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

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## 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.*

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The authors report no conflict of interest.

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## 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

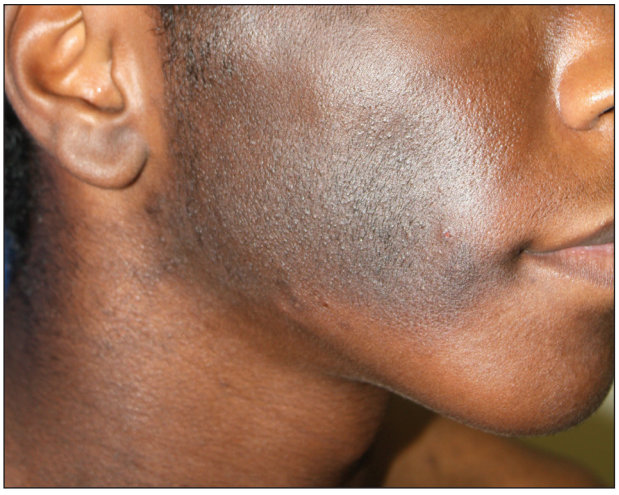
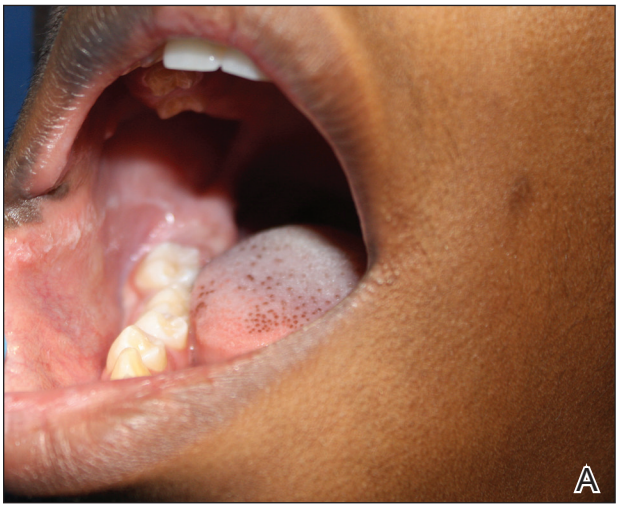


**Figure 1.** Hyperpigmented hairy patch on the right cheek (Becker nevus) in a patient with hemimaxillary enlargement, asymmetry of the face, tooth abnormalities, and skin findings (HATS syndrome).



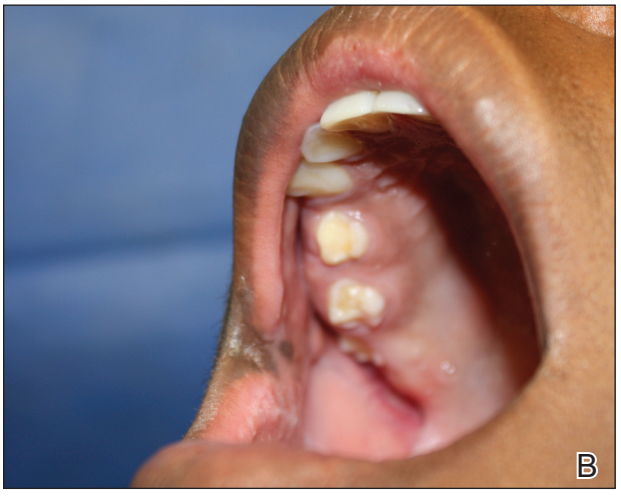




**Figure 2.** Some hyperpigmentation involving the oral mucosa on the right side (A) and dental abnormalities (B).

(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.[1] It was first described by Miles et al[2] 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[3] 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[4] 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[5] described the radiographic features of 12 cases of SOD. In subsequent years, other cases of HD or SOD were reported in the literature.[1,6-16] In 2004, Welsch and Stein[17] reported 1 patient with a Becker nevus of the skin and recommended the acronym HATS. Armstrong et al[18] 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[19] reported a case of HD in which maxillary hypoplasia rather than hyperplasia was noted, which emphasized the

