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Duplication of Structures around the Stomatodeum

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Summary

In this case report a patient with multiple duplications in the cervival spine, skull-base and oral cavity is described and a discussion on the possible origin of the duplications is presented.

Key-Words

Polygnathia – Double mouth – Duplication of mandible – Duplication of pituitary gland – Duplication of tongue – Duplication of cervical vertebrae – Duplicitas anterior – Occipital vertebrae – Split notochord syndrome

Introduction

Cases of facial duplication are rare and mostly unique. In the following an other one will be described, a possible explanation for its developement will be given and it will be compared to other cases in the literature.

Case History

A female infant, born prematurely in August 1964 was admitted to the clinic at the age of 3. The family history was negative and the first months of pregnancy had been trouble-free.
Extra orally, hypertelorism and macrostomia and accompanying duplication of the lower lip were observed (Fig. 1).
Intra orally a cleft palate (Fig. 2) and an anterior open bite were present, but the most striking features were the presence of two mandibular arches and the duplicated anterior part of the tongue both parts of which were lying inside their respective mandibular arches. The two mandibular arches had an osseous connection in the midline and the following deciduous teeth were clinically present: 75, 74, 73, 72, 71, 81, 83, 85, 75, 74, 73, geminated tooth, 83, 84, 85, (Fig. 3). Radiologically some permanent tooth-germs were visible.

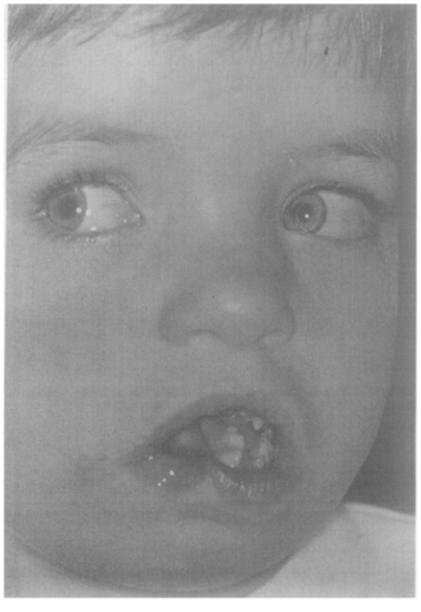

Fig. 1 Patient 3 years of age.

Enquiries at the hospital where the patient was born revealed that at birth a 3–5 cm tumour, attached to the vomer and palate and protruding outside the mouth, was removed directly after delivery. Pathological examination of the tumour revealed that the resected tumour was covered with a thin epidermis, containing a lot of delicate hair, under which fatty tissue with fibrous sheets and some striated muscle tissue were present. The base of the tumour revealed some "respiratory mucosa" with, underneath, some bony tissue and two tooth-germs. The conclusion was therefore that we were dealing with a hamartoma.
Because of abnormal radiological configuration in the region of the sella turcica and the cervical spine, it was decided to make computerized tomograms. They revealed paired sphenoid sinuses (not an abnormal finding) (Fig. 4) and duplication of the sella turcica (Fig. 5).
A broad base of the skull besides duplication of the ventral aspect of the foramen magnum with a tendency of the occipital bone to form an occipital vertebra, mainly on its ventral aspect was present. Vertebrae C_1, C_2 and C_3 were flat and broad ventrally and C_2 showed two odontoid processes (Fig. 6).
The cerebrum and the ventricles did not shown any tendency to duplication. Radiographs of the cervical spine revealed that vertebrae C_2 and C_3 were fused (Fig. 7).

