CERVICO-AURICULAR FISTULAE

A REVIEW OF PUBLISHED CASES WITH A REPORT

BY

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Congenital anomalies of the first branchial cleft are rare. Approximately 32 cases of cervico-auricular fistulae have been reported in the past hundred years: this includes the writer's own case in this review. The majority of sinuses and fistulae derived from the lateral cervical vestiges are concerned with the second pharyngeal pouch and its corresponding cleft.

Auricular and pre-auricular fistulae are not excessively rare, but cervico-auricular fistulae are still infrequently recorded. In some cases reported as cervico-auricular fistulae, there is little doubt from the description that these are auricular or pre-auricular fistulae.

Harding (1890) and König (1896) both recorded cases, but it was not until 1908 that Flint published a well-documented case report on this condition. Frazer (1923) postulated defects of closure of the first branchial cleft, though up to that time he thought that none had been reported. He stated that, 'If a cyst of such a vestige were present it would lie below the tube, behind (at any rate in part) the tensor palati and in front of the carotid and stylopharyngeus and if by any chance it opened in to the pharynx it would do so through the sinus of Morgagni.'

In the following review only those cases that are well documented have been considered.

Embryology

The branchial apparatus appears as but a transient stage in the development of the human embryo, appearing between the 3 mm. and 14 mm. stage. The embryology of these cases is concerned with the branchial or lateral cervical vestiges.

At the fifth week, when the human embryo is about 5 mm. in length, five ridges are visible on the outer surface in the region of the primitive pharynx (Fig. 1a). The walls of the pharynx are lined by five bars

or arches with four intervening clefts, the internal clefts being lined by foregut endoderm and the external clefts by ectoderm, but divided by a thin layer of mesoderm. Though there is confusion in the nomenclature, the general consensus of opinion names the pharyngeal clefts *pharyngeal pouches*, and the corresponding external clefts *branchial clefts*. Each arch has a central core of cartilage, a main blood vessel, and nerve.

FIG. 1a.—Horizontal section through a reconstructed 5-week human embryo to show the pharyngeal arches and their constituents. Externally the arches are covered by ectoderm, internally by foregut endoderm with an intervening layer of mesoderm. The roman numerals (i-iv) indicate endodermal pouches.

FIG. 1b.—A diagram of the pharyngeal region of a human embryo at approximately 6 weeks, demonstrating the relative overgrowth of the first and second arches.

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The muscular derivatives of each arch are supplied by the nerve of that arch. The nerve of the first arch is the mandibular division of the trigeminal nerve and supplies the muscles of mastication, the tensor palati, the tensor tympani, the mylohyoid, and the anterior belly of the digastric. The muscles of the second arch are supplied by the facial nerve, these muscles being those concerned with facial expression, the posterior belly of the digastric, the stapedius, and the stylohyoid. The third arch muscles make up part of the soft palate and the stylopharyngeus muscle, the nerve of supply being the glossopharyngeal. The constrictors of the pharynx and part of the soft palate are derived from the fourth arch and the nerve is the superior laryngeal. The fifth arch nerve, which is the recurrent laryngeal, supplies the intrinsic muscles of the larynx.

The vascular derivatives of each arch are modified, and there is a reduction in the number of arch vessels to facilitate pulmonary respiration and suit the new conditions of life. The original arteries of the first and second arches persist as the mandibular and stapedial arteries. The third arch arteries remain; the right third arch artery contributes to the development of the right common carotid artery and the commencement of the right internal carotid artery. The left third arch artery forms the left common carotid artery and distally the proximal part of the left internal carotid artery. The right fourth arch artery becomes the right subclavian artery and part of the arch of the aorta, and the left fourth arch artery forms the arch of the definitive aorta. The fifth arch arteries are present for only a short time and the sixth arch arteries persist as the pulmonary arteries.

The cartilaginous part of the first arch gives rise to the malleus, incus, the spleno-mandibular ligament, and the central core of the body of the mandible. The second arch cartilage forms the stapes, the lesser cornu of the hyoid bone, the styloid process, the stylohyoid ligament persisting as the intervening portion. The greater cornu and the lower part of the body of the hyoid bone is formed from third arch cartilage, and the cartilage of the fourth and fifth arches make up the skeleton of the larynx.

