HATS Syndrome: Hemimaxillary Enlargement, Asymmetry of the Face, Tooth Abnormalities, and Skin Findings

Jasem M. Alshaiji, MD; Marc Z. Handler, MD; Ran Huo, MD; Ann Freedman, DMD; Lawrence A. Schachner, MD

**Practice Points**
- Appropriate management and treatment of hemimaxillary enlargement, asymmetry of the face, tooth abnormalities, and skin findings (HATS syndrome) requires a multidisciplinary team including a dermatologist, dentist, radiologist, and orthopedic surgeon.
- Becker nevus is among the most common cutaneous manifestations of HATS syndrome and can be treated effectively with the Q-switched laser or the erbium:YAG laser.

Hemimaxillary enlargement, asymmetry of the face, tooth abnormalities, and skin findings (HATS syndrome) is a rare developmental disorder involving the first and second branchial arches. Physical manifestations may present at birth or during early childhood. Characteristic findings include unilateral abnormalities of the face involving the bones, teeth, gums, and skin. Among the characteristic cutaneous manifestations of HATS syndrome, Becker nevus is the most common. A variety of modalities have been utilized in the treatment of HATS syndrome, but no standardized therapy has been established. We report a case of this rare condition in a 14-year-old adolescent boy.

**Case Report**
A 14-year-old adolescent boy presented to the dermatology clinic at our institution for evaluation of a hyperpigmented hairy patch on the right side of the face that had been present since birth. The patient reported the lesion originally had involved the right cheek, neck, and back but had gradually expanded to include the right side of the upper lip and oral mucosa. His medical history was remarkable for acne, which was currently being managed with topical treatments. There was no family history of similar conditions. There were no mental or developmental deformities since birth.

Physical examination revealed a hyperpigmented patch with hypertrichosis on the right side of the body involving the back, neck, and cheek (Figure 1), as well as hyperpigmentation involving the right side of the upper lip and oral mucosa (Figure 2A). Slight facial asymmetry also was noted. Dental examination revealed irregular spacing and decreased growth of the teeth on the right side of the mouth (Figure 2B).

A biopsy of the hyperpigmented patch on the back revealed mild regular acanthosis, basal hypermelanosis, slight papillomatosis, and hair structures within the dermis with features that were consistent with a Becker nevus. A dental radiograph demonstrated hyperplasia of the right maxillary alveolus and basal bone area with 2 missing permanent teeth.
(fourth and fifth premolars) (Figure 3). Computed axial tomography revealed enlargement of the maxillary bone on the right side.

The constellation of clinical, histopathologic, and radiologic findings was consistent with a diagnosis of hemimaxillary enlargement, asymmetry of the face, tooth abnormalities, and skin findings (HATS syndrome). The treatment plan involved surgical modification of the maxillary bone to correct the hyperplasia on the affected side and implanting 2 artificial premolars. Additionally, laser therapy using a Q-switched ruby laser, frequency-doubled Nd:YAG, 1550-nm erbium-doped fiber laser, or 755-nm alexandrite laser was considered to treat the hyperpigmentation associated with the Becker nevus.

Comment
HATS syndrome is a rare, local developmental defect involving the first and second branchial arches. It generally is detected at birth or in early childhood and is associated with unilateral abnormalities of the bones, teeth, gums, and skin. It is more common in boys than girls (1.8:1.0 ratio), with an age range of 2 to 28 years; there is a peak in the first decade of life. It was first described by Miles et al in 1987 in a case of congenital mild facial asymmetry, unilateral enlargement of the maxillary gingiva and alveolar bone, hypoplastic teeth, and hypertrichosis in the affected area. The investigators at that time suggested the term hemimaxillofacial dysplasia (HD). In 1990, Danforth et al reported 8 additional cases with similar features but without known skin changes; they proposed the term segmental odontomaxillary dysplasia (SOD). In 1996, Desalvo et al reported a case of SOD involving a 7-year-old girl with an area of hypopigmentation of the lip on the affected side, and Packota et al described the radiographic features of 12 cases of SOD. In subsequent years, other cases of HD or SOD were reported in the literature. In 2004, Welsch and Stein reported 1 patient with a Becker nevus of the skin and recommended the acronym HATS. Armstrong et al reported 2 cases of SOD with new histopathologic findings of the teeth (eg, fibrous enlargement of the pulps, an irregular pulp-dentin interface displaying many pseudoinclusions, pulp stones). In 2008, Porwal et al reported a case of HD in which maxillary hypoplasia rather than hyperplasia was noted, which emphasized the
HATS Syndrome

The differential diagnosis includes hemifacial hyperplasia, monostotic fibrous dysplasia, and regional odontodysplasia. Little information is available concerning the treatment of patients with this condition. The reported treatment modalities include combined surgical and orthodontic treatment of unerupted teeth (premolar/canine), prosthetic treatment, gingivoplasty, recontouring osteotomy for severe facial asymmetry, and reconstructive jaw surgery. Successful treatment of Becker nevi with the Q-switched ruby laser, erbium:YAG laser, and 755-nm alexandrite laser have been reported.

**REFERENCES**


