

Case Study

INFANTILE HAEMANGIOMA IN A 'BEARD' DISTRIBUTION: A CASE REPORT

ABSTRACT

Aim: To highlight infantile haemangioma as a possible cause of upper airway obstruction in young infants presenting with stridor especially when there are associated cutaneous lesions.

Presentation of Case: We report a 10 week-old baby girl admitted into the emergency paediatric unit of the Federal Medical Centre Azare, Nigeria with a 3-week history of a skin lesion initially noticed as a flat discoloured area over the sternal region. It progressively spread to involve the anterior aspect of the neck and chin and became associated with progressive difficulty in breathing. Examination findings revealed a restless, acutely ill child with inspiratory stridor. She had purplish plaques in the areas mentioned above as well as the lower lip and ventral aspect of the tongue. She improved remarkably following oxygen, intravenous dexamethasone and oral propranolol administration and was discharged after 4 days of admission.

Discussion: Infantile haemangiomas are benign vascular neoplasms that have a characteristic clinical course marked by early proliferation and followed by spontaneous involution. Infrequently, infantile haemangiomas may impinge on vital structures provoking certain related symptoms. Airway obstruction is an uncommon complication and may be particularly challenging for young infants who are obligate nose breathers. Subglottic haemangiomas are also uncommon compared to the cutaneous forms and may be life threatening with attendant airway

obstruction. This was the foremost concern in our patient. Nevertheless, she responded to dexamethasone and propranolol.

Conclusion: It is essential to recognize the association of infantile haemangiomas presenting in a beard distribution with airway obstruction and promptly institute management of respiratory complications.

Keywords: Infantile haemangioma; Benign neoplasm; Beard distribution; Respiratory distress; Stridor; Azare.

INTRODUCTION

Infantile haemangiomas are the most frequently encountered benign vascular tumours of infancy and childhood, they are bright red, protuberant (may be macular or papular at onset), compressible, sharply demarcated lesions that may be found in any part of the body. [1,2] They affect nearly one in ten infants with a higher incidence seen in preterm neonates and those who were exposed to chorionic villus sampling.[3,4] Haemangiomas are commoner in girls than boys with incidence ratios ranging from 3:1 – 4:1 respectively being variously quoted.[1,4] The exact aetiology of infantile haemangiomas is not known. However, it has been postulated that an association between immature endothelial cells and pericytes as well as angiogenic peptides, such as beta-fibroblast growth factor, vascular endothelial growth factor (VEGF), and proliferating cell nuclear antigen, give rise to the development of hemangiomas.[5,6]

Most infantile haemangiomas undergo a phase of rapid expansion, followed by a stationary period and finally spontaneous involution. Even though most haemangiomas are not challenging, requiring no treatment, about 10% may cause significant morbidity mainly through airway obstruction, ocular compression, functional impairment and ulceration.[1,7] Complications

include ulceration, secondary infections and rarely haemorrhage. In the majority of cases haemangiomas are diagnosed based on history and examination findings. If diagnosis remains in doubt, colour doppler ultrasonography and/or MRI may prove helpful.[1]

Treatment modalities currently in use in the management of problematic haemangiomas include, drug therapy, laser therapy and surgery. Medications presently being used include, propranolol, timolol, corticosteroids, alpha-interferon, cyclophosphamide, vincristine, pinyangmycin, and imiquimoid.[1]

Hemangiomas in a 'beard' distribution are uncommon and may present with involvement of the upper airways or the subglottic areas and associated respiratory complications.[8]We present this exceptional case underscoring the need for a high index of suspicion and anticipatory management of life threatening respiratory complications in children with infantile cutaneous haemangiomas in a 'beard' distribution.