The glandular derivatives of the pharyngeal arches form parathyroid and thymic tissue, and the palatine tonsils. The palatine tonsils develop from the endodermal elements of the second pouch. The thymus, together with the inferior parathyroid gland, is developed from the third pouch, the parathyroid gland in this case being drawn caudally by the thymic rudiment. The superior parathyroid gland is derived from the fourth pouch.

The external ear is developed from a series of six

tubercles which develop on the first and second arches on the dorsal end of the first groove which lies between. By growth and fusion, the tubercles and the immediately surrounding area give rise to the primitive pinna. This is situated around the end of the developing external auditory meatus. The tragus and its immediate area is derived from the first arch, and the greater contribution to the formation of the pinna comes from the second arch (Wood-Jones and I-Chuan, 1934; Streeter, 1922). The external auditory meatus is formed from two elements: the dorsal end of the first groove and an ingrowth of epithelium from this groove, the ventral end becoming flattened then disappearing. The endodermal derivatives of the first pouch are concerned with the epithelium of the internal part of the tympanic membrane, the tympanic cavity, mastoid air cells, the eustachian tube, and epithelium of much of the mouth and body of the tongue.

By the end of the sixth week, the growth of the first and second arches has exceeded that of the third and fourth and a sinus is formed, the pre-cervical sinus of His (Fig. 1b). The third arch still contributes in part to the skin over the anterior triangle of the neck; the fourth arch is completely overgrown. Two deep recesses are formed, and these are the cervical vesicles: meanwhile, the first pouch fills up ventrally to become part of the tongue, and the dorsal end, together with the dorsal end of the second pouch, forms the tubo-tympanic recess, this recess forming the eustachian tube and the middle ear; the mesodermic membrane forms the tympanic membrane.

Ectodermal remains may persist from the ventral part of the first groove, from any part of the second groove, and from the cervical vesicles. Endodermal remains may persist as cysts or sinuses and accessory glands.

The following anomalies of the branchial apparatus are recorded: branchial cysts, external sinuses, internal sinuses, complete pharyngo-cutaneous fistulae, cervico-aural fistulae, branchial cartilages, and cervical auricles.

Case Report

A girl aged 2 years was admitted with a tender swelling in the left submandibular region. This had been present for three days and the child had been pyrexial. Since birth there had always been a 'dimple' in the region of the swelling.

Physical examination revealed a tender cystic swelling approximately 5×2 cm. in the left submandibular region anterior to the sternocleidomastoid muscle, at the junction of the upper and middle third. No other abnormality was noted. Radiographs of the chest and mandible were normal. The swelling was incised and brownish pultaceous material was evacuated. This was sterile on culture.

FIG. 2.—Artist's impression of operative area.

In the ensuing three months there was a continuous discharge from the site of the incision and for the first time it was noted that the left external auditory meatus was moist with a 'mucus like' discharge. No opening was seen in the external ear and the tympanic membrane was normal.

The child was readmitted for curettage of the sinus, and histopathological examination of the curettings demonstrated brownish granulation tissue, heavily infiltrated with polymorph plasma cells, lymphocytes, and macrophages, and one or two foreign body giant cells. There was no evidence of acid-alcohol fast bacilli, mycelia, or other specific aetiology. Amylase content of the exudate was 250 Somogyi units; this low level suggested tissue fluid or plasma rather than salivary gland excretion. A sinogram using 45% Hypaque demonstrated a small cavity below the left angle of the mandible. There was no evidence of deep extension.

It was decided to explore the sinus and dissect out the tract. Methylene blue, when injected into the sinus in the neck, emerged from a small pit in the floor of the external auditory meatus about 1 cm. from the external surface of the ear.

An ellipse of skin was excised around the sinus opening in the plane of Langers lines, and two tracts were seen, one running posteriorly up to the region of the external ear and the other running anteriorly beneath the mandible. The incision was then extended similar to that used in parotid gland surgery. The posterior tract was dissected out and appeared to be cylindrical, 5 mm. in diameter and with a fibro-muscular wall. This tract admitted a silver wire probe 1 mm. in diameter. The tract passed deep to the inferior part of the parotid gland and the greater auricular nerve, posterior to the mandibular division of the facial nerve but superficial to the posterior belly of the digastric muscle. The main trunk of the facial nerve was not seen. The tract ended in a vestibule 1.0 cm. in

diameter, lined by epithelium with hairs clearly visible and surrounded by its own wall of cartilage, the cartilage being continuous with that of the external ear.