Surgery

Apart from the excision of the tumour performed elsewhere as mentioned previously, at our clinic, in the year of admis-

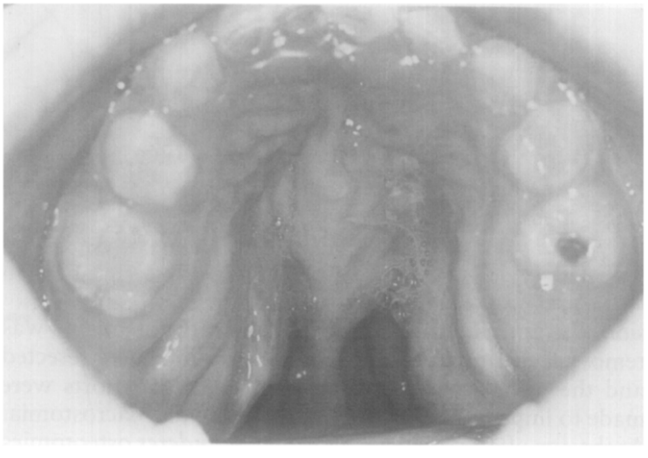

Fig. 2 Patients cleft palate.

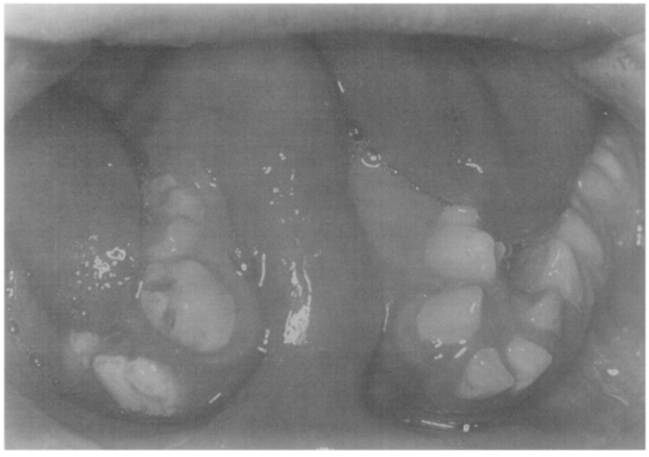

Fig. 3 W-shaped mandible double and anterior part of the tongue.

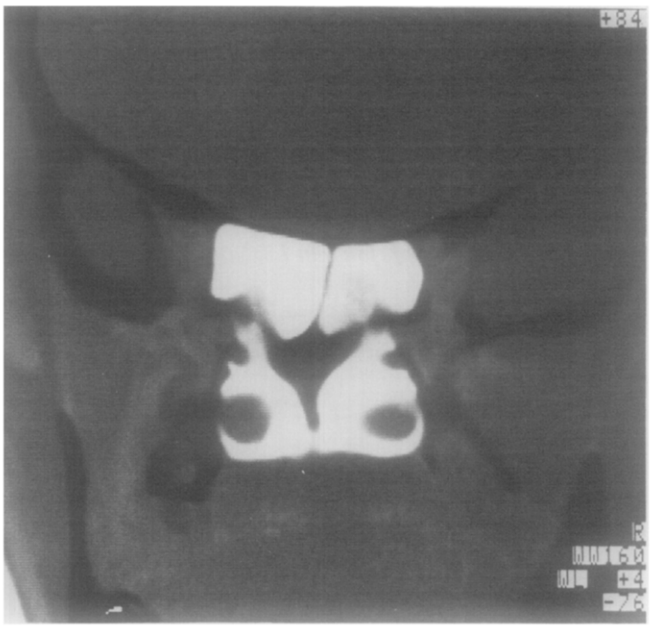

Fig. 4 Paired sphenoid sinuses.

