Heterotopic Brain Presenting as a Lip Lesion

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Heterotopic brain tissue in the upper lip of a newborn child is presented and discussed. This rare developmental anomaly is usually present at birth and may simulate hemangioma. Before any surgical procedure can be performed, thorough radiographic and neurosurgical examination is essential to rule out eventual communication of the tumor with intracranial space.

KEY WORDS: *heterotopic brain tissue, glioma in the upper lip*

Hetertopic brain is mature brain tissue that appears in areas distant to the central nervous system. A recent review of the literature reveals multiple extracranial locations of heterotopic brain tissue including the nose (New and Devine, 1947: Black and Smith, 1950: Dawson and Mun, 1956; Crosby, 1957; Walker and Resler, 1963; Christianson, 1966; Vistnes et al, 1968; Lowe et al, 1971; Kubo et al, 1973; Krebs et al, 1976; Kopf and Bart, 1978; Goravalingappa et al, 1979; Gorenstein et al, 1980; Hughes et al, 1980; Gopal 1981; Brunsting, 1981; Mirra et al, 1981; Azumi et al, 1984; Swift and Singh, 1985), nasopharynx (Low et al, 1956: Zarem et al, 1967; Cohen and Abt, 1970; Okulski et al, 1981; Feldman et al, 1982; Seibert et al, 1984; Ruff and Diaz, 1986), soft palate (Shapiro and Mix, 1968; Ibekwe and Ikerionwu, 1982), tongue (Ofodile et al, 1982), scalp (Lee and McLaurin, 1955; Orkin and Fisher, 1966; Zook et al, 1984), orbit (Emamy and Ahmadian, 1977; Call and Baylis, 1980; Newman et al, 1986), face (Kern and MacDonald, 1961; Kurzer et al, 1982), and lungs (Kanbour et al, 1979; Gonzalez-Crussi et al, 1980). Heterotopic brain has also been observed intracranially but extracerebrally, (Marubayashi and Matsukado, 1978). There is no report in the literature of heterotopic brain tissue located in the lip, although meningeal heterotopics have been noted adjacent to or in the midline (Orkin and Fisher, 1966; Lopez et al, 1974).

The purpose of this report is to present clinical and histopathological findings of heterotopic brain tissue of the lip as well as to discuss briefly its pathogenesis.

CASE REPORT

A newborn white male was referred to us with a tumor approximately 3 × 3 cm on the upper lip (Fig. 1). This tumor was partially purplish superiorly and yellowish inferiorly. The mass, located just to the left of the midline consisted of two components. The superior aspect was firm and blanched on gentle pressure. The inferior aspect was soft and spongy on palpation. On deep palpation, a bony small element was noted centrally. The tumor created distortion of the lower lip secondary to compression as well as displacement of the left alar base superiorly and obstruction of the left nasal airway. Eversion of the mass revealed a depression in the maxillary alveolus, but no abnormality of either the hard or soft palates. The lateral aspect of the lip measured approximately 20 mm to a peak from which the vermillon of the lateral lip segments continued inferior to the mass. The remainder of the physical examination was normal.

Computerized axial tomography did not show intracranial communication of the mass. The cribriform plate and cranial base were completely intact, and no evidence of sinus tract was visible in the nose or midface. A fragment of bone was noted within the lesion confirming palpation (Fig. 2). At 4 months of age the infant was taken to the operating room and under general anesthesia a thorough examination revealed that the lateral lip segments, commissure to peak,