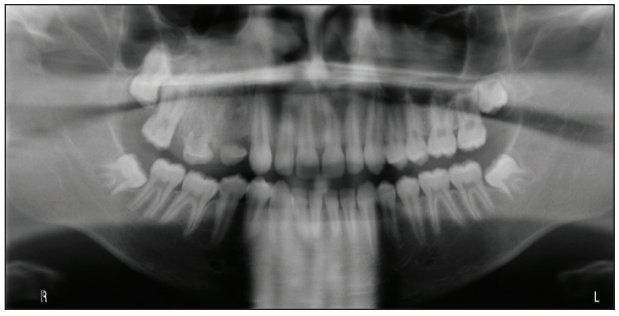


**Figure 3.** Dental radiograph demonstrated hyperplasia of the right maxillary alveolus and basal bone area with 2 missing permanent teeth (fourth and fifth premolars).

variability of the maxillary dysplasia. Koenig et al[20] reported a case of SOD with facial hypertrichosis, commissural lip clefting, and hyperlinear palms. Bhatia et al[21] reported another case of SOD with a new finding of unilateral ectopic eyelashes.

The etiology remains unknown, but theories include an alteration that occurs in utero or in infancy; the possibility of a systemic or endocrine aberration; a postzygotic mutation resulting in genotypic and phenotypic mosaicism of bone and skin, similar to McCune-Albright syndrome; and viral or bacterial infection along the branches of the maxillary division of the trigeminal nerve.[1,15] Bone defects include unilateral enlargement of the maxillary alveolar process and thickening of the vertically oriented trabeculae, which is detected radiographically. A reduction in size of the maxillary sinus and nasal airway was reported in about one-half of cases[1] and can be detected easily by computed tomography scanning. Missing permanent premolar teeth, tooth shape abnormalities, delayed eruption of teeth, abnormal spacing of teeth, hypoplastic teeth, enlarged teeth, and gingival thickening also are common oral findings.[1] The skin manifestations of HATS syndrome are not static but progress well into adolescence[15] and can include facial asymmetry, hypertrichosis, Becker nevus, hairy nevus, lip hypopigmentation, discontinuity of the vermilion border, depression of the cheek, and facial erythema.[17]

The differential diagnosis includes hemifacial hyperplasia, monostotic fibrous dysplasia, and regional odontodysplasia.[1] Little information is available concerning the treatment of patients with this condition.[15] The reported treatment modalities include combined surgical and orthodontic treatment of unerupted teeth (premolar/canine), prosthodontic treatment, gingivoplasty, recontouring osteotomy for severe facial asymmetry, and reconstructive jaw surgery.[1,6,11,15] Successful treatment of Becker nevi with the Q-switched ruby laser, erbium:YAG laser, and 755-nm alexandrite laser have been reported.[22-24]

## Conclusion

There is a need for continued reporting of cases of HATS syndrome in addition to long-term follow-up to document the natural history of the condition and to establish the appropriate treatment.

