficiency anemia, in addition presented with arterial and ureteral abnormalities.

Case Report

A 17-year-old boy was admitted to the Hacettepe University Hospital with the diagnosis of iron deficiency anemia. There was a 7-year history of geophagia. Flat, blue, non-tender lesions varying in size from 0.5 to 1.0 cm were seen on the hands, feet, tongue, and lip (Fig. 1). The hemoglobin was 7.6 g/dl and hematocrit 24%. Blood smear revealed microcytic, hypochromic red blood cells. The serum iron was 13 μm/L and total iron binding capacity 375 μm/L. Tests for occult blood in the stool were positive.

Esophagogastroduodenoscopy confirmed the presence of multiple 0.5-1.5 cm papillary bluish hemangiomas in the stomach and duodenum (Fig. 2). Colonosco-

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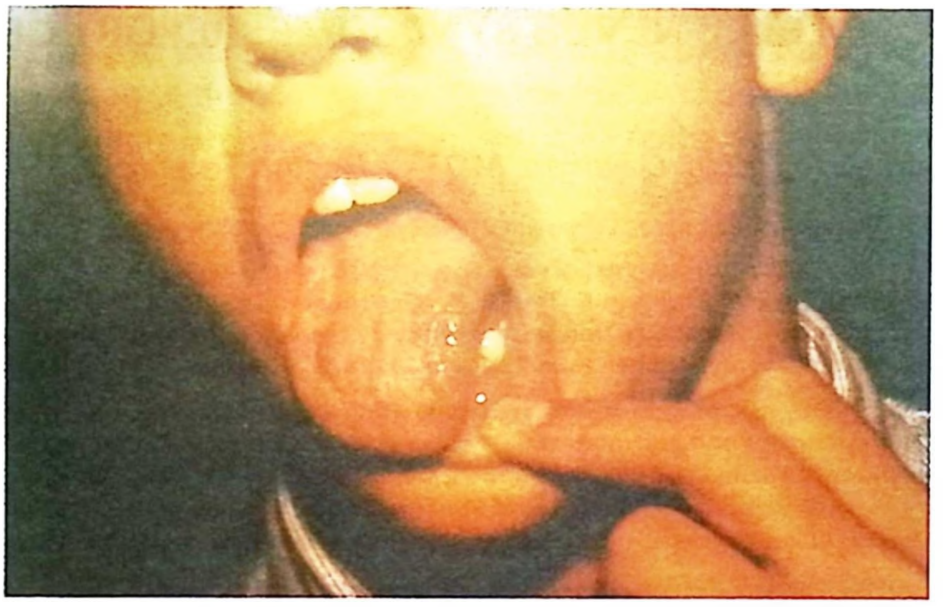

Fig. 1: Blue rubber bleb hemangiomas on the tongue.

Fig. 2: Endoscopic view of hemangiomas in the stomach.

pic examination revealed multiple 0.5-1.5 cm dark blue pedunculated polypoid lesions between the cecum and 20 cm from the anal orifice.

An aortagram showed the right hepatic artery originating from the superior mesenteric artery and two renal arteries originating from the aorta on the left side. There was an ectopic insertion of the left ureter into the bladder.

Abdominal ultrasonography revealed splenomegaly. Hepatitis B surface antigen was negative. Liver biopsy was normal. The splenomegaly was believed to have been caused by pica.

The patient was transfused with red blood cells and was placed on continuous oral iron supplementation.

At follow-ups his hemoglobin level remained at about 10 g/dl.

Discussion

The characteristic finding of BRBNS is a few to hundreds of skin lesions varying in size from 1 mm to 10 cm[3,5]. Three types of skin lesions have been described. Type I is a large disfiguring cavernous angioma that may increase in size and obstruct vital tissues. Type II is a "blue rubber nipple" covered with thin skin that is compressible, leaving an empty, wrinkled sac that rapidly refills. This is the commonest type and was presented in our patient. Type III is an irregular, blue-black macule or papule that may be punctate or merge with adjacent pigmented nevi and may blanch on pressure. The limbs, trunk, and face are the main sites of involvement. Lack of nail bed involvement distinguishes BRBNS from Osler-Weber-Rendu syndrome.

Intestinal hemangiomatous lesions are more frequently found in the rectum and distal colon than in the proximal colon[6]. Moreover, the number of lesions may increase with age[7]. In our patient, there were widespread lesions in the colon, stomach, and duodenum. The intestinal lesions were not biopsied because of the risk of severe bleeding.

Patients usually present with iron-deficiency anemia which is due to chronic blood loss from the gastrointestinal tract, as in our case. As in our patient, most cases of BRBNS do not have a positive family history[7]. Associated orthopedic abnormalities have been described in 14 patients[8]. Two patients with BRBNS presented with ophthalmologic manifestations[2,9]. Our patient is unique because he had vascular and ureteral abnormalities. Whether this is a coincidental association can only be made clear by performing angiographic studies more frequently in these patients.

REFERENCES

1. Bean WB. Blue rubber bleb naevi of the skin and gastrointestinal tract. In Bean WB (ed). *Vascular Spiders and Related Lesions of the Skin.* Springfield: C.C. Thomas, 1958, pp 178-185.
2. Cromptom JL, Taylor D. Ocular lesions in the blue rubber bleb nevus syndrome. *Br J Ophthalmol* 65: 133, 1981.
3. Rice JS, Fischer DS. Blue rubber-bleb nevus syndrome: generalized cavernous hemangiomatosis or venous hamartoma with medulloblastoma of the cerebellum: case report and review of the literature. *Arch Dermatol* 86: 503, 1962.
4. Waybright EA, Selhorst JB, Rosenblum WI, Suter CG. Blue rubber bleb nevus syndrome with CNS involvement and thrombosis of a vein of Galen malformation. *Ann Neurol* 3: 464, 1978.

5. Baker AL, Kahn PC, Binder SC, Patterson JF. Gastrointestinal bleeding due to blue rubber bleb nevus syndrome: a case diagnosed by angiography. *Gastroenterology* 61: 530, 1971.
6. Berlyne GM, Berlyne N. Anaemia due to "blue-rubber-bleb" naevus disease. *Lancet* 2: 1275, 1960.
7. Sandhu KS, Cohen H, Radin R, Buck FS. Blue rubber bleb nevus syndrome presenting with recurrences. *Dig Dis Sci* 32: 214, 1987.
8. McCarthy JC, Goldberg MJ, Zimbler S. Orthopaedic dysfunction in the blue rubber-bleb nevus syndrome. *J Bone Joint Surg* 64-A: 280, 1982.
9. Rennie IG, Shortland JR, Mahood JM, Browne BH. Periodic exophthalmos associated with the blue rubber bleb naevus syndrome: a case report. *Br J Ophthalmol* 66: 594, 1982.