Hemifacial hyperplasia is a rare developmental anomaly characterized by asymmetric growth of ≥1 part of the face. In this report of a female child born with facial asymmetry, CT and MR imaging revealed predominantly lipomatosis. The case suggests that hemifacial lipomatosis may be a subtype of partial hemifacial hyperplasia. We discuss the differential diagnosis of HFH and emphasize the importance of screening for embryonal malignancies.

Case Report
A 15-month-old female child presented to our department with right-sided facial fullness (Fig 1). The abnormality was noted at birth, and the child’s parents stated that the fullness did not fluctuate in size. The right cheek appeared to grow at a rate similar to the left side. Physical examination was also remarkable for a slightly enlarged right auricle. There were no other significant medical problems. No biochemical or chromosomal abnormalities were detected. The child was otherwise healthy. A sonographic examination showed diffuse right facial adipose hypertrophy but no discrete vascular malformation.

CT examination at this time demonstrated predominantly increased fatty tissue on the right side of the face. The distribution of fat was largely in the malar region but also infiltrated the ipsilateral muscles of mastication and the parotid gland and extended into the ipsilateral parapharyngeal fat. The right parotid gland and muscles of mastication had a slightly marbled appearance, and there was enlargement of the right prestyloid parapharyngeal space (Fig 2A). These findings were demonstrated again on a follow-up MR imaging examination 4 months later (Fig 2B).

The bony structures of the right face showed slight asymmetry, with hypertrophy of the right maxilla and the right lateral pterygoid plate. There was also subtle enlargement of the right zygoma and the right mandible (Fig 3). Regarding dental development, there was slight early rupture of the right maxillary deciduous canine teeth, and enlargement of the premolar and molar teeth was also noted (Fig 4).

MR imaging examination performed as a 6-month follow-up revealed stable and similar findings (Fig 5). No abnormal enhancement was identified. No discrete fibrous capsule to suggest an encapsulated lipoma was seen. The tongue appeared symmetric; however, accurate assessment was difficult because the patient was intubated for the study. Findings of an MRA examination were within normal limits, and no intracranial abnormalities were demonstrated. The child was otherwise developing normally and achieving normal milestones.

Discussion
HFH was first described by Meckel in 1822.1 It is a rare morphologic developmental anomaly characterized by asymmetric growth of ≥1 body part. Its true incidence is unknown and, to our knowledge, has not been published, and very little about this syndrome has been published in the radiology literature. Although it is often called HFH, it may be more accurately termed “hemifacial hyperplasia” to reflect the underlying pathology.2,3 Little is known about the etiology of this condition. Various theories, none clearly favored, have been proposed. These include vascular lymphatic, hormonal, and asymmetric.

Received July 8, 2009; accepted after revision July 15.
From the Departments of Radiology (P.B.-H., P.S.) and Plastic Surgery (P.T.), Mount Sinai School of Medicine of New York University, New York, New York.

DOI 10.3174/ajnr.A1857
development of the neural fold, with hyperplasia of neural crest cells. It is a condition that appears to affect the derivatives of the first pharyngeal arch, though the middle ear structures seem spared.

Isolated hemifacial hyperplasia should be a diagnosis of exclusion, because there are other more clinically burdensome conditions that can cause facial asymmetry. These conditions include the following: Proteus syndrome, BWS, neurofibromatosis type 1, and vascular malformations such as Klippel-Trenaunay syndrome and lymphaticovenous malformations.

A full neurologic examination and brain imaging are prudent, and the radiologist may be the first to suggest screening for organomegaly and malignancies. Tumor surveillance, especially for malignancies of embryonal origin, is supported by a prospective study of children with isolated hemihyperplasia or hemihypertrophy. These tumors include Wilms’ tumors, hepatoblastoma, and adrenal cell carcinoma.

