



## **Infantile Haemangioma in a 'Beard' Distribution: A Case Report**

**Iragbogie Al-Mustapha Imoudu<sup>1\*</sup>, Maimuna Orahachi Yusuf<sup>1</sup>  
and Hayatu Ahmad<sup>1</sup>**

<sup>1</sup>*Department of Paediatrics, Federal Medical Centre, PMB 005, Azare, 751101, Bauchi State, Nigeria.*

### **Authors' contributions**

*This work was carried out in collaboration between all authors. Author IAMI wrote the draft of the manuscript. Author MOY wrote the case summary. Author HA conceptualised the report. All the authors read and approved the final draft of the manuscript.*

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**Case Study**

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### **ABSTRACT**

**Aim:** To highlight infantile haemangioma as a possible cause of upper airway obstruction in young infants presenting with stridor particularly 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 with 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 intranasal oxygen, intravenous dexamethasone and oral propranolol administration and was discharged after 4 days of admission on oral prednisolone and propranolol.

**Discussion:** Infantile haemangiomas are benign vascular neoplasms that have a characteristic clinical course marked by early proliferation and followed by spontaneous involution. Infrequently,

\*Corresponding author: E-mail: tomhe164@yahoo.com;

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.

## 1. 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 [1-4]. These tumours 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 [1,5]. 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 [6,7].

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 [1,8]. 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 for problematic haemangiomas are, drug therapy, laser therapy and surgery. Effective medications 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 [9]. 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.

## 2. 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 with breathing, as well as 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 breast milk. 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. These lesions were soft, non-tender, the largest measuring 8x3 cm in its widest diameter on the anterior chest wall. She had no dysmorphic features and anthropometric measurements were normal. The respiratory rate was 48 cpm, 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 at 200 mg/kg/day in 6 hourly divided doses, gentamycin at 2.5 mg/kg 8 hourly and dexamethasone at 0.15 mg/kg/dose 6 hourly as well as oral propranolol 2 mg/kg/day bid. Her full blood count and differential count results as well as serum urea and electrolytes were normal. However, chest radiograph showed features consistent with bronchopneumonia.

Response to management was remarkable as she was able to breathe without distress, showed a gradual regression in the size of the lesions, became afebrile and was able to feed within 48 hours of commencement of treatment. Following complete recovery, she was discharged home on oral propranolol at 2 mg/kg/day, prednisolone at 2 mg/kg/day and oral ampiclox at 200 mg/kg/day 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 (she did not have any clinic visit).

### **3. DISCUSSION**

Haemangiomas typically show rapid proliferation in infancy which may manifest with potentially life threatening complications as demonstrated by our patient [10,11]. Subglottic haemangiomas are uncommon compared to the cutaneous forms and may be life threatening in the proliferative

phase with attendant airway obstruction [11]. This was the foremost concern in our patient. Yet, due to limited facilities in our centre we were unable to carry out some investigations eg. direct endobronchoscopy and MRI or magnetic resonance angiography (MRA) of the head and neck as well as immunohistochemistry for confirmation of diagnosis. These would have also aided in the determination of the extent of involvement of the airway and confirmation of subglottic affectation.

The index patient also presented with clinical and radiological features of bronchopneumonia, which is not a common finding in 'beard' presentation of infantile haemangiomas. However, infections have been known to complicate infantile haemangiomas particularly when they are ulcerated [1,4].

The effectiveness of corticosteroids and propranolol in infantile haemangiomas (as shown in our patient) is well documented [1,9,10]. Propranolol, a non-selective beta blocker is thought to act by vasoconstriction, decreasing growth factors and perhaps preventing the haemangioma stem cells from differentiating into endothelial cells or pericytes leading to involution. Steroids have a direct inhibitory effect on angiogenesis and cause enhanced expression of genes coding for markers of apoptosis [8,12-15]. These medications may have facilitated the regression of the tumours in this case resulting in marked improvement of symptoms. However, a lack of compliance to medications led to a resurgence of symptoms necessitating readmission. This has been well documented by some studies [16,17].

Steroids are mostly useful in the early proliferative phase of infantile haemangioma and are continued until tumour growth has stopped or a complete reduction in size is demonstrable. They are then gradually tapered [18]. Oral propranolol on the other hand is known to have early, intermediate and long-term effects [16]. The initial effect which is due to  $\beta_2$  adrenergic inhibition (vasoconstriction) is usually obvious within 1-3 days of commencing therapy as demonstrated in this case [16]. The length of propranolol therapy is determined by the extent of involvement, the treatment indications and the morphology of the tumour. Some authors have suggested 6 months of treatment with adjustments for certain morphological subtypes. For instance, treatment may be continued for up to 12 months in cases of deep and mixed

infantile haemangiomas [19]. We were unable to track the ultimate response and outcome in this case as a result of poor compliance to the management plan. There is the need to stress the importance of continued, uninterrupted medical therapy and follow up for such patients as they are at risk of tumour resurgence and life threatening airway compromise.

#### 4. 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 haemangiomas 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.

#### CONSENT

As per international standard or university standard written patient consent has been collected and preserved by the authors.

#### ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the authors.

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#### COMPETING INTERESTS

Authors have declared that no competing interests exist.

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