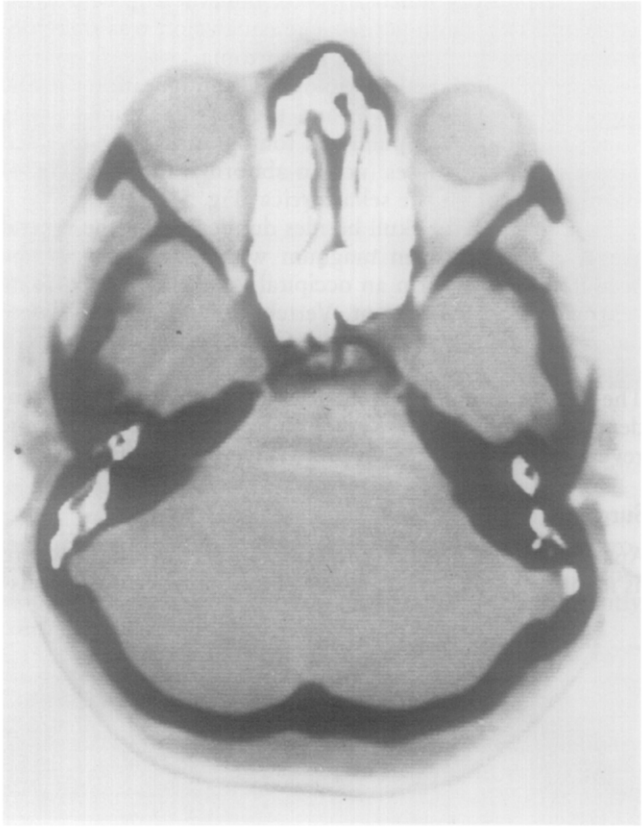

Fig. 5 Duplication of sella turcica.

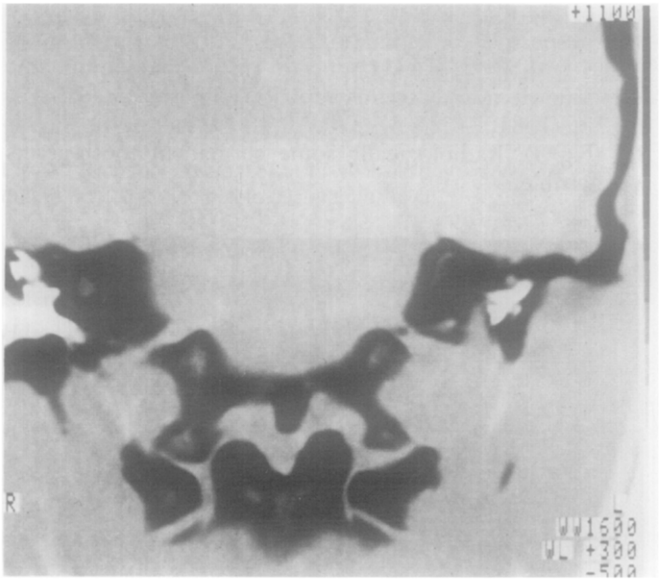

Fig. 6 Broad cranial base, duplication of the ventral aspect of the foramen magnum tendency of the occipitial bone to form an occipital vertebra. Vertebra C_2 showing two odontoid processes.

sion, the anterior part of the tongue on the right side was removed, the medial bodies of the mandible were resected and the palate was closed. At the age of 10 efforts were made to improve lip closure by correcting the macrostomia. At the age of 16 combined *Köle* and *Wunderer* osteotomies were performed to close the anterior open bite. Postoperative orthodontic treatment is still continuing (Fig. 9). At the age of 18 a pharyngoplasty was performed to correct nasal escape during speech. The mental development of the patient was just below average but at least in part, this might well be related to speech and hearing problems caused by the cleft palate sequellae.

