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Mandibular Asymmetry Associated with Perimandibular Hemangiomas

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mation seems to take place mainly within the first 2 years. After this time there appears to be functional equilibrium between the condyle and the transplant. This functional stimulation protects the graft from beeing resorbed completely. Thereby, we were able to establish the remarkable fact that no changes in the form of condyle were observable in the X-rays. In conformity with orthopedic methods (*Bergmann* 1948), bulky bone transplants should complement and augment the rim of the glenoid fossa in its possible largest circumference. Contrary to this, parts of articular eminence or zygomatic arch are too tiny and can only be placed to the ventro-lateral site of the condyle. The position is unstable, the conditions for bony union are poor. They therefore do not fulfil the requirements of a broad arthroereisis. The same view applies to procedures where the transplants are wedged into a gap inbetween the zygomatic arch (*Aubry* and *Freidel* 1952).

Essential in our procedure is the nonviolation of the articular cavity. Undesirable haemarthros and damage of cartilaginous articular surfaces are thus avoided. We have kept to this procedure till today, because our earlier postoperative controls (*Selle* and *Haschemi-Nejad* 1967) had already revealed similar good results as the check-up described above. These findings confirm that the theoretical design of an arthroereisis of the temporo-mandibular joint was correct and that the method is reliable, provided the operative technique and the postoperative immobilisation of the mandible are adequate.

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Summary

Two cases of unilateral hypoplasia and dysplasia of the mandible with involvement of the maxilla existing simultaneously with perimandibular cavernous hemangiomas are described. In both cases the clinical picture resembled that of otomandibular dysostosis. In one case there existed simultaneously v. Recklinghausen's neurofibromatosis. In both cases severe venous bleeding which was difficult to control was encountered.

Key-Words: Mandibular asymmetry, mandibular dysplasia secondary to hemangioma.

Introduction

Most forms of mandibular asymmetry lend themselves to diagnosis with relative ease. Once the etiology of the pathologic picture is known, adequate surgical therapy without unpleasant surprises can be undertaken. However, there are always individual cases involving forms of dysplasia in the region of the maxillofacial skeleton in which the etiology remains unclear. In two such cases, with unilateral mandibular dysplasia with extension to a portion of the midfacial skeleton, we unexpectedly encountered cavernous perimandibular hemangiomas. These lesions gave rise to severe bleeding which was difficult to control and in our opinion are regarded as possibly etiologic in these unilateral maxillofacial anomalies. Since the literature to date apparently contains no information dealing with the etiologic role of vascular malformations in maxillofacial skeletal anomalies, this would seem to indicate that it is seldom observed. However, the undertaking of surgical correction in such cases can create a life threatening situation and our experiences with this entity should provide the surgeon with valuable advance knowledge.

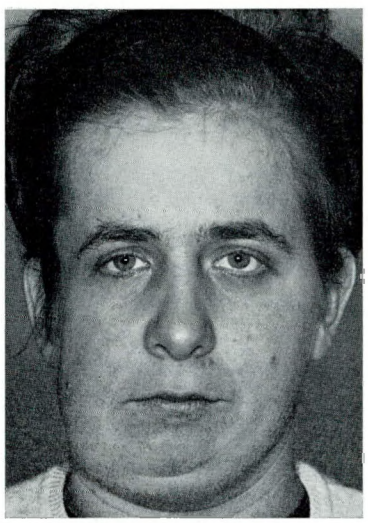


Fig. 1 Flattening of the area encompassing the right temporomandibular joint, zygomatic bone and temporal region. The pupillary plane is inclined downward toward the right and the caudal position of the right external ear is illustrated.

Patients

Case 1: A 26-year-old female was referred to us for correction of a unilateral otomandibular dysostosis. The history revealed an interruption in the development of the lower half of the face on the right beginning at the age of six years. One year prior to the present hospitalization the mandibular right first and second molar teeth had been extracted without incident regarding unusual blood loss. Trauma of the facial skeleton could not be elicited. There were no general or local complaints.

