UNUSUAL GROWTH - OROFACIAL REGION

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MJAFI 1999; 55 : 81-82

KEY WORDS : Epignathus, Oro-facial growth.

Introduction

Growth involving the orofacial region in the new born, is rare. A careful evaluation is mandatory to rule out the involvement of any other element of the cranio-maxillofacial region or other parts of the body. Epignathus defines any type of growth arising from the upper jaw or palate and projecting from the mouth [1]. Although most are benign, their size and location often result in respiratory or feeding difficulty. The unusual nature of the large congenital growth and non-availability of any previous mention of such growth in the literature, so far, has prompted us to report this case of growth of the orofacial region.

Case report

A 6-month-old child presented with complaints of a large, firm and fleshy mass protruding from the upper jaw and lip. The mass was noticed since birth. The general status of the child indicated malnourishment (body weight 4 kg) and respiratory tract infection. The child had difficulties in taking his feeds. He was not breast fed since birth. Locally the swelling was roundish, 6 cms in diameter, sessile, firm to hard in consistency involving the midline pre-maxillary region, two-third of the upper lip structure and vestibule. The mass was enveloped by skin dorsally and mucosa on the ventral aspect which blended with the adjoining structure. The colour and texture of the skin and mucosa was normal. The deformity was associated with cleft hard and soft plate (Fig 1). Radiologically a dense radiopaque snouty curvilinear shadow could be seen in extension from the pre-maxillary region. The shadow was enveloped with a soft thick radiopaque contour.

With the improvement of the general health status, resolution of respiratory tract infection and increase in body weight to 5.5 kg, the patient was operated upon in July 97.

Excision of the growth combined with upper lip and vestibular reconstruction were carried out. The excised mass measured 6.5 X 6 x 4.0 cms. The Microscopical findings of the department of Pathology, Northern Command Hospital stated "the covering skin contains normal hair roots, sweat glands and appendages deep to which there is abundant fat. There are bony trabeculae of varying thickness and bony density separated by abundant haemopoietic marrow. There are two unerupted milk teeth partly in relation to the bone. The vermillion border of skin is identifiable which contains on the buccal aspect numerous mucus salivary glands. No elements of tissue not native to the site of biopsy are seen so as to

suggest a teratoma". Opinion was Congenital Hamartoma Upper Jaw.

The post-operative period was uneventful. Patient was discharged on 10th post operative day. Review after 6 weeks showed satisfactory healing of soft tissue, acceptable reconstruction of the upper lip and increase in body weight to 7 kg.

Discussion

The protruding mass in our case, involved the pre-maxilla and the upper lip structures. Embryologically the median nasal, lateral nasal and the maxillary processes initiate the sequential activities in the 4 week intra-uterine life to form the premaxilla and the upper lip (Fig 2). The morphogenesis depends on two principal interactions—firstly there are genetic instructions and secondly the capacity in the local tissues for the unhindered cell replication, cell survival and programmed cell death [2]. Melanocytic neuro ectodermal tumor [3] of infancy has been classified as a hamartoma and as a benign neoplasm, the latter terminology being more favourable by recent authors. It is a rare tumour showing characteristic histologic picture of melanin containing epithelial cells lining cuboidal or slit like spaces. Recent immunohistochemical studies confirm that it is a tumor with polyphenotypic expression of neural and epithelial markers, melanin produc-

Fig. 1 : Swelling in the midline premaxillary region

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Fig. 2: Post operative photograph showing satisfactory reconstruction of lip.

tion and glial and rhabdomyoblastic differentiation [3]. Mention of different terminologies such as Pigmented Neuroectodermal tumor of infancy or Melanotic progonoma or Retinal anlage tumor [4] has been also reported for growth presenting similar clinical and histological picture as mentioned by Peltinato et al [3]. However the clinicoradiological behaviour and the

histological findings in our case do not conform with the tumors of the reported cases in the literature.

Poswillo [2] mentioned that at the end of 60 days gestation period, the embryo enters into foetal stage, when the influences of genetic informations are overcome by the local environmental factors. He commented that it is extremely important to protect the foetus from teratogenic insults to avoid any future catastrophe. The mother of the patient narrated that during her pregnancy for the birth of the child patient she suffered from severe general body weakness, vomiting, diarrhoea etc, between 10-12 week of gestation period. She was treated with intravenous fluids, injections etc (details of treatment not known) in a private nursing home in Bihar.

Based on the above findings there is a strong presumption that the oro-facial mass in the child is a malformation of the palate, premaxilla and the upper lip. In the absence of any reported case in the literature a new terminology of the congenital deformity may be coined as-"Maxillofacial dysplasia with Cleft Palate'.

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