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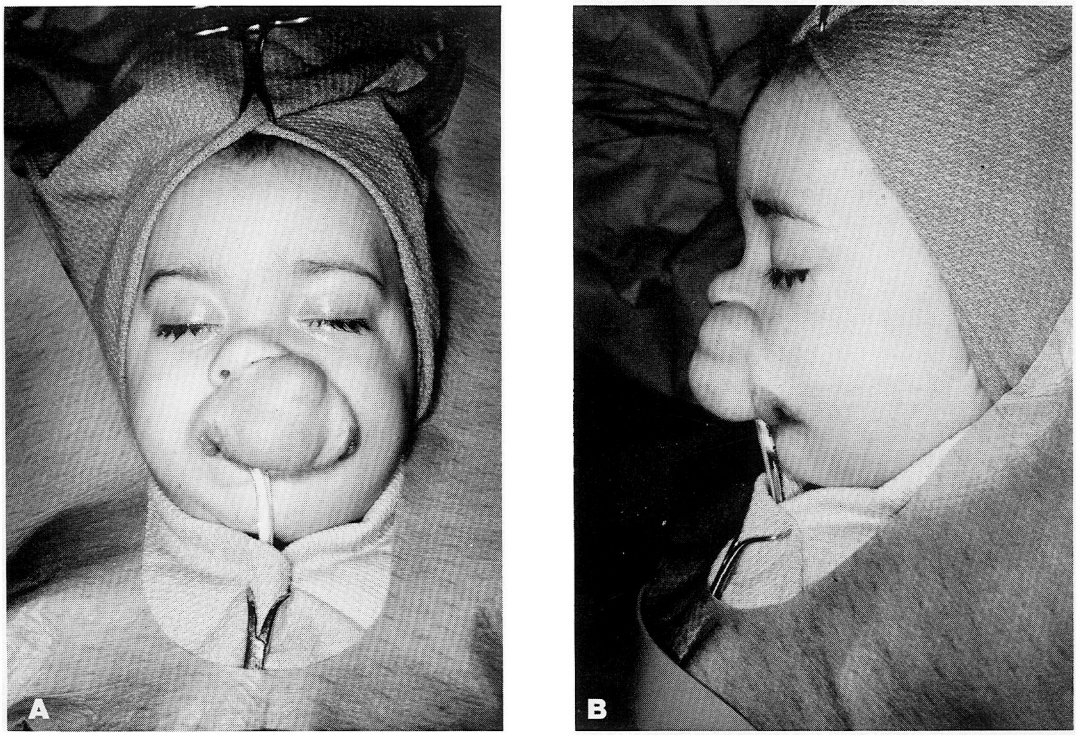

FIGURE 1 Heterotopic brain tissue on the upper lip in a male infant: *A*, frontal view; *B*, lateral view.

measured 20 mm bilaterally. A distorted Cupid's bow appeared to be present on the right with an intact right philtral column, but no left philtral column was identified. The distance from alar base to oral commissure was 28 mm on the left, and 13 mm on the right. The mass was surgically explored, with the two components excised as well as the segment of bone, and sent to pathology for histologic evaluation. The bone was connected to the maxilla but not connected to the tumor. The lip repair was then performed in a fashion similar to that employed for closure of a unilateral cleft lip, with orbicularis muscle reapproximated across the midline. A wedge of skin and subcutaneous tissue inferior to the left alar base was then excised to help minimize the vertical asymmetry (Fig. 3).

Postoperatively, the patient remained free of airway obstruction. A slight asymmetry of the left alar base and lower lip were still evident, and prominence of the left maxilla was also noted at 3 years of age (Fig. 4). It is anticipated that these can be further improved at a later time.

Histopathologically, the firm superior portion of the tumor was composed of neural tissue (glial cells and intercellular glial substance), which was separated by dense bands of collagen resembling fascia or even meninges (Figs. 5 and 6). The soft, inferiorly located mass, also showed neural tissue between bands of collagen fibers and fibroblasts. Neuroglial tissue of this part of the tumor was covered from the top by the dermis and subcutis with lobulated adipose tissue (Fig. 7). No tissue consistent with dermoid cyst, teratoma, or facial duplication was identified.

DISCUSSION

Heterotopic brain tissue is an extremely rare developmental anomaly in which brain tissue located outside the cranial cavity forms an asymptomatic tumor. This tumor usually is firm, smooth, incompressible, red to blue, and slow growing. Most frequently this tumor appears on the bridge of the nose in the newborn resembling hemangioma (Christianson, 1966; Kopf and Bart, 1978; Brunsting, 1981). When located intranasally, such tumor may cause respiratory obstruction (Gopal, 1981; Mirra et al, 1981; Seibert et al, 1984) and deformity of the nasal bone (Baran et al, 1973). In our case, tumor of the upper lip did cause obstruction of the left nasal airway and nasal deformity.

This entity was first described in 1852 by Reid, and subsequently many synonyms associated with such lesions have appeared in the literature: ganglioma, astrocytoma, fibroglioma, ganglioneuroschwannospongioblastoma, ganglioblastoma, glioma, nasal glioma, encephalo choristoma-naso-frontalis, encephaloma, encephalocoele, and choristoma (Walker and Resler, 1963).