General Health: No significant pathologic findings.

Extraoral Examination: On the right side there was a flattening of the region encompassing the temporomandibular joint, zygomatic bone and temporal area. Soft tissues overlying the body of the mandible were slightly turgid and the chin was receded. The external ear and canal on the right were more caudally placed than on the left and the pupillary plane was inclined downward toward the right *(Fig. 1)*. There was a clearly palpable mandibular hypoplasia on the right. Motor and sensory nerve functions were normal bilaterally.

Oral Examination: The maxilla was edentulous and the mandible was partially edentulous, all teeth were vital. The midline of the mandible was shifted to the right which produced a gross occlusal discrepancy. Maximal

mouth opening with the maxillary denture in place was 40 mm and there was a concomitant deviation of the mandible to the right. There was absence of the protrusive movement on the right. In the right edentulous mandibular region there was an indolent, compressible thickening of the mucosa. Beginning in the area of the mandibular right cuspid, the body and ramus of the mandible were palpably smaller. The uvula was adherent to the glossopalatine arch on the right. No other pathologic findings were noted.

Radiographic Examination: Orthopantomogram *(Fig. 2a)*, posteroanterior mandible *(Fig. 2b)*, right and left lateral oblique of the mandible, temporomandibular joints (Parma and Schüller views), posteroanterior skull: There was a striking asymmetry of the mandible. The body and ramus of the mandible on the right, in both the vertical and sagittal planes, were more than half reduced in size. Cortical bone was missing. The inferior alveolar canal was clearly visualized and over part of its course formed the posterior border of the ramus. The angle of the mandible was missing. The semilunar notch was extensively deepened, the neck of the mandibular condyle was elongated and thinned, as well as the coronoid process. There was an asymmetry of the maxillary and zygomatic bone regions with distinct hypoplasia on the right side. The left side appeared normal.

Tomograms of the temporomandibular joints: There was extensive hypoplasia of the right condyloid and coronoid processes *(Fig. 2c)*. The articular fossa was irregularly defined and the joint space was widened.

Provisional diagnosis: Mandibular and maxillary hypoplasia of undetermined origin.



Fig. 2a The reduction in size of the body and ramus of the mandible on the right as well as the elongated and diminished condyloid and coronoid processes are readily evident in the orthopantomogram. The maxilla and zygomatic bone on the right are hypoplastic.



Fig. 2b The posteroanterior view of the mandible shows clearly the pronounced asymmetry and the decreased opacity of the right side of the mandible due to resorption of the cortical bone.



Fig. 2c The extensive hypoplasia of the right temporomandibular joint, neck of the condyle and coronoid process as well as deepened semilunar notch and poorly defined articular fossa and widened joint space, are all clearly visible in the tomogram of the joint.

Treatment Plan (Prof. *H. L. Obwegeser):* Unilateral or bilateral sagittal splitting of the ramus as described by *Obwegeser* (1957) for the correction of the asymmetric mandibular retrognathia. Relocation of the chin border prominence as described by *Obwegeser* (1958), simultaneous reconstruction of the defective ramus, angle and body of the mandible with free rib grafts. Later vestibuloplasty with skin graft.

Operation (PD Dr. *M. Perko*): Under nasal endotracheal general anesthesia a typical exposure for sagittal splitting of the right ramus was done. Immediately profuse venous bleeding was encountered. It could not be controlled with clamps. In order to get better access a submandibular approach was made. Here too, life threatening hemorrhage was encountered from fragile hemangiomatous tissue extending from the mandible to the mastoid and to the base of the skull. Ligation of the external carotid artery had no influence upon bleeding, likewise a trial compression of the

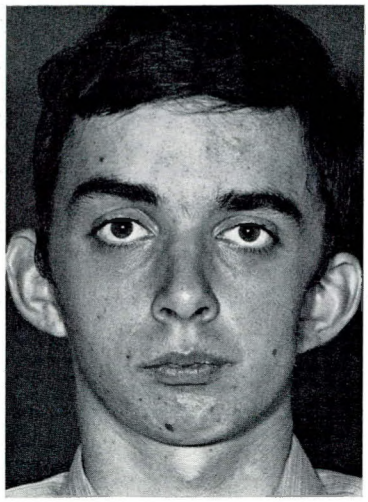


Fig. 3 Flattening of the lower left half of the face, the hypoplasia in the region of the temporomandibular joint and ramus of the mandible the downward inclination of the pupillary plane toward the left and the caudal position of the left external ear are evident.

