MIDLINE CONGENITAL CERVICAL FISTULA OF TRACHEAL ORIGIN

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Up to about 1880 it was generally believed that midline congenital fistula of the neck might be due to tracheal maldevelopment, but according to the well established doctrine of today, they are supposed never to be of tracheal origin. If one studies the subject in any of the larger bibliographic reference works, he is impressed by the comparatively large number of reported cases of so-called congenital tracheal fistula. On consulting the articles referred to, however, one becomes aware that the error of referring these fistulas to tracheal maldevelopment persists only by virtue of the titles of the various articles. As a matter of fact, within three years after Dzondi (1829) first created the term tracheal fistula, Ascherson recognized the error; but in correcting it, fell into a somewhat similar one by assuming that all midline cervical fistulas communicated with the pharynx. In 1864, Heusinger coined the phrase branchial fistulas to explain these defects. Heusinger states: "Some authors classify congenital fistulas of the neck into (*a*) pharyngeal fistulas and (*b*) tracheal fistulas." He then adds that he will preserve this classification, although "a congenital tracheal fistula has never been demonstrated." He might have stated his point even more emphatically; for an analysis of all the case reports up to the time of Heusinger's publication discloses neither clinical nor pathologic evidence that would warrant more than a faint suspicion of tracheal participation in midline fistula formation. In 1848, Luschka published an article that evidently carried much weight as authority, for his views were accepted by Koenig as late as 1875 in that year's edition of his "Specielle Chirurgie," and by Fisher in the 1880 volume of "Deutsche Chirurgie," devoted to diseases of the neck. Luschka assumed that in midline cervical fistulas there exists, originally, a communication between the fistulous tract and the trachea.

An article published by Kostanecki and Mielecki in 1890 marked the establishment of the fundamental pathologic principles underlying cervical fistulas. These authors showed that the so-called tracheal midline fistulas had, in reality, nothing to do with the trachea. In their opinion, no significance was to be attached to the site of cervical fistulas; all cervical fistulas were branchiogenetic, and whether the openings were centrally or laterally placed depended merely on which

branchial cleft was responsible for the fistula. In their opinion, when the second branchial cleft was involved, a midline fistula resulted, as a rule; but when the third or fourth cleft was involved, a lateral fistula resulted. This, of course, was an error. It persisted, however, for only a short time.

The year after the publication of Kostanecki and Mielecki's article in 1891, His published his study of the thyroglossal duct, demonstrating its descent from the foramen cecum of the tongue to the permanent site of the thyroid isthmus, causing abnormalities in this descent that are responsible for all midline cervical fistulas. The views expounded by His have met with universal agreement. Koenig, himself, in a

Fig. 1.—Anterior view of the defect. Inset shows enlargement of the teat, rosette, cicatrix and fistulous opening.

later edition of "*Specielle Chirurgie*" (1895) says that "midline tracheal fistulas of congenital origin are merely figments of the imagination" (*phantasiegebilde*). And thus the question has stood since 1891: Midline congenital fistulas of the neck are related in some way to the maldevelopment of the thyroglossal duct (barring, of course, the cases in which branchiogenetic retentions have burrowed over toward the midline), and are never due to maldevelopment of the trachea.

A case encountered recently in private practice seems to establish quite clearly the fact that midline congenital fistulas of the neck may be due to tracheal anomalies, and that we cannot unreservedly group all these fistulas under the head of thyroglossal duct anomalies.

REPORT OF CASE

History.—A man, aged 26½ years, with a totally negative family and past history, reported that since the day of his birth he had had a defect in the middle of his neck. There had never been any associated pain or discomfort, and professional advice had never been sought regarding a cure. There had always been a small amount (a drop a day) of purulent discharge from the lower part of the defect, and a minimal amount of glairy secretion from the upper part ("not enough to bother about wiping off"). There had never been any retention with consequent signs of inflammation. The patient sought relief merely because the defect had become unsightly to him.

Physical Examination.—This was negative except as regards the defect itself. There were no other visible congenital defects.