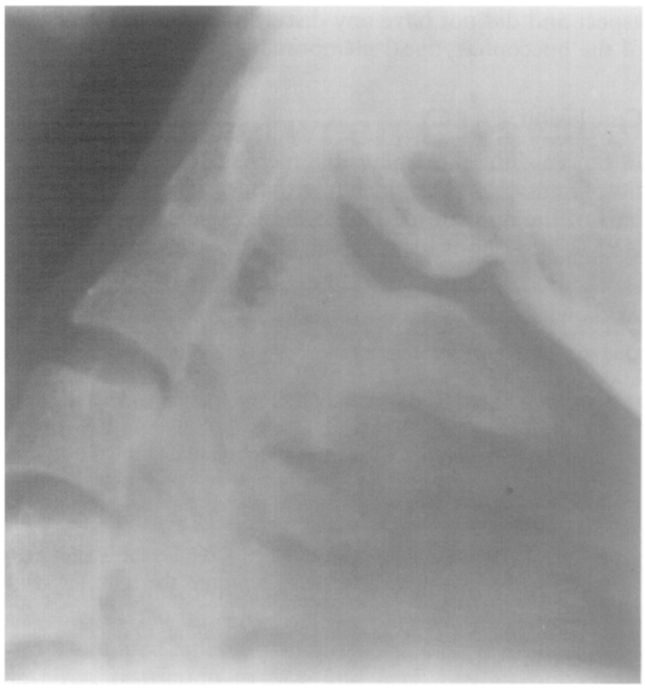

Fig. 7 Vertebrae C_2 en C_2 are fused.

Review of the Literature

Almost every case of facial duplication reported in the literature is in one way or another unique and therefore not quite comparable with other ones described.
Duplication of the lower lip and mandible was described by *Meijer* (1883), *Price* and *Zarem* (1979) and recently by *Maisels* (1981). A case mentioned by *Kawamoto* et al. (1977) seems to us to be the same case as that described later by *Price* and *Zarem* (1979).
A case with oral manifestations nearly identical with our case was described by *Morton* (1957). Here, also two pituitary glands were present.
Other cases of duplications in which a second more or less rudimentary oral cavity was present, were described by *Israel* (1877), by *Quinard*, quoted by *Herbst* and *Apffelstaedt* (1928), *Bjerrum* (1930), *McLaughlin* (1948), *Beatty* (1956), *Goulian* and *Conway* (1964), *Bacsich* et al. (1964) (here two pituitary glands and partial duplication of the cervical vertebrae were found), *Davies* et al. (1973), *Kawamoto* et al. (1977) and *Borçbakan* (1978).

Discussion

In the cases presented the degree of organization of the tissues and organs makes the possibility of a teratoma unlikely.

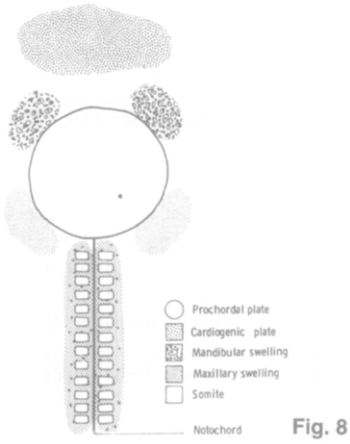

Fig. 8a

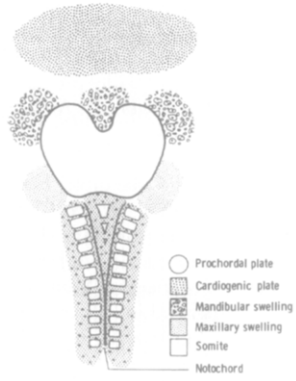

Fig. 8c

Fig. 8a Schematic drawing of normal embryological structures.

Fig. 8b Normal embryological development.