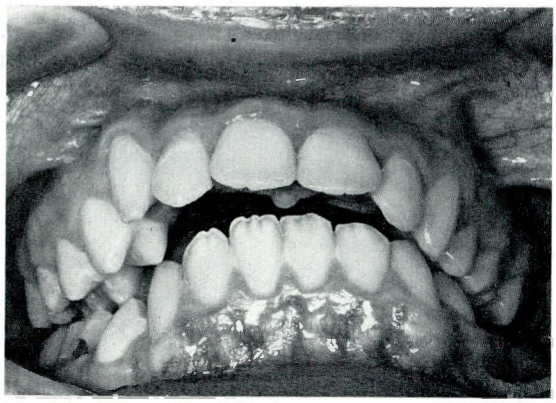


Fig. 4a Abnormal occlusion.

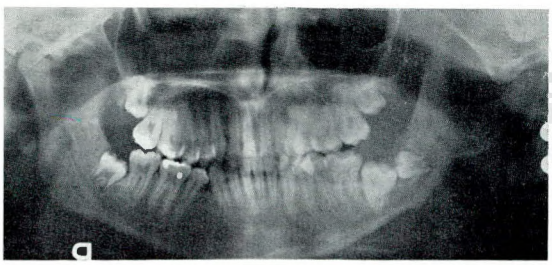


Fig. 4b The orthopantomogram shows diminished size of the ramus and posterior body region on the left, thornlike projections in the area of the missing angle, deepening of the semilunar notch with apparent elongation of the coronoid and condyloid process.

common carotid artery was without influence upon the bleeding. The ascending ramus, which was extensively erroded posteriorly, was resected in order to gain access for the ligation of some of the dilated, thin walled blood vessels. A definitive control of the hemorrhage could be achieved only with massive tamponment. Total blood loss was approximately 7200 cc. Blood replacement was with fresh whole blood, Ringers solution and PPL.

Postoperative course: On the seventh postoperative day under general anesthesia the packings were changed. On the 14th postoperative day a secondary closure of the wounds was done. As a consequence of the extraoral manipulations the patient sustained a total right facial paralysis.

Case 2: A 17-year-old male was referred with a diagnosis of a left otomandibular dysostosis. The lesion was first ascertained at the age of 5 years. Two months prior to the present hospitalization the patient had undergone removal of approximately 40–50 neurofibromas from the arms, legs and ventral trunk due to v. Recklinghausen's neurofibromatosis. Three months previously, the mandibular right third molar and left second and third molar teeth had been extracted without incident regarding blood loss.

General appearance: Numerous scars were present on the skin, secondary to excision of neurofibromas. No other significant pathologic findings were noted.

Extraoral examination: The left side of the face appeared flattened and in the region of the temporomandibular joint and ramus of the mandible there was an obvious hypoplasia. The mandible was micrognathic with deviation of the chin prominence to the left. The pupillary plane was inclined downward to the left and the external ear and canal on the left were more caudally placed than on the right *(Fig. 3)*. The distance from the deepest point on the infraorbital rim to the buccal cusp of the maxillary second premolar was 55 mm on the right, corresponding to a measurement of 50 mm on the left. The vertical distance from the infraorbital rim to the inferior border of the mandible on the right was 95 mm and on the left it was 87 mm. The temporomandibular joint on the left was tender to pressure and the mandibular angle could not be palpated.

Oral examination: The maximal spontaneous mouth opening was 28 mm. The mandible deviated 6 mm to the left during protrusion. The maxillary and mandibular dentitions were almost completely intact.