Fig. 2.—Lateral view of the defect. Inset shows the details from this view.

From 1 inch below the chin, in the midline of the neck, running down to 1 inch above the jugulum, there was a prominent fold of skin, which felt like a subcutaneous cicatrix and which became tense when the chin was elevated. About 1½ inches above the jugulum in the midline, there was a firm fleshy teatlike process, three-fourths inch long and one-fourth inch in diameter. This process hung, tip downward, and was attached to the skin of the midline by a fold of skin, running from base to apex of the teatlike process. This fold which was creased on itself at rest, could be spread out fanwise by picking up the teat and pulling it away from the neck.

Directly beneath the low point of the fold of skin, there was a small circular rosette-shaped bud of granulation-like tissue, about one-fourth inch in diameter, presenting radiating sulci and rugae, and a surface that had a glazed appearance, as if painted with mucus. This glairy surface could be wiped dry and dull, but shortly afterward it became moist and glazed again. With

the finest silkworm gut probe, it was not possible to discover a fistulous opening in this rosette.

From this rosette, running downward toward the jugulum was an elliptical cicatrix, 1 inch long and one-half inch wide. The surface of this cicatrix was thoroughly epithelialized, dry and glazed. This cicatrix at its lower pole, ended in a fistulous opening one-fourth inch in diameter, which in turn led into a sinus that ran down to a point slightly below the upper margin of the sternum. This sinus was immediately under the skin, throughout its course, and could be partly everted through its mouth, showing a moist, rugose and bright red mucous membrane lining (Figs. 1 and 2).

Operation and Result.—The opening of the fistula was circumscribed and the tract was dissected out with a minimal amount of the surrounding tissue in which it lay. During this dissection, it was noted that in the wall of the

Fig. 3.—Photomicrograph of the teat.

tract, at about its center, there was a small pea sized cartilaginous-like mass. After the tract had been dissected out, two elliptical incisions were made, circumscribing the entire defect, running from just above the teat, along the lateral margins of the cicatrix already described and ending in the incision utilized for dissecting out the lower tract. Through these incisions, the defect was removed *en masse* and all removed tissue was dropped into 10 per cent. liquor formaldehydi. At no point could there be made out anything resembling a patent thyroglossal duct, or any other type of fistulous deep running tract.

The wound was closed around a small rubber tissue drain, and was perfectly healed in two weeks. It has remained healed now for more than ten months.

Histologic Examination.—The tissue removed was examined by Dr. M. T. Burrows, whose report is given herewith:

The specimen examined lay in the midline of the neck and consisted essentially of three parts. The upper end was located at a point just below the

level of the lower edge of the thyroid cartilage and the lower end was well down beneath the skin over the upper end of the sternum. The upper part consists of a teatlike protuberance which is covered with skin and looks like the lobule of the ear. This projects out and hangs over a small sessile tumor mass which has a glairy and fissured surface. The surface of this tumor mass is covered with a glairy secretion. The third part is a tube closed below and opening above, just below the middle part into a floor of what appears to be very thin skin. This tube admits a 3 mm. probe and extends down into the subcutaneous tissue over the sternum for about 3 cm. There

Fig. 4.—Low power magnification of section through rosette, showing cartilage, muscle layer, submucosa and papillary mucosa.

is a small mass of cartilage attached to the wall of this tube in its lower half. There is no definite connection between these structures and the deeper tissue of the neck. The specimen is fixed in liquor formaldehydi and sections are taken from three parts.

Section 1 is of the upper teatlike process. It is composed of a central mass of connective tissue. This is divided into two parts. An outer part which is covered by the epithelium of the skin. This part looks like true skin. It contains hair follicles and sebaceous glands, and transmits the sweat ducts from the glands below to the surface. The sweat glands appear normal. The internal layer consists of connective tissue in the center of which are a few

striated muscle fibers, a part apparently of the superficial muscle layer of the neck (Fig. 3).