The anterior 'tract' was 2 cm. long and fanned out in the sub-mandibular region as a series of tails that ran superficial to the sub-mandibular structures. There was some doubt as to the nature of this 'tract' and its extensions, as it was thought that it might be a branch of the facial nerve. Faradic stimulation of these structures provoked no response: the mandibular division of the facial nerve was positively identified using this technique (Fig. 2).

The two tracts and the vestibule were excised. After operation there was slight weakness of the corner of the mouth but this quickly resolved.

Histopathology. The specimen consisted of a 5 cm. fistula of varying diameter. Blocks were taken through the entire length, and over a length of 1.5 cm. at the cephalic end, serial sections were cut and every tenth section was examined.

The superior or cephalic end, 1 cm. long, was the widest portion of the tract, forming an ampulla 1 cm. in diameter. Here a ring of cartilage was included in the anterior three-quarters of the wall, the posterior wall consisting of fibrous tissue only. The ring of cartilage was partly continuous and partly formed by discrete curved plates. The cartilage consisted of mature hyaline cartilage, the central rounded cells becoming flattened peripherally and showing transition to the cells of the perichondrium. The cartilage stopped abruptly at the lower end of the ampulla.

The lumen of the fistula was 5 mm. in diameter in the ampullary region and 2-3 mm. elsewhere. The wall was lined by stratified squamous epithelium showing numerous pilo-sebaceous follicles and islands of cartilage; many short fair hairs projected into the ampullary lumen. The characteristic four layers of normal epidermis were present in the lining. A few melanoblasts were scattered through the basal layer and a moderate number of apocrine sweat glands were present in the deeper dermis (Fig. 3).

About the middle of the tract, the epithelium was partially replaced by granulation tissue and a moderate inflammatory infiltrate was present in the adjacent dermis. The ampullary region was free of inflammation.

Bundles of voluntary muscle formed the cords tethering the inferior end of the fistula to the submental tissues.

Discussion

In reviewing the published case reports of cervico-aural fistulae, the ages ranged from 8 months to 45 years. There were 23 female and 9 male cases; in 14 it occurred on the left and in 18 on the right side of the head and neck.

In the 32 cases, the presenting signs and symptoms were as follows: 11 had a cystic swelling, either quiescent or inflamed below the angle of the jaw and concomitant with a chronic discharge from the

FIG. 3.—Photomicrograph of transverse section of middle of fistula. (H. and E. $\times 25$.)

external ear; 10 presented with a cystic swelling in the same site; and 11 presented as a chronic draining sinus below the angle of the jaw, chronic ear discharge being present in 7, and no ear discharge evident in 4.

Topography. In the majority of cases the site of the lower end of the fistulae was in an area inferior to the angle of the mandible and anterior to the upper and middle third of the sternocleidomastoid muscle. The tracts ran cephalad, inclining dorsally to the external ear and in all cases running superficial to the posterior belly of the digastric muscle. The anatomical relation to the parotid gland varied, in 2 cases running deep (Caldera, 1922; De Bord, 1960 and own case), in 3 within (Aimi and Takino, 1962; De Bord, 1960; Lallemand and Poncet, 1961), and in 4 superficial to the gland (Gore and Masson, 1959; Ladd and Gross, 1937; Pagano, 1955; Stark, 1959). The opening of the tract in the external ear varied from deep in the fundus of the external ear, to superficial between the tragus and antetragus. In 4 cases, though the tract was attached to the external ear cartilage, there was no opening of communication.

Facial Nerve. It is well established that nerves grow to muscles and follow their subsequent migrations, but why a particular nerve filament reaches the

muscle of supply is still uncertain. Once established, this connexion between nerve and myotome is permanent.

The position of the facial nerve in relation to the tract varies. In some cases the nerve is reported as being deep and in other cases superficial to the tract. Exploration and positive identification of the nerve were considered unwarranted in certain cases and the exact anatomical position was not recorded.

In one case the tract was medial to the zygomatic and mandibular branches of the nerve (De Bord, 1960). Following paresis of the facial muscles in another case, the facial nerve was explored from the stylomastoid foramen to the parotid gland; it was found that the nerve had been displaced by the cartilaginous bulb, posteriorly and inferiorly; it had then passed along the anterior surface of the mastoid process of the temporal bone, then forward and superficial to the cartilaginous bulb before entering the gland (Rankow and Hanford, 1953).