There was an overjet of 15 mm with no occlusion and a lingual inclination of the posterior maxillary and mandibular teeth on the right *(Fig. 4a)*.

Radiographic examination: Orthopantomogram *(Fig. 4b)* and posteroanterior view of the mandible *(Fig. 4c)*: There was an obvious mandibular asymmetry. The ramus and body of the mandible distal to the first molar tooth was approximately 1/3 smaller on the left than on the right. The mandibular angle on the left was non-existent and thornlike projections were seen in this



Fig. 4c The asymmetry of the mandible is apparent in the postero-anterior view.

region. In the remaining portions of the mandible smooth borders and normal bony structure were seen. The inferior alveolar canal was well visualized. The semilunar notch was deepened with elongation of the coronoid and condyloid processes. There was a hypoplasia in the region of the left zygomatic bone. Tomograms of the temporomandibular joints showed a pronounced hypoplasia of the left condyle and flattening of the glenoid fossa. The right side appeared normal.

Provisional diagnosis: Left mandibular and maxillary hypoplasia of undetermined origin.

Treatment plan:

1. Rotation of the caudal half of the left side of the face down and forward as advocated by *Obwegeser* (1970) in the management of otomandibular dysostosis.
2. Relocation of the chin border prominence.
3. Corrective operations as indicated.
4. Orthodontic treatment as indicated.

Operation (Prof. *Obwegeser*): Under nasal endotracheal general anesthesia a typical sagittal splitting operation was begun. The right side was completed uneventfully. On the left side, after the mucosal incision was made, and while elevating the periosteum laterally and posteriorly from the ramus, unusually copious venous hemorrhage was encountered. Attempts to achieve hemostasis by clamping, ligatures and electrocautery were unsuccessful. All attempts to achieve hemostasis from an intra-oral route were met with futility, therefore an extra-oral incision was made below the angle of the mandible. Here also copious bleeding occurred from hemangiomatous tissue which extended to the base of the skull. Extensive packing of the operative site from extra-oral was done, the intra-oral wound was closed and a pressure dressing was applied. Blood loss measured approximately 3000 cc. Blood replacement was with fresh whole blood and glucose solution.

Postoperative course: On the twelfth postoperative day under general anesthesia the packings were removed, whereupon fresh bleeding again occurred. The area was tamponated by packing. Following this the packing was removed in increments over the next 10 days without further bleeding. Carotid arteriogram on the left four months after the first operation disclosed a normal arterial distribution in the facial skeleton. No arterial aneurysms, arteriovenous fistulas or vascular malformations were noted.



Fig. 5a Postoperative occlusion achieved through sagittal splitting of the rami of the mandible.

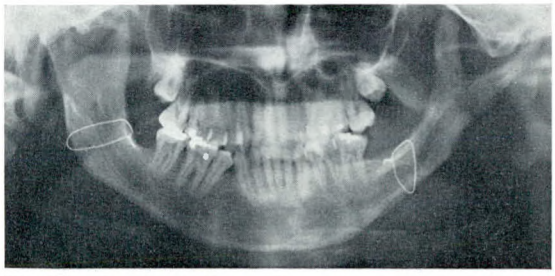


Fig. 5b Postoperative orthopantomogram.

Reoperation 4 months after the first operation: Upon exposure of the ramus of the mandible on the left side no unusual problems were encountered. Upon elevation of the periosteum on the lingual aspect of the ramus copious venous bleeding was again encountered. After hemostasis was achieved by tamponage packs, the sagittal splitting of the mandible on the left was completed as well as that on the right side. Thereby, correction of the mandibular asymmetry was achieved, as well as the correction of the micrognathia *(Fig. 5a, b)*. The rotation of the maxilla was deferred. Blood loss measured approximately 2300 cc. Blood replacement was with fresh whole blood, Ringer's and glucose solutions. On the twelfth postoperative day an attempt to remove the packing was again met with copious bleeding. A fresh packing was placed for additional 10 days. Further corrective operations to be performed at a later time.

