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Proboscis lateralis, a rare malformation

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INTRODUCTION

Selenkoff, in 1884, was the first to describe this congenital abnormality. Since then a number of cases have been published, all with the same morphological and histological findings. Clinically, proboscis usually appears as a trunk-like appendage coming from the upper and inner wall of the orbital roof and replacing the external nose. It is club-shaped with a small opening at its distal end, which is the outlet of the canal that runs through the whole trunk. The proximal end of the canal is blind and has no connection with the nasal cavity. In addition to proboscis, various deformities may be present probably depending on the time during foetal development at which some form of damage occurred. Examples of such deformities are absence of the frontal sinus, antrum Highmori, vomer and premaxilla (Selenkoff, 1884), absence of the olfactorius and cribriform plate on the affected side (Seefelder, 1910), absence of the isolateral wall and a lower lid cleft at its inner angle (Landow, 1890; Skevas, 1988), a high arched palate, clefts in the upper lid, iris and retina (Tendlau, 1918; Biber, 1949; Rondal and Duritz, 1977), a cleft between the nasal and maxillary bones, occasionally lack of the lower lacrimal pundum (Kirkpatrick, 1970), lack of the olfactory or optic nerves, hypertelorism, medial canthal cleft and nasal chamber absence (Kirkpatrick, 1970; Rondal and Duritz, 1977).

The incidence of proboscis lateralis is less than one in 100,000 infants, born alive, and unrelated to the nationality or sex. Bilateral proboscis has been reported once in the literature (English, 1984). On the other hand, proboscis can be present with an otherwise normally developed nose. Embryologically the external nose is formed by the union of the medial and lateral nasal processes. The medial nasal process gives rise to one half of the cartilaginous septum and the medial crus of the lower lateral cartilage, while the lateral nasal process produces the nasal bone, the lateral crus of the lower lateral cartilage, the upper lateral cartilage and the anterior end of the lower turbinal complex. The lateral nasal process is the one which supplies the material for the proboscis lateralis. The onset of this proboscis is believed to be between the fourth and fifth week of foetal life.

Stupka (1938) suggested that most likely an exogenous cause, probably a traumatic injury (amniotic cords) was responsible for this malformation. He suggested that the time of onset is not earlier than the period corresponding to the appearance of the lacrymal-nasal cleft and the approximation of the nasal and maxillary processes, but not later than the time at which fusion of the physiological optic nerve occurs. Biber (1949), considering all cases described, agreed with Stupka that it was the lateral nasal process which supplied the material for the formation of the proboscis. Peter (1910) and Tendlau (1918) both observed cases in which the proboscis was dislocated to the upper and outer margin of the orbit. Peter believed that the new bone production and consequent dislocation of the orbit might be regarded as a healed encephalocele, while Tendlau believed that a germinal anomaly was the causative factor of the malformation. Blair (1931) and Bishop (1964) stated that lacrimal duct overgrowth gave rise to this anomaly, while Binns (1969) suggested that hypertrophy of the lateral nasal process was responsible. Rondal and Duritz (1977) assumed that the lesion responsible for the anomaly must be related to the primary organizer of the nasal portion of the face. This was either the nasal placode or the forebrain that induced the nasal placode.

CASE REPORT

A female child was brought into our clinic a few days after delivery. She was the first child of a 20 year old woman, who had had no problems during her pregnancy except for a common cold in the 24th week. No drugs were taken during pregnancy except iron tablets and delivery was normal.

The complete E.N.T. examination revealed no malformation other than a left-sided proboscis lateralis 1.5 cm in length (Figure 1). Both nostrils were present and the X-rays of the nasal sinuses were normal. The proboscis was suspended from the area of skin covering the left site of the root of the nose, which is unusual since the most typical site is the upper and inner wall of the orbital roof. There was no connection between the proboscis canal and the inner nose. At the age of eight days and at the urgent request of her parents, the baby was taken to the operating room and the proboscis was removed under general anaesthesia. The small skin incision was sutured with Nylon 6/0 and no complications were observed after the operation. Histological study showed the proboscis lateralis measured 1.4 x 0.8 x 0.7 cm. The wall of the duct was lined partly with keratinizing stratified squamous epithelium and partly with pseudostratified columnar epithelium. Some of the epithelial cells showed cilia extending into the lumen. Varying numbers of goblet cells were interspersed. The epithelium rested on a prominent continuous thick easinophilic basement membrane. The underlying lamina propia contained a variable number of mucus and serous glands, blood vessels, adipose tissue, islands of cartilage and many striated muscle fibers