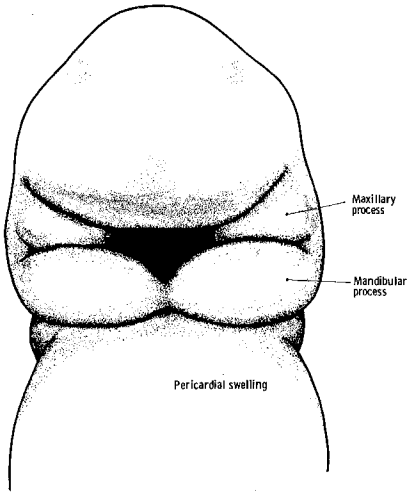

Fig. 8b

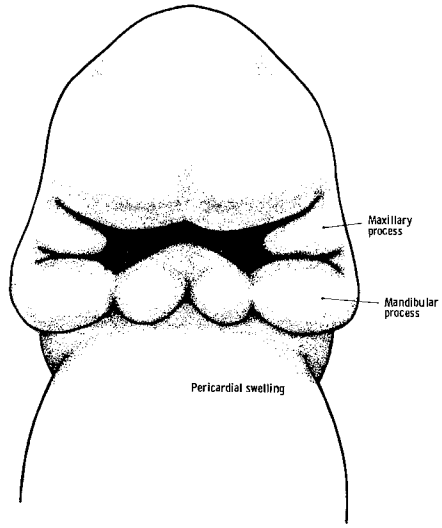

Fig. 8d

Fig. 8c Hypothetical configuration of embryological structures; splitting of the notochord with its disturbing effect an the anteroventral part of the prochordal plate.

Fig. 8d Hypothetical embryological development.

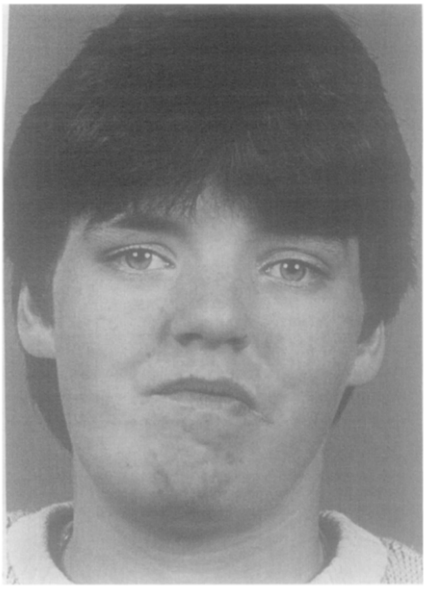

Fig. 9 Actual condition after surgery at the age of 16 years.

An epignathus is less probable because of the presence of oral tissues only. *Magitot* (1875) considers the possibility of splitting of one of the branchial arches whereas *Meijer* (1883) thought a supernumerary branchial arch to be the causative factor. *Meijer* (1873) was also the first to suggest the possibility of splitting of the foetus in the head region. *Morton* (1957) supposed that these abnormalities were part of the split notochord syndrome, the most extreme manifestation of which is a complete duplication of the head.

In tracing this extreme abnormality back to its embryological development we arrived at the following hypothesis. If a duplicated anterior notochord had been present, it may have caused a disturbance of the antero-ventral part of the prochordal plate, i. e. the future buccopharyngeal membrane. A more or lesse heartshaped prochordal plate as shown in Fig. 8 c might have been the result.

Moreover, the two notochords in the anterior region may have induced the development of two Rathkes pouches, giving rise to two pituitary glands and the development of two sellae turcicae. Other phenomena suggesting a double anterior notochord are the osseous duplication of the cranial base and cervical vertebrae.

The prochordal plate or buccopharyngeal membrane becomes surrounded by mesenchyme of the first branchial arch. Later, from these mesenchymal condensations two maxillary processes and four mandibular processes may have developed in accordance with the typical shape of the buccopharyngeal membrane (Fig. 8 d).

The ventro-medial aspects of the two sets of mandibular processes may have given rise to the two anterior parts of the developing tongue. Broadening of the structures which form the base of the skull and are responsible for positioning of the maxillary process may result in the hypertelorism observed and in the failure of the secondary palate shelves to fuse.

Our case can only indirectly be compared with the case of Antonio, (*Obwegeser* et al., 1978). There, a split notochord may as well have been the causative factor although its effects on the surrounding tissues were limited to the dorsal aspect and did not have any disturbing effect on the shape of the buccopharyngeal membrane.

Conclusion

In our case the most plausible mechanism for the duplication seems to be a splitting or duplication of the anterior part of the notochord. This is in agreement with *Morton*'s (1957) idea about his case.

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