Discussion

Mandibular hypoplasia can be congenital, post-traumatic, postoperative, neoplastic in origin, due to osteomyelitis, result from irradiation or from

a restriction of growth due to the formation of scar tissue. In the two cases described the precise etiology could not be proved. Operations in the regional jaw anatomy had not been performed. The removal of the third molar teeth (case 2) and the extractions (case 1) were operations carried out on the asymmetric mandible "per se". The hemangiomatous tissue was not disturbed in these procedures. A great similarity exists between the cases herein reported and otomandibular dysostosis. However, in these cases the facial asymmetry was not congenital in origin. Also the characteristic mal-development of the ear and commissural macrognathia were missing. Other factors clearly distinguishing these cases from classical otomandibular dysostosis are the deepening of the semilunar notch with resultant elongation of both the coronoid and condyloid processes. Otherwise, the morphologic appearance of both syndromes is almost identical. Congenital and progressive forms of hemifacial atrophy are readily excluded in the differential diagnosis since all structures in the involved hemiface are affected. Also a unilateral form of dysostosis mandibulofacialis *Franceschetti* is not considered on the basis of its typical symptom complex. The perimandibular cavernous hemangiomas discovered at operation may be considered as primarily etiologic in the reported cases. The concomitant hypoplasia of the maxilla and zygomatic bone may be attributable to an extension of the hemangioma into this region. Additionally, it is considered possible that the inability to protrude the mandible is a result of extension of the lesion into the interarticular space and that this phenomenon also accounts for the widened joint space, the hypoplasia of the condyle and the thinning of the subcondylar neck.

In contrast to other skeletal areas, hemangiomas of the jaw regions are seldom encountered (*Lucas* 1972). In the majority of reported cases the hemangiomas have been central in the mandible *with* (*Erich* 1947, *Pizer* 1954, *Pusey* 1967) *or without significant* mandibular asymmetry (*Williams* and *Gilbert* 1955, *Villa* and *Limchayseng* 1955, *Battersby* 1957, *Kaburda* and *Taylor* 1967). A number of case reports involving a unilateral facial asymmetry resulting from the bulging produced by enosseous hemangiomas have been published (*Weinstein* et al. 1963, *Marie* and *Barety* 1953). Instances of perimandibular hemangiomas have not been found in the literature by the author.

With regard to site of origin *Thoma* and *Goldman* (1960) and *Lucas* (1972) name basically two variations of hemangiomas of bone, one of which arises within the bone as an anomaly of the blood vessels of the bone marrow and the other arising in the periosteum and resorbing the underlying bone. The last mentioned type of hemangioma is probably the one which was found in the described cases.

In the second case of this report there existed *von Recklinghausen's* neurofibromatosis. *Landing* and *Farber* (1956) state that hemangiomas frequently occur in the presence of neurofibromatosis. *Rittersma* et al. (1972) describe two cases of neurofibromatosis with deformity of the mandible, the clinical and radiographic findings being very similar to those observed in the patients discussed in this report; however, operation reports are not included. One must conclude that there is imminent possibility that in those cases too perimandibular hemangiomas could have led to the development of mandibular asymmetry.

Conclusions

1. Perimandibular hemangiomas can be responsible for hypoplasia and dysplasia of the mandible and maxilla and consequently lead to facial asymmetry which may appear similar to otomandibular dysostosis.
2. Individual, isolatable blood vessels supplying the cavernous hemangiomas cannot be identified and the surgical bleeding which occurs can be controlled only by massive tamponading for approximately 14–20 days.
3. To assist in the diagnosis of unclear cases, exploratory aspiration with a thin needle is suggested.
4. Previous experience of uncomplicated tooth extractions provides no clue as to the existence of a perimandibular hemangioma.
5. In certain cases, as in case two of this report, correction of the mandibular anomaly can eventually be completed.
6. Because of the danger of profuse operative hemorrhage, inexperienced operators should not undertake surgical correction of jaw anomalies in cases where the diagnosis is unclear.

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