Heterotopic Neuroglial Tissue Associated with Bilateral Palatine Cleft

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Isolated heterotopic lesions of neuroglial tissue may rarely present in the head and neck, or they may be associated with other congenital deviations. In this article we present the case of a 7–month-old patient with a central lesion that emerged through the anterior part of a bilateral palatine fissure.
Keywords: *Heterotopic neuroglial tissue, children*
J Clin Pediatr Dent 32(4): 305–308, 2008

INTRODUCTION

Heterotopic central nervous system (CNS) tissue at an extracranial site is rare, which is defined as the presence of normal neural tissue outside the cranial cavity with or without being associated with the meninges. When it does occur, it presents in locations adjacent to the cranial base, such as the nasopharynx, the orbit, and the mastoid or auricular region. Exceptional cases have been reported in distant locations such as the tongue, lips, and lungs. Early descriptions of this lesion classified it as a variant of the encephalocele; also, the terms *glioma* or *gliomatosis* have been used to describe similar lesions.[1-2]

Heterotopic neuroglial tissue can be an isolated entity or, in 25 percent of the cases, it can be related to other anomalies of the CNS and head and neck syndromes. The association of this lesion with patients suffering from palatine fissure is not frequent.[3]

In this article we present a description of a case of heterotopic neuroglial tissue emerging through the septum in the bilateral palatine fissure of a 7-month-old patient, and a review of the literature.

CASE REPORT

A 7½-month-old female was referred by the Pediatrics and Otolaryngology Departments, having been diagnosed with complete bilateral palatine fissure (CBPF), associated with a mucous retention cyst in the nasal septum. Hereditary antecedents were negative. The patient was the daughter of young parents, with no blood relation between them. It was the first pregnancy, with no complications during gestation. Normal birth; weight 3.475 kg; no signs of neonatal hypoxia. Since birth, a bilateral palatine fissure was observed. The patient was sent to the Department of Maxillofacial Surgery to have the defect corrected; also because a lesion, measuring approximately 4 x 2 cm, was observed protruding through the complete palatine cleft. The neurological exam revealed slight psychomotor slowness with horizontal nistagmus. The preoperative exams were normal. The patient was scheduled for surgical intervention to remove the lesion and close the palatine fissure.

Once the patient was in the operating room under general anesthesia, the operating crew proceeded to review the defect, which was a central lesion in the palatal midline,

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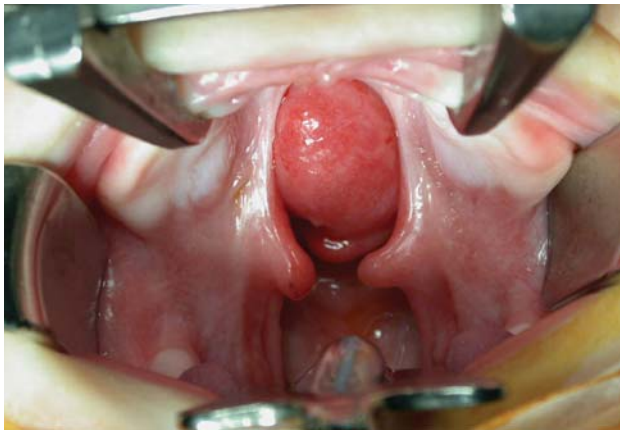

Figure 1. Intraoral aspect of lesion protruding through the palatine fissure.

sessile, and pulsating (Fig 1). The surgeons first aspirated this lesion, obtaining 3 mL of a clear and crystalline-liquid, thought to be cerebrospinal fluid.

By means of reactive strips, a concentration of 112 mg/dL of glucose was obtained. The surgery was postponed while the Department of Neurosurgery was called in for consultation. Magnetic Resonance Imaging (MRI) showed a large defect beginning in the lower portion of the knee of the callous body extending caudally at the level of the midline, in front of the hypothalamus toward the base of the skull. It projected through the anterior clinoid and prolonged itself through the palate.