The extremely rare Proteus syndrome, suspected as the primary condition in the historic “elephant man,” causes a progressive overgrowth of all soft tissues and bones, including the skull, not limited to the region of isolated hemifacial hyperplasia. Patients with Proteus syndrome will often have involve-
ment of the hands and feet and veracious epidermal nevi.\textsuperscript{7} BWS is a congenital cancer-predisposition syndrome associated with macroglossia, macrosomia, ear pits or ear creases, and midline abdominal wall defects. The associated hemihy-

pertrophy seen in BWS may represent the mosaic form of macrosomia.\textsuperscript{8} The hemihypertrophy in neurofibromatosis type 1 can be due to even subtle plexiform neurofibromas and/or skeletal dysplasia. When vascular malformations result in facial asymmetry, it may be due to the bulk of abnormal vessels alone or combined with induced hypertrophy due to local increased vascular flow and local lymphedema.\textsuperscript{9} Occasionally, hemifacial atrophy (Parry-Romberg syndrome) can give the false appearance of hemifacial hyperplasia on the normal side, though these patients present later, between 5 and 15 years of age.

Rowe\textsuperscript{10} in 1962 classified HFH into THFH and PHFH. THFH manifests unilateral enlargement of all the viscerocrania
tal structures bounded by the frontal bone, excluding the eye; inferiorly by the inferior border of the mandible and the mid-
line; and laterally by the ear. THFH is characterized by en-
largement of the teeth, bones, and soft tissues. Not all the included structures are enlarged in PHFH, as in our patient. Other findings in hemifacial hyperplasia include an enlarged tongue on the involved side, enlarged lips, and widening of the palate of the affected side.\textsuperscript{5}

Our case, similar to a previous case report,\textsuperscript{11} demonstrated lipomatosis as the dominant feature and lacked significant facial musculature hyperplasia. Hemifacial myohyperplasia is a proposed newly described separate entity in which facial muscular hyperplasia is the dominant feature.\textsuperscript{3,4} This may explain the lack of the nasal deviation toward the affected side in our patient but rather deviation to the opposite (normal) side. Nasal deviation toward the affected side may be observed when the facial musculature around the nasolabial fold is in-
volved and when the hypertrophied turbinates cause nasal septal deviation.\textsuperscript{4} The teeth, mandible, and maxilla should be carefully scrutinized for deformities to help one reach the cor-
rect diagnosis. This is especially important if only lipomatous overgrowth is seen initially.

Conclusions
Hemifacial hyperplasia is a rare developmental condition of asymmetric growth of $\geq 1$ body part. MR imaging and CT illustrate hemifacial lipomatosis as the dominant feature, sug-
gest ing that it may represent a subtype of partial hemifacial hyperplasia.

References
1. Gorlin RJ, Meskin LH. Congenital hemihypertrophy: review of the literature and report of a case with special emphasis on oral manifestations. J Pediatr 1962;61:870–79
2. Yoshimoto H, Yano H, Kobayashi K, et al. Increased proliferative activity of osteoblasts in congenital hemifacial hypertrophy. Plast Reconstr Surg 1998;102:605–10
3. Lee S, Seo R, Murakami C, et al. Hemifacial myohyperplasia: description of a new syndrome. Am J Med Genet 2001;103:326–33
4. Urban PP, Roland B. Congenital isolated hemifacial hyperplasia. J Neurol 2009;256:1566–69. Epub 2009 May 8
5. Lawoyin JO, Daramola JO, Lawoyin DO. Congenital hemifacial hypertrophy: report of two cases. Oral Surg Oral Med Oral Pathol 1989;68:27–30
6. Hoyne HE, Seaver LH, Jones KL, et al. Isolated hemihyperplasia (hemihypertrophy): report of a prospective multicenter study of the inci-
dence of neoplasia and review. Am J Med Genet 1998;79:274–78
7. Zingade ND, Zingade NN. A rare case of Proteus syndrome. J Laryngol Otol 2008;122:e7
8. DeBaun MR, Niemitz EL, McNeil DE, et al. Epigenetic alterations of H19 and LIT1 distinguish patients with Beckwith-Wiedemann syndrome with cancer and birth defects. Am J Hum Genet 2002;70:604–11
9. Blei F. Vascular anomalies: from bedside to bench and back again. Curr Probl Pediatr Adolesc Health Care 2002;32:72–93
10. Rowe NJ. Hemifacial hypertrophy: review of the literature and addition of four cases. Oral Surg Oral Med Oral Pathol 1962;15:572–87
11. Sugiyama M, Tanaka E, Ogawa I, et al. Magnetic resonance imaging in hemi-
facial hyperplasia. Dentomaxillofac Radiol 2001;30:235–38