## REFERENCES


1. Friedlander-Barenboim S, Kamburoğlu K, Kaffe I. Clinical and radiological presentation of hemimaxillofacial dysplasia/segmental odontomaxillary dysplasia: critical analysis and report of a case. *Oral Surg Oral Med Oral Pathol Oral Radiol*. 2012;113:268-273.
2. Miles DA, Lovas JL, Cohen MM. Hemimaxillofacial dysplasia: a newly recognized disorder of facial asymmetry, hypertrichosis of the facial skin, unilateral enlargement of the maxilla, and hypoplastic teeth in two patients. *Oral Surg Oral Med Oral Pathol*. 1987;64:445-448.
3. Danforth RA, Melrose RJ, Abrams AM, et al. Segmental odontomaxillary dysplasia: report of eight cases and comparison with hemimaxillofacial dysplasia. *Oral Surg Oral Med Oral Pathol*. 1990;70:81-85.
4. Desalvo MS, Copete MA, Riesenberger RE, et al. Segmental odontomaxillary dysplasia (hemimaxillofacial dysplasia): case report. *Pediatr Dent*. 1996;18:154-156.
5. Packota GV, Pharoah MJ, Petrikowski CG. Radiographic features of segmental odontomaxillary dysplasia: a study of 12 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 1996;82:577-584.
6. Paticoff K, Marion RW, Shprintzen RJ, et al. Hemimaxillofacial dysplasia: a report of two new cases and further delineation of the disorder. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 1997;83:484-488.
7. Jones AC, Ford MJ. Simultaneous occurrence of segmental odontomaxillary dysplasia and Becker's nevus. *J Oral Maxillofac Surg*. 1990;57:1251-1254.
8. Prusack N, Pringle G, Scotti V, et al. Segmental odontomaxillary dysplasia: a case and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2000;90:483-488.
9. Becktor KB, Reibel J, Vedel B, et al. Segmental odontomaxillary dysplasia: clinical, radiological and histological aspects of four cases. *Oral Dis*. 2002;8:106-110.
10. Velez I, Vedrenne D, Cralle P, et al. Segmental odontomaxillary dysplasia: report of two cases and review of the literature. *Todays FDA*. 2002;14:20-21.
11. Drake DL. Segmental odontomaxillary dysplasia: an unusual orthodontic challenge. *Am J Orthod Dentofac Orthop*. 2003;123:84-86.
12. Gavalda C. Segmental odontomaxillary dysplasia. *Med Oral*. 2004;9:181.

13. Ozpinar B, Gokce B, Uzel G, et al. Prosthetic rehabilitation of segmental odontomaxillary dysplasia: seven-year follow-up. *Cleft Palate Craniofac J*. 2009;46:103-107.
14. Kuklani RM, Nair MK. Segmental odontomaxillary dysplasia: review of the literature and case report [published online ahead of print December 14, 2010]. *Int J Dent*. 2010;2010:837283.
15. Minett CP, Daley TD. Hemimaxillofacial dysplasia (segmental odontomaxillary dysplasia): case study with 11 years of follow-up from primary to adult dentition. *J Oral Maxillofac Surg*. 2012;70:1183-1191.
16. Whitt JC, Rokos JW, Dunlap CL, et al. Segmental odontomaxillary dysplasia: report of a series of 5 cases with long-term follow-up. *Oral Surg Oral Med Oral Path Oral Radiol Endod*. 2011;112:E29-E47.
17. Welsch MJ, Stein SL. A syndrome of hemimaxillary enlargement, asymmetry of the face, tooth abnormalities, and skin findings (HATS). *Pediatr Dermatol*. 2004;21:448-451.
18. Armstrong C, Napier SS, Boyd RC, et al. Histopathology of teeth in segmental odontomaxillary dysplasia: new findings. *J Oral Pathol Med*. 2004;33:246-248.
19. Porwal R, Ousterhout DK, Hoffman WY, et al. Hemimaxillofacial dysplasia: a variable presentation. *J Craniofac Surg*. 2008;19:1554-1557.
20. Koenig LJ, Lynch DP, Yancey KB. Segmental odontomaxillary dysplasia presenting with facial hypertrichosis, commissural lip clefting, and hyperlinear palm. *Pediatr Dermatol*. 2008;25:491-492.
21. Bhatia SK, Drage N, Cronin AJ, et al. Case report: segmental odontomaxillary dysplasia—a rare disorder. *Eur Arch Paediatr Dent*. 2008;9:245-248.
22. Raulin C, Schönermark MP, Greve B, et al. Q-switched ruby laser treatment of tattoos and benign pigmented skin lesions: a critical review. *Ann Plast Surg*. 1998;41:555-565.
23. Trelles MA, Allones I, Moreno-Arias GA, et al. Becker's naevus: a comparative study between erbium: YAG and Q-switched neodymium:YAG; clinical and histopathological findings. *Br J Dermatol*. 2005;152:308-313.
24. Choi JE, Kim JW, Seo SH, et al. Treatment of Becker's nevi with a long-pulse alexandrite laser. *Dermatol Surg*. 2009;35:1105-1108.