Heterotopic brain tissue is composed predominantly of glial cells (or astrocytes) within intercellular glial substance and sometimes focal clusters of neurons (Feldman et al, 1982)

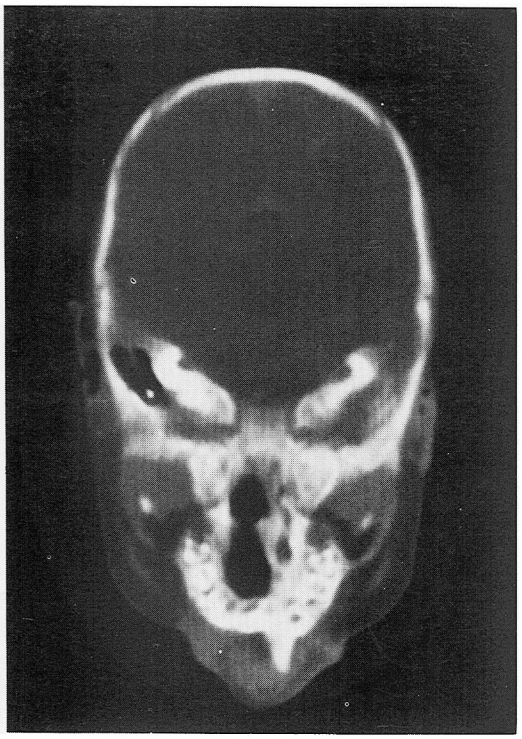

FIGURE 2 Computerized tomography scan of the head at the level of the lesion. Note fragment of bone connected to the maxilla.

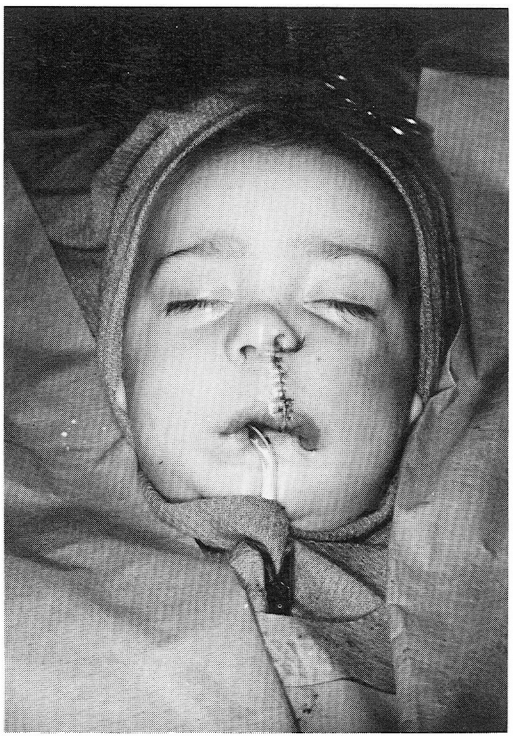

FIGURE 3 Result immediately after excision of heterotopic brain tissue from upper lip.

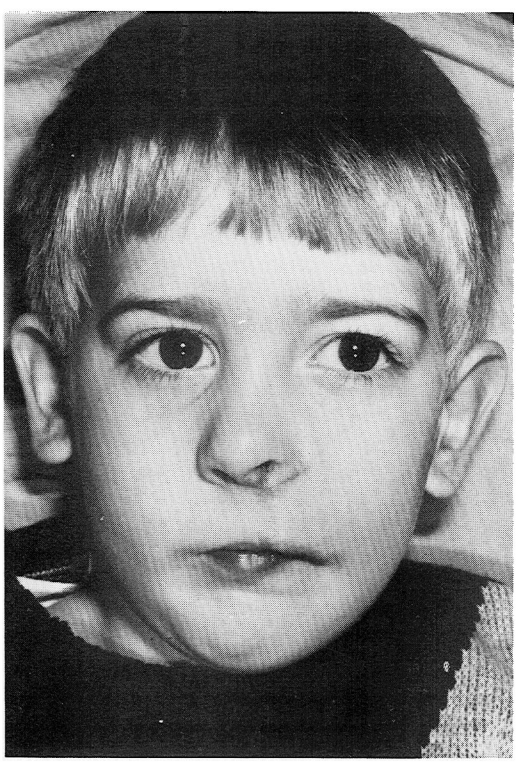

FIGURE 4 The same child 3 years after excision of heterotopic brain tissue from upper lip.

