THE BLUE RUBBER BLEB SYNDROME

Pages with reference to book, From 20 To 21

Tariq Shaft, Tahir Hussain, M. Shaft Quraishy (Department of Medicine, Unit 5, Dow Medical College and Civil Hospisal, Karachi.)

INTRODUCTION

The blue rubber bleb syndrome is a Condition in which multiple hemangiomas of the skin are associated with hemangiomas of the gastrointestinal tract[1-4]. The syndrome was first clearly delineated by Bean[2]. It consists of compressible soft blue swellings of the skin present predominantly on the trunk and upper arms. They are frequently tender or spontaneously painful. Localized sweating may occur over the lesions. There are associated hemangiomas in the gastrointestinal tract which are most common in the small intestine. These can bleed and lead to anaemia[1-3]. Aetiology is unknown with reported sporadic cases and a number of reports indicating an au tosomal dominant pattern of transmission[1-5].

CASE REPORT

An 11 year old girl presented in the outpatient department with a 6 years history of small multiple swellings on the limbs and back. She had first noticed a swelling on her left thumb which gradually increased in size and was followed by similar swellings appearing on other fingers, palms, on dorsum of feet, back and on the oral mucosa. These were initially painless but later became painful, with occasional bleeding mostly following trauma and this led her to seek medical advice. On clinical examination swellings were bluish, soft, compressible, varying in size from 1x1 ems to 1x2 ems, arising from skin and subcutaneous tissue, non-fluctuant and non- translucent. These were present on all fingers of the hands (Figure 1),

Figure 1. Hemangiomas on the hands

the left palm, dorsum of the feet, back and the oral mucosa. There were no other positive findings. Her hemoglobin concentration was 13.6g/dl and platelet count was 2,72,000/cmm. Stool I)R showed no RBC's and no occult blood. All other biochemical and haematological investigations were normal. 11cr upper G.I. endoscopy was done using an Olympus GIF XQ2O fibre optic endoscope which revealed a hemangioma on the lesser curvature of stomach (Figure 2).

Figure 2. Endoscopic view of the hemangioma on the lesser curvature of stomach.

Biopsy was not attempted. Choice of either surgical resection or photo-coagulation or cryodessication was offered. Patient opted for the latter facility which is expected to be acquired by our hospital in near future.

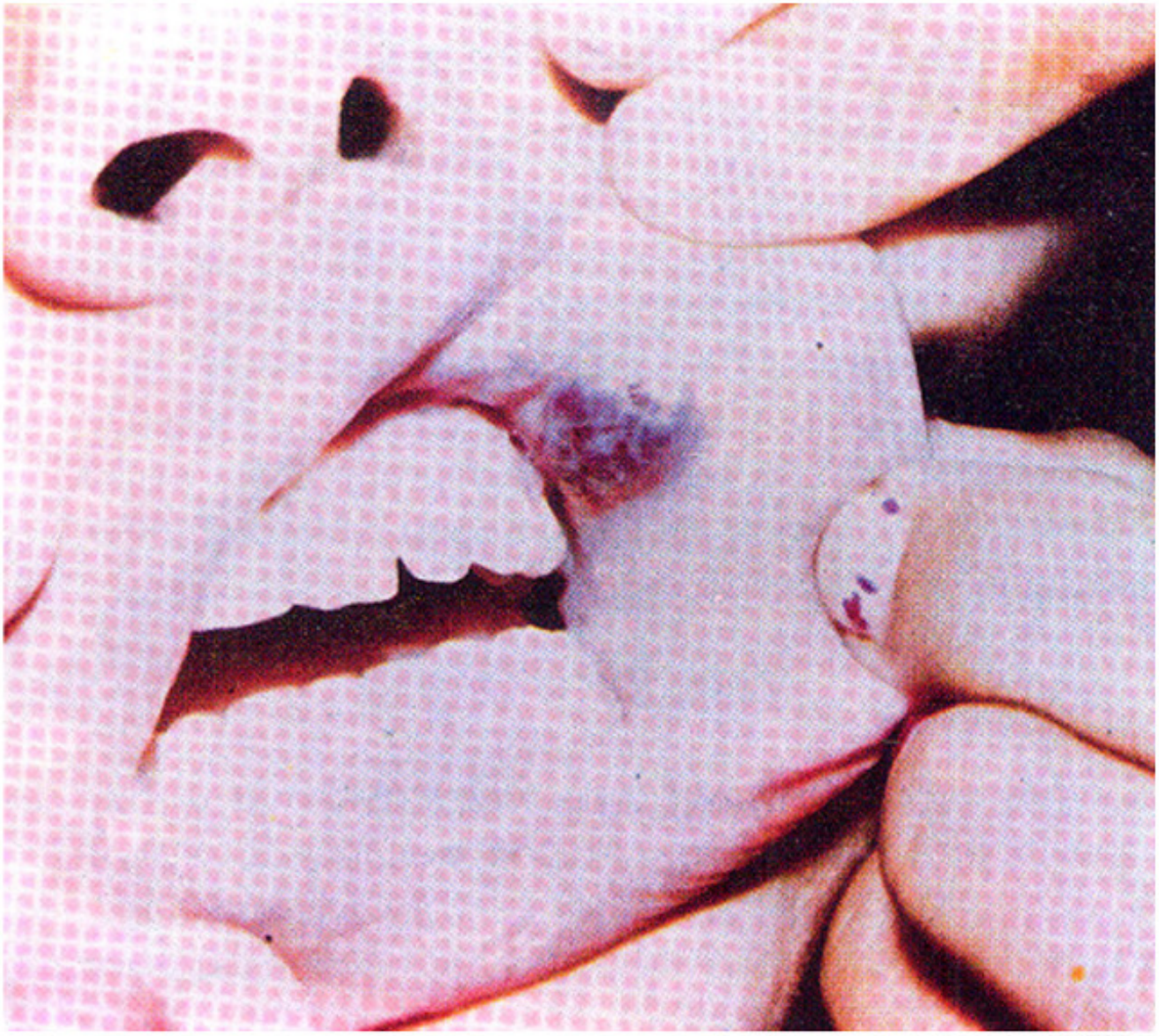

Figure 3. Hemangioma on the buccal mucosa.

DISCUSSION

Hemangiomas are benign vascular neoplasms composed of endothelial lined vascular spaces. Differentiation between capillary and cavernous types is based on histopathology; 20-30% are present at birth and almost all develop by 9 months and rarely they may occur for the first time at a later age. In 80% of cases only a solitary lesion is present but in 20% multiple hemangiomas may be present[1]. These may occur either alone or in association with hemangiomas of other internal organs giving rise to a number of different syndromes[1-4]. A variety of treatments have been suggested indicating a lack of consensus. They vary from medical treatment such as administration of corticosteroids to surgical excision after identification of feeding vessels (which can either be embolized or ligated at time of surgery) to the use of carbondioxide lasers2 to such innovative techniques as the use of monoclonal antibodies directed against oestrogen receptors detected in some of these tumours[6].

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