A tentative diagnosis of glioma was made (Fig 2). Later, the Neurosurgery Department proceeded to close the neurological defect and take a biopsy, which tested positive for neuroglial heterotopy (Fig 3 A-F). The Department of Maxillofacial Surgery closed the palatal defect using the Veau-Wardill technique. After surgery, the patient recovered without complications, and was discharged from the hospital 4 days later. A month after surgery the patient was doing well, but postoperative follow-up was not possible.

DISCUSSION

The discovery of heterotopic neuronal tissue in the palatine region associated with fissures is very rare.[4] Uemura et al, in an extensive review of world literature in 1999 with relation to this condition found only 7 cases involving the palatine fissure.[5] Giannas et al presented 1 more case in 2004, just as Anjaneyulu et al did.[2,6]

These defects can present themselves at any age, but frequently they are diagnosed during infancy. In prenatal studies, its diagnosis is difficult except in the case presented by Cohen and Abt of a gestational process associated with polyhydramnios.[7]

Heterotopy has been considered a unique condition, but in a limited number of cases it may be accompanied by micrognathia, glossopexy, bifid uvula, pectum excavatum, polidactily, anencephaly, cardiovascular anomalies, or broncopulmonary displasia.[2,3]

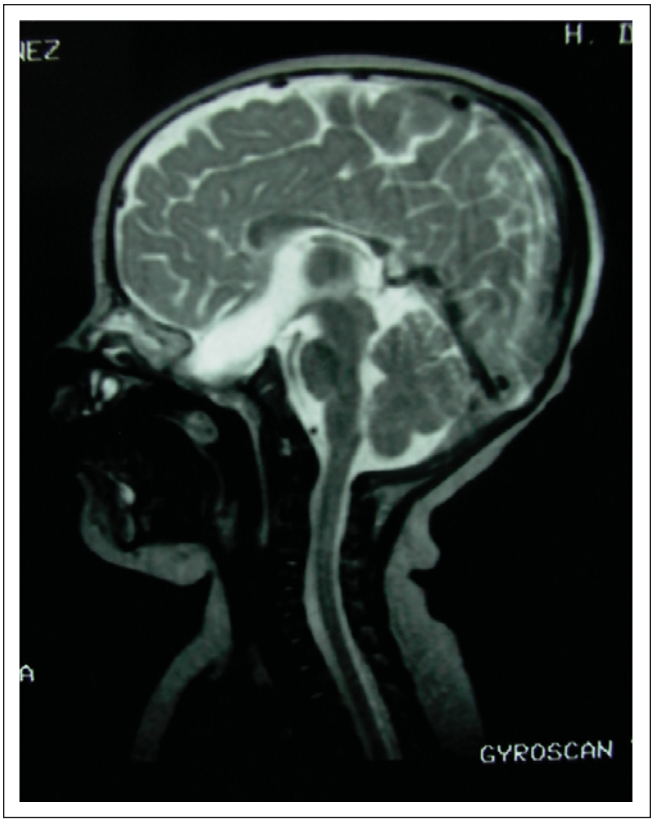

Figure 2. The contrasted Magnetic Resonance Imaging that shows the infundibulum of the hypophysis visibly elongated accompanying the lesion that extends caudally through the palate.

Clinically, neuronal heterotopias can simulate other congenital anomalies in the head and neck, such as lymphatic lesions, encephaloceles, meningoceles, teratomas, branchial cysts, and mucous retention cysts, as in the present case, where the lesion protruded into the oral cavity.[1-6]

The mechanism that generates this anomaly is uncertain. Some have tried to explain its relation in patients with cleft lip and palate as an entrapment of neuronal tissue during embryonic development in inadequate sites. Giannas et al[6] have described 3 additional elements: (a) encephaloceles that lose their intracranial connections, (b) islands of totipotential cells that generate glial tumors, and (c) abnormal entrapment or migration of glial tissue beginning in the olfactory bulbs.

