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Case Report

Large infantile haemangioma of the cheek: A case report

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ABSTRACT

Infantile haemangiomas (IH) or Strawberry naevi as were previously called are common neoplasms of childhood. The tendency to spontaneously involute differentiates this tumour from other vascular neoplasms. Though many treatment modalities have been described with advances in understanding of tumour pathology, it has been consistently found that minimal intervention under watchful observation offers the best chance of getting the best result in the long term. Though parents are extremely nervous, seeing the grotesque nature of these tumours, patient, firm counselling and reassurance is all that is required in most cases for successful management.

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1. Introduction

A 5-month old baby was brought by anxious parents with a progressively increasing swelling of the face disfiguring the right cheek and lip. The swelling started from an oozy red spot in the centre of the right cheek at 7 days of birth and grew in a sustained fashion. There were intermittent brisk bleeding episodes that subsided with pressure and healed with scars. The child was born at term and there was no significant antenatal or family history.

On examination, there was a boggy swelling of the entire right cheek, reddish brown, variegated surface, extending from the right lower eyelid into the dorsum of the nose and involving the right half of the upper lip. The swelling was pulling down the right half of the face (Figures 1 and 2). There was no tenderness. Did not bleed on touch. A diagnosis of Infantile haemangioma (IH) was made. The parents were counselled in detail about the course of the lesion, assured that it was best followed up as it would

spontaneously involute, unless the swelling obstructed vision or interfered with feeding the baby. Though the lesion was large, but had started flattening and greying out in places, no topical or oral medicine was started for the child and parents advised to report promptly, if there was any complication and discharged.

2. Discussion

Infantile haemangiomas occur in 4 to 5% of infants,¹ more common in female children than male in the ratio of 1.4: 1 to 3:1.² In children with PHACES syndrome (posterior fossa defects, Haemangioma, Abnormal arteries in the brain and heart, Coarctation of aorta, eye problems and sternal clefts) the incidence among female children is much more (9:1).³ The incidence is higher in Caucasian race, children with low birth weight, (30% in children less than 1 kg at birth), maternal Chorionic villous sampling, amniocentesis, older maternal age, breech presentation, placental abnormalities all seem to increase the incidence of IH.⁴ The common factor in these cases seems to be placental hypoxia. For

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Fig. 1: Infantile haemangioma right cheek- frontal view



Fig. 2: Infantile haemangioma right cheek- worm's eye view

some unknown reason being the first-born increases risk.²

The pathogenesis of IH has been proposed to be tissue ischemia that leads to neo vascularisation from Endothelial proliferation cells.⁵ GLUT 1, a placental glucose transport protein is the unique immune marker for IH which is not present in any other vascular malformation.⁶

The clinical evolution of IH involves an initial proliferative phase and a later involution phase. An IH appears within 4 weeks of birth, rapidly grows up to 3 to 5 months of age, and then starts regressing after 1 year.⁷ Between 5 months to 1 year is a plateau phase where proliferation and involution occur at different sites simultaneously. The involution phase proceeds slowly, with most of the tumour resolving by 4 years of age. During the proliferative phase if the IH is superficial, it will appear as a raised, reddish, well circumscribed lesion with a nodular surface (strawberry naevus). A deep IH below the skin will give a blue hue and creates a diagnostic confusion.

Mixed or compound IH is when both superficial and deep components are present like in the described case. When involution starts, the lesion starts flattening and shrinking from center outwards. Even after maximal involution, redundant skin with fibrofatty tissue, telangiectasias, scars and varied pigmentation may remain.⁸ Anatomically, IH are classified as focal, segmental, indeterminate and multifocal. In the head and neck region, the segments involved are fronto-temporal, maxillary, mandibular and frontonasal (as in this case).⁹ Though focal lesions are more common in the face, segmental IH is associated with 11 times more complications like ulceration, bleeding, distortion and destruction of structures than local lesions and 8 times more likely to need treatment.¹⁰

The diagnosis of IH is usually by history and clinical examination. Sometimes, when there are complications necessitating intervention, then imaging may be required to assess the extent of the lesion (especially when syndromes are suspected), the vascularity of the tumour and the response to treatment. The most common imaging modalities are Ultrasound and MRI scan.¹¹

More than 60 % of IH can be managed by just leaving them alone and watching their progress / regression over the years. Intervention is necessary when 1) A life threatening complication is anticipated like airway obstruction, 2) When there is a functional impairment like blocking the line of vision or feeding impediment, pain or bleeding or 3) to prevent permanent disfigurement and scarring.

Nowadays, Propranolol administered as an oral therapy in the dose of 1 to 3.4 mg/kg body weight has been shown to be highly efficacious in reducing the growth of IH and safe in children. The mechanism of action is most probably by vasoconstriction, stopping angiogenesis and inducing apoptosis. The other treatments tried are corticosteroids - topical, intralesional and systemic, Interferon alpha, Imiquimod and other anti-angiogenic agents. Pulse dye laser has also been tried with varying results. Surgery is reserved for emergency relief of complications, for very small lesions in critical locations or for scar improvement after resolution.

3. Conclusion

Infantile haemangiomas are common childhood neoplasms. Clear understanding of pathology has made the classification of these lesions more streamlined and confusing nomenclatures like “capillary” , “Cavernous“ haemangiomas have been dispensed with. As IH have the unique ability to involute after a rapid growth phase, parents of affected children need to be counselled effectively to submit to a watch and wait policy. Complications should be anticipated, and intervention instituted early in deserving cases. Except for propranolol therapy, any other intervention, has not been superior to the result obtained after natural involution, therefore unless indicated intralesional injections, lasers or surgery should not be

irrationally offered. A “benign neglect” policy may still be the best approach to infantile haemangiomas.¹²

4. Conflict of Interest

The authors declare that there are no conflicts of interest in this paper.

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