The nerve is that of the second arch, but terminates in part of the area derived from the first arch and, as the muscles of facial expression spread out to cover part of the first arch territory, one might expect to find the nerve superficial as it passes forward to reach the facial muscles. Conversely, if there is overgrowth of the first and second arches over the third and fourth, then the nerve should lie deep to the tract.

When the nerve passes superficial, the tract must be running fairly deep and the possibility arises of involvement of the endodermal pouch. If this is so, the tract should not communicate with the external ear.

In the 15 patients in whom the facial nerve was positively identified, 8 had the nerve running superficial and 7 had the nerve running deep to the tract.

The time of onset of the anomaly in relation to the change in character of the lateral surface of the foetal pharynx may well be an important factor in determining the final course of the seventh nerve. When interference with the normal process of growth in the region of the branchial cleft is early, then the tract may well be deep to the nerve and maintain this position; conversely, if the time of interference is late the nerve could be deep to the tract.

Radiology. Investigation of these anomalies by radiological methods proved unreliable. Use of radio-opaque media demonstrated the tract fully in only 4 cases (Lyall and Stahl, 1956; Oppikofer, 1946; Pagano, 1955; Stark, 1959). In 2 (Aimi and Takino, 1962 and our case), a localized cystic space was demonstrated, there being no evidence of extension, and in one (De Bord, 1960) a tract running deep to the parotid gland was noted.

Operative Technique. Nearly all the tracts were excised through an incision similar to that used in parotid gland surgery. 'Step ladder' incisions in Langers lines were occasionally used.

A useful adjunct to the operative technique was the use of methylene blue, which was injected into the lower end of the fistula either as a 0.5% solution, or as a mixture of hydrogen peroxide and methylene blue 5 : 1.

Faradic stimulation was used to identify the facial nerve in 2 cases.

Histopathology. Macroscopic examination of the tracts showed some as thin threads of tissue and others as cylindrical fibro-muscular structures, up to 5 mm. in diameter and 9 cm. in length. At the upper end of some of the tracts, cartilaginous structures were seen. These were noted in 4 cases as partly semicircular vestibules (Alvarez and Jacobson, 1961; Flint, 1908; Rankow and Hanford, 1953, own case), and in one as a well-developed cartilaginous sheath 4 cm. long and up to half the length of the tract: all were proximal to but communicating with the auricular cartilage.

Microscopic examination of the tracts showed stratified squamous epithelium lining the inner surface with the characteristic four layers of the

epidermis present in the lining. Sweat glands, hair follicles, and focal lymphocytic infiltration were also present. The cartilage consisted of mature hyaline cartilage.

Embryology. The fate of the branchial clefts in the human embryo is still extremely vague. Growth is so complex that the anomalies must be even more complex.

It is reasonable to suggest that a cervico-auricular fistula represents part of the ectoderm of the first cleft which has been pulled dorsally as the ear migrates backwards; this ectoderm may be manifest as a solid tract, or hollow tube, the first ectodermal cleft not having been closed. Necrosis plays an essential part in the development of the human embryo and the fistula may be a result of a breakdown in a solid ectodermal tract. This may be due to a poor blood supply. The term 'failure of closure' of the first ectodermal cleft is misleading, because this presupposes that there is a gap between the first and second; this is not so.

The presence of a cartilaginous bulb or sheath in 4 cases may be explained as part of a misplaced auricular tubercle, or as part of some inductive action of the ectodermal tract on the surrounding mesenchyme.

Until further evidence is available concerning the fate of the external clefts in the human embryo, it is convenient to define a cervico-auricular fistula as a fistula derived from a persistence of part of the first ectodermal cleft, the skin roofing in the middle portion of the tract.

Summary

A series of 31 reported examples of cervico-auricular fistulae have been collected and one further case added.

The diagnosis is made more certain by noting two physical signs, namely a chronic discharging sinus in the neck and a discharge from the external ear when other aural pathology has been excluded. There is little doubt that this anomaly is more common than previously realized, and that cases are missed.

Radiological investigation may well prove more satisfactory if a less viscous radio-opaque solution is used.

At operation the facial nerve may lie superficial or deep to the tract. The exposure of the main trunk of the facial nerve is unwarranted if it is not readily visualized. There has been a low incidence of paresis of the facial muscles.

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