

Hemifacial Hyperplasia: Two Extremes of Age Presentation

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Abstract

Congenital hemi hyperplasia is a rare congenital developmental disorder characterized by asymmetrical overgrowth of one or more body parts. Described initially by Meckel in 1822 and later reported by Wagner, et al. In 1962 Rowe classified hemi hypertrophy into complex hemi hypertrophy which involved entire half of the body, simple hemi hypertrophy affecting one or both limbs, and hemifacial hypertrophy which involved half of face. He classified hemifacial hypertrophy further into true hemifacial hypertrophy which exhibits unilateral enlargement of all tissues, teeth, bones, and soft tissues, inferior border of the mandible inferiorly, midline medially, and ear including the pinna laterally and partial hemifacial hypertrophy. Were not all structures are enlarged to the same degree or limited to one structure. Here we report two cases of true hemifacial hyperplasia with the aim to add the existence of this condition and knowledge on true hemifacial hyperplasia and its differentiating condition, along with enhanced skin pigmentation. CASE 1 A 12 years old boy reported to our Bokaro general hospital maxillofacial OPD with the chief complaint of swelling on right side of face since 3 years.

9 Patient was asymptomatic 3 years back when he complained of swelling on right side of his face. Patient gives a history of fall from stairs 5 years back while playing with his cousin. He and his mother both are unaware of side from which patient fell. According to his mother he had only small laceration on upper lip, with mild bleeding which stopped after sometime. There is no history of unconsciousness, vomiting or bleeding from any other region. Figure 1: Showing facial profile. Swelling increased from front of right ear to, right cheek region and then involvement of eye region. Increase in swelling was not associated with pain. Pain

was only experienced while touching and eating hard food rest of the time he had no complains. No significant past history of illness. No significant dental history.

On general examination patient was well oriented to person, place and time. He weight 24 kg, Height- 128.5 cm with left shoulder slightly drooped down. No abnormality detected on examination of CVS, Respiratory examination. His vital were Journal of Genetic Syndromes & Gene Therapy ISSN: 2157-7412 Journal of Genetic Syndromes and Gene Therapy Case Report This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited. J Genet Syndr Gene Ther, within normal limits. Facial examination revealed entire enlarged right half of face. Submentovertex view showing deviated nasal tip to left side, enlarged right nostril and right upper half lip. Submentovertex view showing deviated nasal tip to left side, enlarged right nostril and right upper half lip.

Temperature of right half of face was slightly raised. The facial swelling had a smooth surface, soft consistency, fluctuant. Also the swelling was not translucent. Patient also complains of recurrent redness of right eye, blurring of vision on right side for which he was referred to ophthalmologist. His right ear pinna was enlarged and tenderness at preauricular region. Intraorally there was swelling which extended from mid palatal suture to posterior palatal seal, anterior faucial pillar, maxillary tuberosity, buccal gingiva upto maxillary anteriors. Buccal sulcus not obliterated. There was spacing between the teeth of involved side, alveolar process thick especially in premolar and

molar region, midline shift was present.

A 58 year old male patient reported to our OPD with a chief complaint of painful mobile teeth after trauma. He had history of road traffic accident. The patient was well oriented with stable vital signs. On clinical examination it was seen that he had gross enlargement of right half of face .While taking history he revealed that he had asymptomatic swelling in his right cheek region since birth which had gradually increased to present size and ceased to grow when he was 20 years. There is no significant family history.

Gross swelling of right side of face. The swelling extended superiorly to upper canthus of right eye, inferiorly up to 5 cm below the lower border of mandible, anteriorly until nasolabial fold, and posteriorly till the tragus of the ear. Nose and chin were deviated to left side. Enlarged soft tissue mass was observed involving maxilla, mandible, and zygoma on the affected side. Right ear (Figure 10) and right half of lower lip were remarkably enlarged and incompetent. On palpation swelling was nontender, hard in consistency, and noncompressible. Skin over the involved area was hyper pigmented. This hyper pigmentation started from pinna upto clavicle. Temporo mandibular joints movements were normal. Regional lymphnodes were not palpable. On intraoral examination, there was sufficient mouth opening. Enlarged right maxillary and mandibular alveolar arches, upper and lower labial mucosa, and buccal mucosa were observed.

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