Oculo-auriculo-vertebral (Goldenhar's) syndrome

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Goldenhar's syndrome (Goldenhar, 1952) consists of epibulbar dermoids or lipodermoids, auricular appendages, pretragal blind fistulas, and vertebral anomalies. A number of other deformities associated with this condition have been summarized by Bowen, Collum, and Rees (1971).

Our case is presented because of the presence of an aberrant salivary gland in the nose. This complication has not been previously reported in Goldenhar's syndrome.

Case report

This baby boy, born at the University of Missouri School of Medicine, was seen in the Ophthalmology and Plastic Surgery Clinics at regular intervals. There was no family history of congenital abnormality.

Examination revealed the following congenital anomalies:

- (1) Dermoid, left eye (Fig. 1).
- (2) White opacity behind pupil, left eye.
- (3) Preauricular appendages, both sides (Fig. 2).
- (4) Deviated nasal septum with mass in nose (Fig. 2).
- (5) Blind fistula near left ala nasi (Fig. 3).
- (6) Right pes equinovarus.

FIG. 1 *Epibulbar dermoid on nasal aspect of left eye*

FIG. 2 *Epibulbar dermoid, left eye. Bilateral preauricular appendages. Nasal deformity*

FIG. 3 *Profile showing large preauricular appendage and blind fistula (arrow)*

Radiological examination

The left orbit was smaller than the right in vertical diameter (Fig. 4). The right optic foramen measured 5 mm. and the left 3 mm. The nasal septum was deviated to the left. The left side of the mandible was hypoplastic and the left zygomatic arch was partially absent (Fig. 5).

FIG. 4 Antero-posterior view of skull, showing calcification of falx. Left orbit smaller in vertical diameter than right orbit

FIG. 5 Water's view of skull, showing deviation of nasal septum to the left side. Left side of mandible and left zygomatic arch are hypoplastic

Treatment

Under general anaesthesia the dermoid of the left eye and the preauricular appendages were removed. The mass in the nose was excised and the fistula repaired. When the baby was 3 years old the left eye was enucleated; it appeared considerably smaller than the right eye and a whitish mass was seen behind an irregular pupil. The pes equinovarus was corrected in the Orthopaedic Clinic.

Pathological findings

The left eye was phthisical. The mass in the nose consisted of glandular tissue composed of mucous and serous acini (Fig. 6). Fat cells were present. The pathological diagnosis was that of an aberrant salivary gland.

Discussion

Doyne (1903) reported a case of Goldenhar's syndrome with underdeveloped right side of the nose and small right nostril. Gupta, Gupta, and Prashar (1968) described a nasal deformity due to malunion of the medial and lateral nasal folds in the same syndrome.

The salivary glands develop as ectodermal epithelial proliferations from the oral cavity, the first being the submandibular gland in the sixth embryonic week (Ferner, 1949). The hard and the soft palate start to form in the seventh embryonic week (Remnick, 1970).

FIG. 6 Portion of aberrant gland with serous and mucous acini. Several fat cells are seen. $\times 100$

Because of the relatively high frequency of occurrence of a cleft palate in this syndrome (7 per cent. according to Gorlin and Sedano, 1970), a delay in the closure of the palate may have taken place in our patient, permitting upward growth of the aberrant salivary gland from the oral cavity.

Summary

A case of Goldenhar's syndrome associated with an aberrant salivary gland in the nose is reported. The orbit and the optic foramen were smaller on the left side than on the right. The left eye was enucleated at the age of 3 years; it was phthisical.

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