Section 2 is from the sessile tumor which lies just below the teatlike process (Section 1). The outer edge of this mass is irregular. It has many papillomatous projections. Its surface is covered by low cuboidal and high columnar and ciliated epithelium (Figs. 4, 5 and 6). In the tissue between the papillae there is a mass of hyaline cartilage. The connective tissue between this cartilage and the surface is poorly divided into mucosa and submucosa. It is areolar in character. There are a few smooth muscle cells about the cartilage, and it contains well formed mucous glands and ducts. The ducts are lined with high columnar epithelium without, and low cuboidal cells within. The glands are like those of the normal trachea and bronchus. In a few glands, on the surface, the epithelium is pseudostratified, like that of the trachea. The whole picture is like that of sections of the trachea or bronchus.

Fig. 5.—High power magnification of portion of section shown in Figure 4, demonstrating the structure in greater detail, particularly the mucous glands.

Section 3 is a section through the lower part of the wall of the tube. It is covered with squamous epithelium like that of the larynx. It differs from this only that in a few places it has a more definite horny layer. The other layers peculiar to the skin are not present however. Ducts extend down from this into the deeper tissue. They are lined with a stratified epithelium of which the inner row of cells are high, columnar and ciliated. Their cytoplasm is clear. They are apparently filled with mucus. No glands are seen in this section. At one end of the section, there are a few sebaceous glands near what appear to be hair follicles. The connective tissue is areolar in type. The part lying just beneath the outer epithelial covering is densely infiltrated with lymphoid and polyblastic cells (Fig. 7).

Section 4 is taken from the wall of the lower end of the tube. It is covered with squamous epithelium like that of the larynx. There are a few lymphoid

cells in the connective tissue beneath, and deep in, there is an irregular mass of hyaline cartilage surrounded by a well formed perichondrial layer and skirted on one side by a mass of mucous glands like those in the larynx or trachea.

Section 5 is taken from just without the mouth of the tube. It is covered by squamous epithelium like that of the mouth. Deep in, there is a gland duct lined by pseudostratified and stratified epithelium of which the inner

Fig. 6.—High power magnification of mucosa (Fig. 5) showing ciliated epithelium.

layer is composed of clear high columnar cells. The connective tissue is dense and fibrous, and near the surface, it is infiltrated in many places by lymphoid and polyblastic cells.

COMMENT

In view of the arrangement, in almost typical fashion, of all the histologic elements that form the trachea, there seems to be no escape from the conclusion that the malformation was of tracheal origin.

Congenital fistulas of the neck are lined by all possible varieties of epithelium, and smooth muscle, and mucous glands are found in the walls of these fistulas. In no recorded instance, however, have I been able to find any notation regarding the presence of cartilage, such as has been described in the case under discussion, and no one has described an arrangement of epithelium, connective tissue, muscle tissue, mucous glands and cartilage, laid down exactly as they are in these sections.

After a most patient search of the literature had failed to disclose an analogous instance, the opinion of Prof. Ludwig Pick was sought.

Fig. 7.—Low power magnification of wall of fistulous tract.

He kindly went through the literature, also without finding an analogous instance, and reported that basing his opinion on the microscopic sections he would state unqualifiedly that the malformation was tracheal in origin.

The explanation of the formation of such a fistula as has been described must of necessity be more or less hypothetical, but it is herewith appended as a matter of record: An anomalous budding process takes place at some point on the ventral or lateral aspect of the primitive trachea. As the primitive trachea develops into its definitive form, the anomalous sprout grows downward in a plane anterior

to the sternum. Pick suggests that this is not a budding process, but rather should be interpreted as a double tracheal anlage, with the accessory trachea taking a course anterior to the sternum. At the point of budding or division, the anterior branch loses its connection with the foregut (from which the tracheal and lung anlage developed), becomes closed off, and gradually dilates to cystic proportions. The cyst stretches the overlying skin in the midline. Finally, rupture occurs, establishing a fistula, and resulting in skin retraction in the shape of the folds, or possibly teats, as in the present instance.