

Discussion

There is a wide spectrum of malformations in the oronasopharynx which occur due to disturbances in the early embryogenesis. Ewing(1) classified these malformations into three broad types—dermoids, teratomas, and epignathi. Teratomas and epignathi are regarded as true neoplasms because they display progressive and uncoordinated growth. They are derived from all three germinal layers. Dermoids, however, are strictly benign lesions with limited growth potential and are bigerminal in origin (ectoderm and mesoderm).

The dermoids arising in the oronasopharynx have also been designated as hairy polyp, complex hamartoma, inclusion dermoid, and even teratoma(2), but probably the term which explains these malformations best is 'choristoma'. Choristoma is defined as a non-neoplastic tumor-like mass of developmental origin in which are found tissues foreign to the site at which it is located(3). It differs from hamartoma which is a non-neoplastic proliferation of cells which are normally present at that site.

Chaudhry *et al.*(2) could find only 111 such lesions till 1978 and added two of their own. Few isolated case reports have been published since then(5). Only a few of these have been described in neonates and young infants. This is because many of them escape recognition due to their small size or lack of symptoms. In the neonatal period, they may present with respiratory and occasionally feeding problems. Female predominance has been reported(2). Microscopically, in addition to skin and its appendages, choristomas may contain adipose tissue, fibrovascular connective tissue, smooth and striated muscle, salivary glands tissue, nerves, lymph nodes, cartilage and bone.

The purpose of this report is to highlight the confusion regarding the terminology of the various types of malformations in the oral cavity and also stress on the necessity to recognize these 'Choristomas' as benign developmental errors.

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Giant Congenital Melanocytic Nevi

L. Kasturi
V. Mashankar
U.A. Desai
A.V. Kulkarni

Congenital melanocytic nevi are common lesions found on the skin in approximately 1% of the newborns(1). Most of the congenital nevi are small(2). The small

From the Department of Pediatrics, BARC Hospital, Anushaktinagar.

Reprint requests: Dr. L. Kasturi, 7-D Everest, Anushaktinagar, Bombay 400 094.

nevi are amenable to simple excision and primary closure. However, giant nevi occupy a major portion of large anatomic site and are difficult to remove(3). The term giant congenital melanocytic nevi is used for nevi measuring more than 20 cm.

Although easy to diagnose the management of congenital nevi is complicated because of the risk of developing malignant melanoma. The risk of developing melanoma in giant congenital melanocytic nevi is 2-42%(4).

Case Report

A 10-year-old Hindu girl, was referred to this hospital, with extensive blackish pigmentation of the trunk and abdomen since birth. The lesions were non-progressive. There was no itching and no change in color. She was born of nonconsanguineous marriage with an uneventful perinatal period. On examination, she had extensive hairy blackish pigmentation suggestive of giant congenital melanocytic nevi. The lesions were on the entire back, right shoulder, anterior aspect of the chest and abdominal wall as shown in the Fig. Small satellite hairy blackish pigmented lesions were also seen over the right upper limb, thigh and cheek. Few similar lesions were seen on trunk and lower limbs. No other congenital anomalies were noted. The neurological and systemic examinations were normal.

Discussion

Congenital melanocytic nevi may be classified as small if they are 1 to 1.5 cm in size, intermediate if 1.5 to 20 cm and giant if more than 20 cm in size(6). The present case had extensive lesions, suggestive of giant congenital melanocytic nevus. Histologically, congenital melanocytic nevi consists of nests of melanocytes in the



Fig. Showing giant congenital melanocytic nevus covering the anterior aspect of the chest, upper abdomen and right shoulder; satellite lesions are seen over the face and the right forearm.

epidermis, dermis or both(6). Congenital nevi may extend to subcutaneous tissue, fascia or even muscle.

These nevi are characterized by medium to dark brown color, a verrucous or lobulated surface and a coarse terminal hair growth. Giant congenital melanocytic nevi frequently show peripheral satellite nevi(7) as was observed in this case. Giant congenital melanocytic nevi involving the scalp and neck may be associated with

lepto-meningeal melanocytosis resulting occasionally in the development of hydrocephalous, seizures and leptomeningeal melanoma(5,7).

Other associated abnormalities include spina bifida or meningocele (when the lesion is over the spine), club foot, hypertrophy or atrophy of the deeper structures of a limb(5,7). These associated anomalies were not observed in the present case, though the skin over the spine was involved. The course is fairly stable in most cases as has been observed in the present case. They may elevate with time, change texture, develop irregular nodularity, variably darken or lighten. Lesions very rarely regress.

Giant congenital melanocytic nevus runs a significant risk of malignant melanoma in 6-10% of the cases(7). Because of the high risk of malignant degeneration during childhood early excision has been recommended(2,4). Aggressive surgical excision early in life is often not practical because of involvement of vital structures as well as the functional and cosmetic disability caused by excising large cutaneous areas.

It is recommended to follow up patients every 6-12 months interval to observe for changes like asymmetry, border irregularity, localized color variation to red, dark brown, blue or black, changing size of atypical area and for appearance of elevated firm nodules(6).

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Esophagopleural Fistula Complicating Suppurative Lung Disease

V. Kohli

R.K. Marwaha

K.L. Narasimhan

R.P.S. Bajwa

S.K. Mitra

Acquired esophagorespiratory fistulas are rare in childhood. They may be esophagobronchial or esophagopleural, the

From the Departments of Pediatrics and Pediatric Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh 160 012.

Reprint requests: Dr. R.K. Marwaha, Additional Professor, Department of Pediatrics, PGIMER, Chandigarh 160 012.