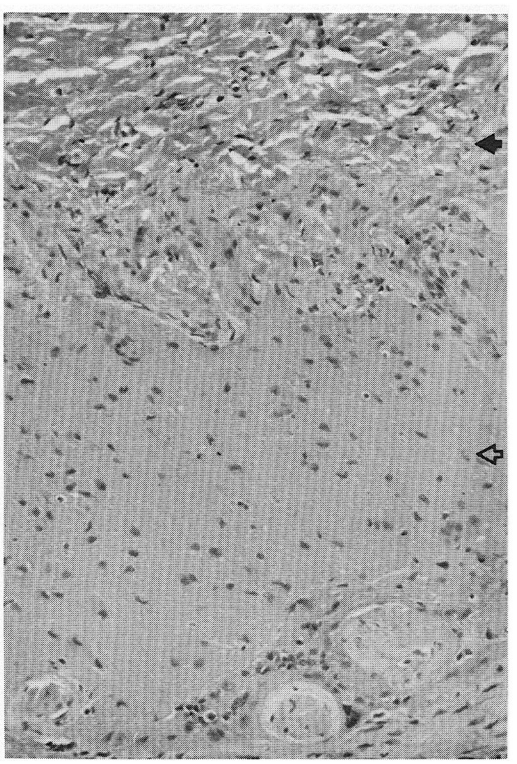

FIGURE 5 Micrograph of firm portion of the tumor found on the upper lip. Neural tissue (open arrow) is covered by dermal collagen fibers (black arrow). (Hematoxylin and eosin. Original magnification 117 ×).

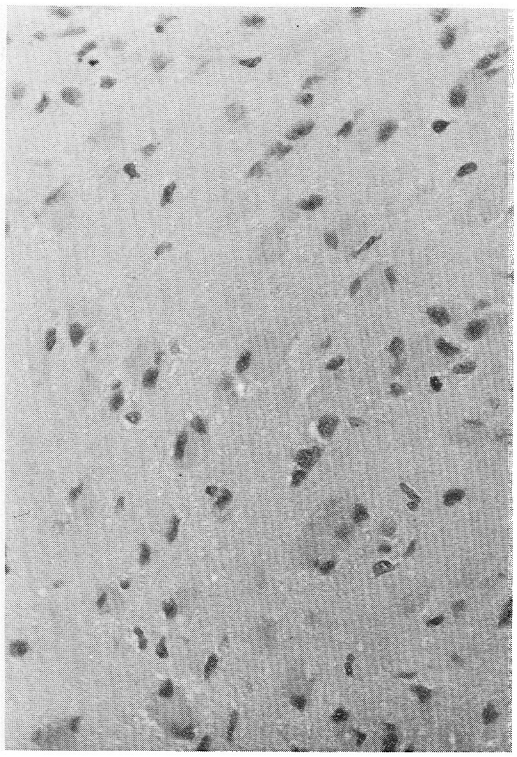

FIGURE 6 Higher magnification shows glial cells and intercellular fine fibrillar glial substance. (Hematoxylin and eosin. Original magnification 315 ×).

FIGURE 7 Histopathology of soft part of the tumor. Neural tissue (open arrow) separated by fibrous connective tissue is in the vicinity of subcutaneous adipose tissue (black arrow). (Hematoxylin and eosin. Original magnification 63 ×).

whose cytoplasm often contains Nissl's granules (Mirra et al, 1981). Scattered foci of calcification in some lesions have also been observed (Mirra et al, 1981; Zook et al, 1984). Diverse theories exist to explain the origin of these tumors, but there is general agreement that they are developmental rather than neoplastic lesions. The most commonly accepted theory about the pathogenesis of these tumors is that an encephalocoele loses its intracranial connection during the course of fetal development (Gorenstein et al, 1980; Hughes et al, 1980). In 15 percent of the cases with heterotopic brain tissue on the nose, communication with the intracranial space was present (Kubo et al, 1973). Incisional biopsy in these cases may cause cerebrospinal fluid rhinorrhea with fistula and meningitis (Gorenstein et al, 1980) or intraoperative cerebrospinal fluid leak (Grundfast et al, 1986). Thus, radiography, including CT scan, brain scintigraphy or angiography, and neurosurgical consultation must always precede surgical intervention (Kopf and Bart, 1978; Gorenstein, 1980), with the mode of therapy dependent on the outcome of these detailed studies.

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