PRESENTATION OF CASE

A 10 week-old baby girl presented to the Emergency Paediatric Unit (EPU) of the Federal Medical Centre Azare, Nigeria on account of a 3-week history of raised multiple reddish rashes over the upper aspect of the chest. These progressively spread upwards to involve the anterior neck, lower jaw and lower lip. The lesions also spread laterally to involve the infra-auricular region bilaterally. They were soft, painful, non-discharging and subsequently coalesced to form a huge mass. There were no swellings in any other part of the body and there was no bleeding either from the lesion or any other part of the body. At the time the lesion was noticed to have spread to the neck, she developed progressive difficulty in breathing, noisy and fast breathing. There was no associated change in voice or drooling of saliva. She also developed an insidious

onset cough, becoming progressively restless and refusing feeds. This was the first episode of such symptoms in the child's life, there was no family history of a similar illness and she was delivered at term at a primary health facility following an unsupervised pregnancy.

On examination, she was acutely ill, in respiratory distress, and with inspiratory stridor. She was febrile with an axillary temperature of 38°C. There were multiple plaques of various sizes on the supra-sternal region, anterior aspect of the neck, lower jaw, lower lip, buccal mucosa and the ventral surface of the tongue. They were soft, non-tender, the largest measuring 8×3cm in its widest diameter on the anterior chest wall (Fig 1). She had no dysmorphic features and Anthropometric measurements were normal. The respiratory rate was 48cpm, she was dyspnoeic, had vesicular breath sounds with widespread coarse crepitations bilaterally. A diagnosis of infantile haemangioma in a beard distribution with upper airway involvement and bronchopneumonia was made.

She was placed on intranasal oxygen via nasal prongs, intravenous ampiclox, gentamycin and dexamethasone as well as oral propranolol. Her full blood count and differential count results as well as serum urea and electrolytes were within normal values. However, chest radiograph showed features consistent with bronchopneumonia. Following complete recovery, she was discharged home on oral propranolol, prednesolone and oral ampiclox after 4 days of admission to be followed up in the outpatient clinic. However, nine days after discharge, she re-presented to the EPU with the same symptoms. Her caregiver admitted to stopping the administration of prescribed medications 5 days earlier. She had the same treatment as in the previous admission with remarkable improvement noticed after a 24-hour period. Prior to discharge 2 days later, the caregiver was re-counselled on the disease including compliance to follow up and medications. Nonetheless, she was lost to follow up.

88 **DISCUSSION**

89 Haemangiomas typically show rapid proliferation in infancy which may manifest with
90 potentially life threatening complications as demonstrated by our patient.[9,10] Subglottic
91 haemangiomas are uncommon compared to the cutaneous forms and may be life threatening in
92 the proliferative phase with attendant airway obstruction.[10]This was the foremost concern in
93 our patient. However, due to limited facilities in our facility we were unable to carry out some
94 investigations like, direct endobronchoscopy and MRI of the neck. These would have aided in
95 the determination of the extent of involvement of the airway and confirmation of subglottic
96 affectation. Our patient also presented with clinical and radiological features of
97 bronchopneumonia, which is not a common finding in ‘beard’ presentation of infantile
98 haemangiomas.

99 Response to corticosteroids and propranolol as shown in our patient is also well
100 documented.[1,8,9] Propranolol, a non-selective beta blocker is thought to act by
101 vasoconstriction, decreasing growth factors and perhaps preventing the haemangioma stem cells
102 from differentiating into endothelial cells or pericytes leading to involution while steroids have a
103 direct inhibitory effect on angiogenesis and cause enhanced expression of genes coding for
104 markers of apoptosis. [7,11,12] These drugs may have facilitated the regression of the tumours
105 resulting in marked improvement of symptoms. However, a lack of compliance to medications
106 led to a resurgence of symptoms necessitating readmission. There is the need to stress the
107 importance of continued, uninterrupted medical therapy and follow up for such patients until the
108 lesions resolve completely as they may recur and cause life threatening airway compromise.

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CONCLUSION

Airway obstruction is an uncommon complication of haemangiomas and may be more challenging for young infants who are obligate nose breathers. We recommend that infantile haemangioma of the upper airway be considered in young infants with respiratory distress and stridor especially when they present with cutaneous lesions. Anticipatory management of these respiratory complications may improve survival in resource-poor settings like ours.

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COMPETING INTERESTS

We declare that no competing interests exist here.

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152 **FIGURES**

153 Figure 1

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