Hemifacial hyperplasia

CASE

A 49-year-old male patient presented to the clinic with a main complaint of a painless swelling of the left mandible that had been present for more than 14 years. On radiological examination, the patient presented with enlargement of the mandibular ramus, angle and corpus on the left, with a normal appearing trabecular bone pattern. The midline deviated to the right, with superior displacement of teeth 43-38 with no associated macrodontia noted (Figures 1, 2).

Figure 1: Panoramic radiographs of the patient 11 years ago (2012) at the age of 38 showing enlargement of the left ramus and corpus of the mandible.

Figure 2: Panoramic radiograph of the same patient in 2023. No significant growth occurred between the two examination periods.
The diagnosis of partial hemifacial hyperplasia was made based on the clinical and radiological findings, including localised involvement of the mandible, cessation of growth and no altered trabecular bony pattern. The patient is currently under continuous follow-up.

**INTERPRETATION**

Hemifacial hyperplasia (HFH), first described by Beck in 1836, is a rare developmental condition characterised by unilateral overdevelopment of the hard and/or soft tissue of the affected side of the face. The terms “hemifacial hyperplasia” and “hemifacial hypertrophy” have been used interchangeably, although the term “hyperplasia” is more appropriate as the tissues show an increase in cellular number rather than an increase in cellular size.

HFH may be a part of hemihyperplasia of the whole body. Asymmetry in HFH may be present at birth, but is only accentuated after puberty. The growth is proportional during this period up until cessation at adulthood. The prevalence of HFH is 1:86,000 with a male predominance. The aetiology of HFH is still unknown with endocrine dysfunction, central nervous system disorders, lymphatic or vascular malformations, somatic mutations and chromosomal abnormalities included among possible theories.

The affected side grows faster than the non-affected side, leading to asymmetry of the face. HFH can affect the facial bones, soft tissue, teeth and/or any associated structures. Unilateral enlargement of the fungiform papillae of the tongue is a common finding when the soft tissue is affected.

Additional facial characteristics on the affected side include wrinkling of the skin, smaller nasal vestibule, nasal deviation, chin deviation and inferior displacement of the auricle.

Rowe classified hemihyperplasia as follows:

- Simple hemihyperplasia (involving a single limb)
- Complex hemihyperplasia (involving half of the body on the same side)
- Hemifacial hyperplasia (involving half of the face) subclassified into:
  - true hemifacial hyperplasia (enlargement of all tissues)
  - partial hemifacial hyperplasia (not all structures are enlarged to the same degree or not at all).

The differential diagnosis to consider for HFH is CLOVES, and Kippel Trenaunay syndrome associated with fibrous dysplasia (FD) and capillary lymphatic-venous deformation. In the case of FD, an altered trabecular bony pattern is seen. Other overgrowth syndromes can also be included.

The initial radiographic method used for assessment is a panoramic radiograph. Other supporting techniques that can be used include posterior-anterior images as well as CBCT (Cone Beam Computed Tomography). The diagnosis can be made from clinical and radiographic information, given practitioners understand the differentiating criteria.

The treatment of HFH is usually for cosmetic rather than functional considerations and is based on individual assessment. Treatment includes facial contouring after growth has ceased. In paediatric cases, surgical intervention can be considered if the patient is suffering from psychosocial issues. Some cases of HFH present with progressive growth, thus long-term follow-up is required. The prognosis of HFH is good, with no reported cases showing malignant transformation.

**REFERENCES**


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**Conflict of interest**

The authors declare that they have no conflict of interest.

**Ethics approval**

This study was approved by the University of Pretoria Ethics Committee (Reference no: 562/2023). All procedures followed the ethical standards of the Helsinki Declaration of 1975, as revised in 2008. A waiver of informed consent is requested from the Ethics Committee as no identifiable features are shown/used.