Recent studies have made several advances in understanding the mechanisms involved in the disordered patterns of neuronal migration, making it possible to determine at the molecular level why neuronal and glial cells migrate differently, generating malformations. Kanatani et al report that cortical formation in the developing brain is a highly complicated process involving neuronal production, interaction of radial glia with neuronal migration, and multiple modes of neuronal migration. This pattern is affected by changes in the molecules that control migration, such as cyclooxygenase,[5] reeling transmitters, and double chain.[8]

Neuroglial tissue can contain solid and cystic elements as a result of the production of cerebrospinal fluid by the choroid plexus.[9] The tissue is usually firm, lobulated of

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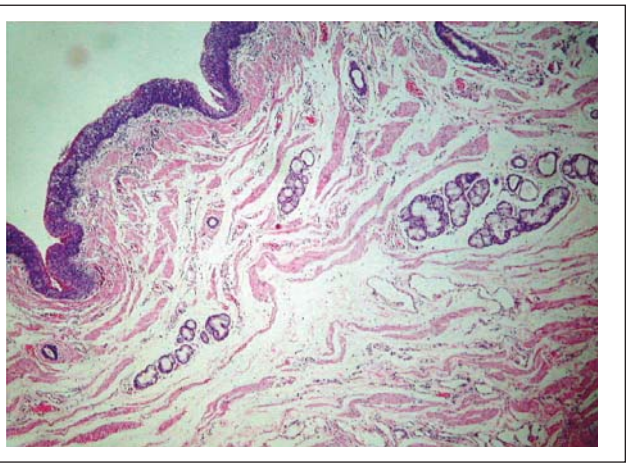

Figure 3-A. Nasal mucosa. Central nervous tissue trabeculae intermingled with fibro/adipose tissue of the submucosal. H&E 4X.

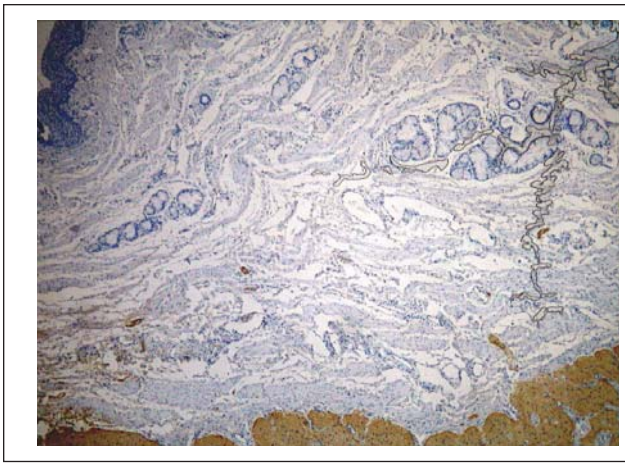

Figure 3-B. Same field. Immunohistochemical image for the specific Neuronal Enolasa 4X.

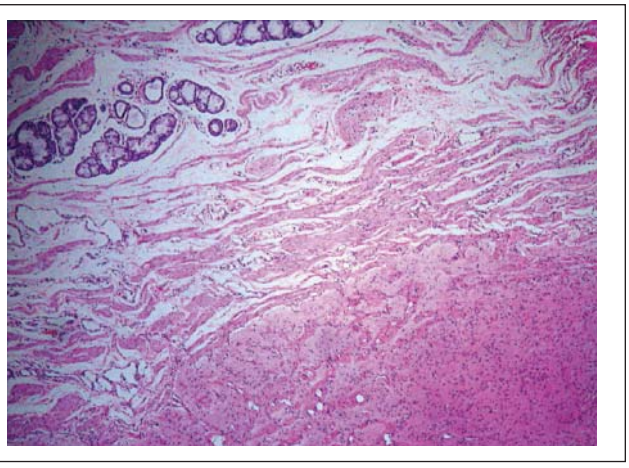

Figure 3-C. Neuronal – glial tissue in nests and fasciculus. Without meninges, between the glandular submucosal acini. H&E 10X