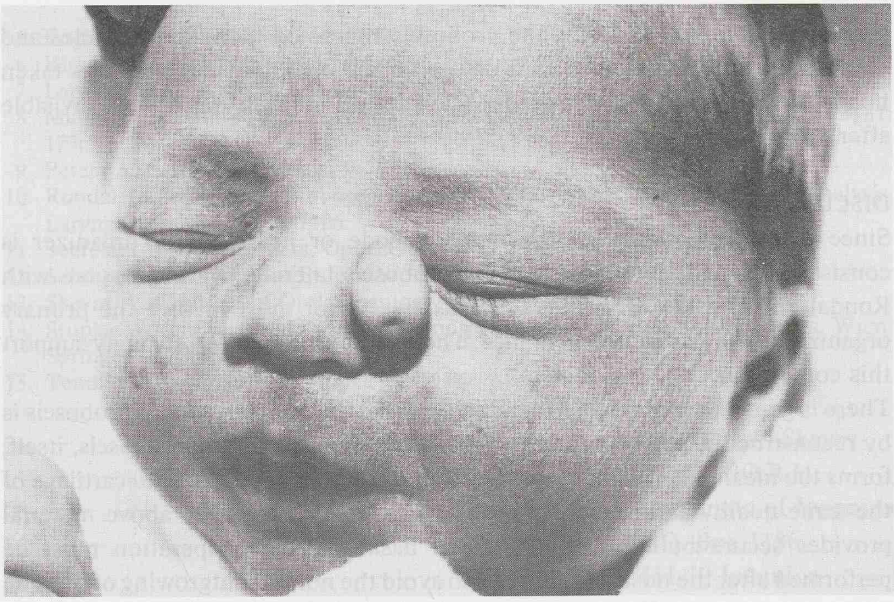

Figure 1. An eight days old baby with left-sided proboscis lateralis.

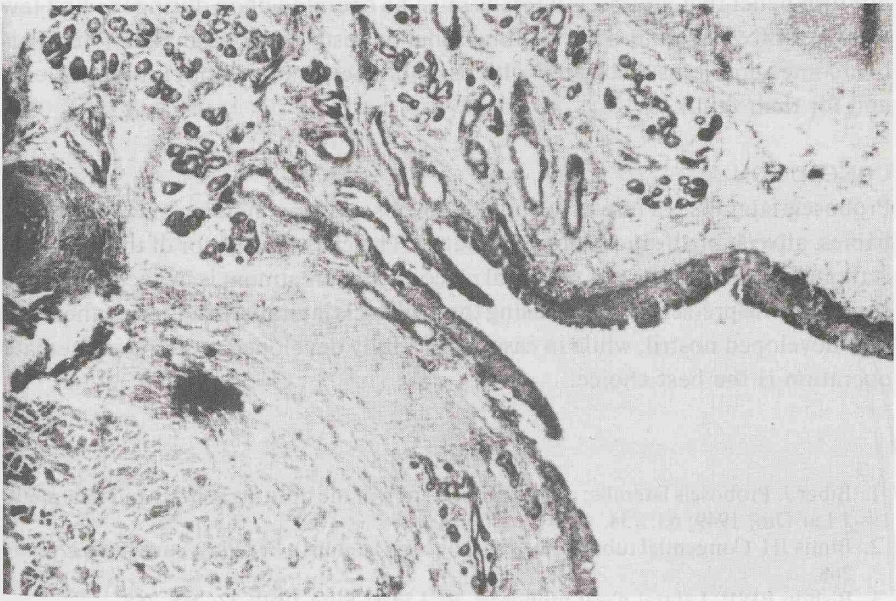

Figure 2. The lamina propria containing mucus and serous glands, vessels, adipose tissue, islands of cartilage and striated muscle fibres.

(Figure 2). The skin covering the proboscis contained many hair follicles and sebaceous and sweat glants. Four days after the operation the baby was taken home and has remained well since. The surgical incision was almost invisible after a few months.

DISCUSSION

Since a localized lesion of the nasal placode or its forebrain organizer is consistent with all autopsied cases of proboscis lateralis, we must agree with Rondal and Duritz (1977) that the causative factor must involve the primary organizer of the nasal portion of face. The histological findings strongly support this contention.

There is general agreement in the literature that the management of proboscis is by reconstructive surgery. In cases with a missing nostril, the proboscis, itself, forms the ideal material for reconstruction since it contains skin and cartilage of the same quality as that on the healthy side of the nose. The above material provides secure building of the lateral nasal wall. The operation must be performed after the nose has matured to avoid the normal outgrowing of the side affected. The best time is the pre-school age so as to reduce psychological trauma. (Blair, 1939; Biber, 1949; Bishop, 1964; Mahindra et al., 1973; Rondal and Duritz, 1977).

On the other hand, in cases similar to our case, with a well developed nose, it is our firm belief that proboscis lateralis must be operated upon during the first few weeks of life, since there are no functional or aesthetic contraindications. This overcomes problems associated with psychological trauma both for the patients and for their families.

CONCLUSION

Proboscis lateralis is a rare deformity of the nose with an incidence of 1 in 100,000 babies, alive at birth, and is not sex-related. The causative factor of this anomaly is thought to be an injury of the nasal placode. The treatment is by reconstructive surgery in the pre-school years, using the proboscis lateralis, itself, when there is a non-developed nostril, while in cases with a fully developed nose the immediate operation is the best choice.

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