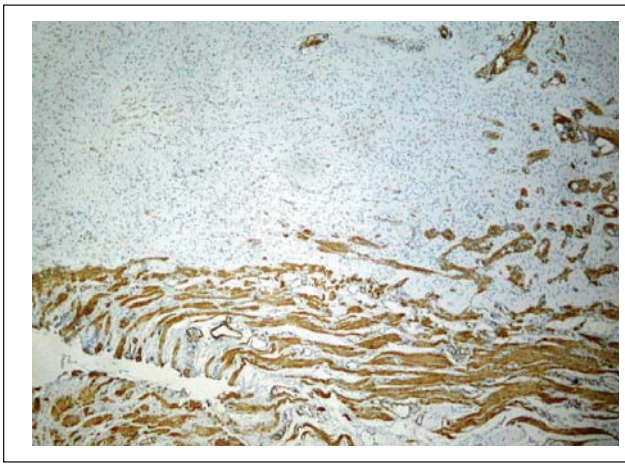

Figure 3-D. Areas with edema, fasciculus of fibrillar astrocyte. PGFA 10X

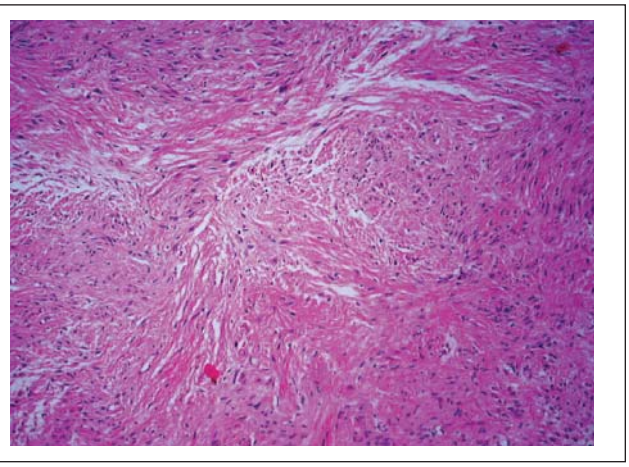

Figure 3-E. Astrocyte fasciculus and oligodentrocytes in a fibrillar bottom H&E 40X.

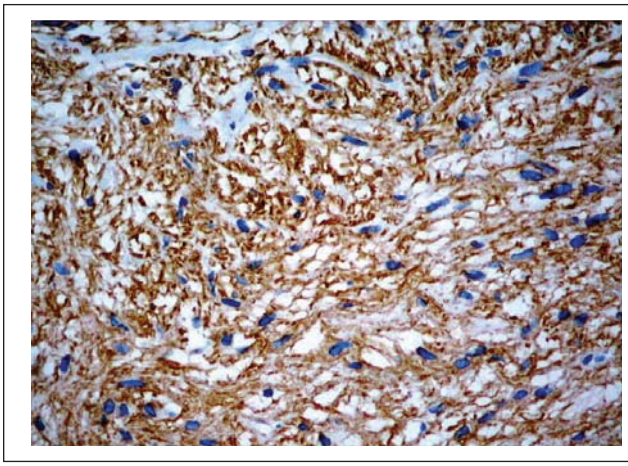

Figure 3-F. Immunohistochemical image for the PGFA 40X.

elastic consistency, and of variable size that can reach several centimeters. Histologically, neuroglial tissue can contain mature glial elements, ependym, and choroid tissue intermingled with nasal and buccal mucosa,[10] as in our patient, where CNS tissue trabeculae can be seen intermingled with glandular acinar and fibroadipose tissue of the nasopharyngeal submucosa.

Computerized tomography and MRI are indispensable in

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determining the origin and size of the lesion and can also help establish the possibility of a frank intracranial communication, which must be considered in central pulsating lesions. Aspirating punctures in lesions suspected of being cystic assist in diagnosis but are not concluyent. There is also the risk of creating fistulas of cerebrospinal fluid that could complicate patient management and prognosis.[11]

Once the diagnosis is confirmed, surgical removal of small lesions must take place. Recurrences are rare and are mostly due to primary incomplete removals. In cases having intracranial communication, craniotomies are needed to close the defects of the base; external resection of the lesion remnants is also needed. Up until now there have been no reports of neoplasic transformation.[12,13]

ACKNOWLEDGMENT

We would like to thank Norman Wahl, for his assistance in editing this manuscript.

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