

Review Article

EMBRYONIC CYSTS, SINUSES AND FISTULAE OF BRANCHIAL ORIGIN

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Abstract :

This brief review hopes to provide basic embryological and anatomical information on location, cause and identification of congenital developmental anomalies related to formation of pharyngeal (branchial) arches to paediatricians, otolaryngologists, obstetricians and neonatologists. The pathological manifestations due to neglect or oversight of these development defects have potential to cause a wide range of clinical disorders.

Correct and timely recognition of these congenital developmental defects and their redress through appropriate surgical and clinical interventions require clinicians to possess a rudimentary grasp of embryological processes

Keywords : branchial, cysts, sinuses, fistulae

Introduction :

Early recognition and correct diagnosis of congenital lateral cervical dysgenic embryological structures require that a paediatrician have a thorough knowledge of embryology. Congenital birth and developmental malformations, especially those involving the head and neck may, require specialised surgical interference. This paper attempts an overview of the 'side of the neck' developmental embryopathies¹.

As early as the 5th week of growth, the 5 mm embryo shows discernible branchial or pharyngeal arches at the cranial end of its foregut and surrounding area^{2, 3}. Despite the complexity and specificity of the differentiation of ecto-endo-mesodermal entities, there are surprisingly a very low percentage of congenital defects in these areas.

F r e q u e n c y
notwithstanding this review may aid the novice clinician embryonic vestiges. The scope of this paper is limited to the pouch / cleft complex only (endoderm / ectoderm).

This malformation discussed here are in their order of frequency of occurrence.

1. Branchial cysts
2. External sinuses
3. Internal sinuses
4. Pharyngo – cutaneous fistulae
5. Accessory or aberrant glandular structures.
6. Other associated anomalies

Branchial cysts

The branchial cysts consist of almost 75% of all the lateral cervical vestiges. The salient features of these cysts are:

- a) Almost always unilateral
- b) No familial incidence
- c) Constancy of position
- d) Predominantly unilocular
- e) Lined in majority of cases by stratified squamous cells.

These cysts lie at about the level of hyoid, on the carotid sheath deep to the anterior margin of the sternocleidomastoid. Though most cysts are encapsulated completely, some may have an upper or lower pedicle or both. When present the upper pedicle extends from the

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supramedial part of the branchial cyst towards the pharynx passing between the external and internal carotid arteries, lateral to the XII (Hypoglossal) nerve.

The inferior pedicle, if present, extends to the medial edges of the sternocleidomastoid attaching itself to the skin at variable levels, as low as the clavicle. Pediculation of a branchial cyst can be confirmed through the application of gentle traction on the cyst: Dimpling of the skin or pharyngeal wall or both, confirms the attachment of a superior, inferior or both pedicles to the cysts.

External sinuses

The distinguishing characteristics of the external sinuses are:

- a) Present at birth
- b) These sinuses open in the lower third of the neck along anterior border of the sternocleidomastoid.
- c) Secondary infections and inflammations are common
- d) Bilateral in about a third of cases.
- e) When unilateral, more common in the right side.
- f) Familiar incidence common
- g) More often seen in the female than the male.
- h) Histologically the epithelium is of the stratified squamous cells type.

Deep sinuses extend upwards between the external and internal carotid arteries, resembling in their extension, to the superior pedicle of the branchial cyst. The calibre of the track is uniform along the length of the sinus, except when the sinus possesses an umbelliform upper extremity. The trumpet shaped external sinus may be confused with the inferiorly pediculated branchial cyst. A search for a patent lower extremity of the tract should confirm the possibility of external sinus.

Internal sinuses

The features of these sinuses are somewhat ambiguous

- a) Their frequency is not known.
- b) Most are symptomless and remain undiagnosed
- c) They open into the fauces, posterior to the palatine tonsil

- d) An extension from the palatopharyngeus invests the sinuses
- e) The lining epithelium is of the respiratory type, ciliated or pseudostratified columnar.

Complete pharyngocutaneous fistulae

These fistulae present in the under mentioned characters

- a) Opening in the same anatomical plane as the internal and external sinuses.
- b) Inflammatory changes frequent
- c) Bilateral in about a third of cases
- d) Familial disposition recorded
- e) Predominantly of the stratified squamous lining epithelium, through often towards the pharyngeal termination, showing ciliated columnar respiratory type cell lining.

Whilst most often the tracts of these complete fistulae are uniform dimension transversely, they may not uncommonly show a distended cyst suspended between two patent small calibre tracts extending to either side.

Other branchial and associated anomalies

Cervicoaural fistulae

- a) Rare
- b) Superiorly open into the external auditory meatus
- c) Inferiorly open into the skin over angle of the mandible
- d) The posterior belly of the digastric muscle lies deep to the course of the tract of these fistulae.

Branchial cartilages

Strictly speaking though these inclusions are not of either ecto or endodermal germ origin, cartilaginous nodules are occasionally found within the wall of branchial cysts or fistulae or even as unassociated findings situated in the cervical skin. Branchial cartilaginous nodules have been often discovered in or about the tonsil or in the tongue. They are considered as vestiges of the second arch cartilage (Reichert's).

Cervical auricles

Tags of skin, sometimes protruding from cutaneous orifices

of sinuses or fistulae are known as cervical auricles. These unusual auricles, when present, show bilaterality and a familial preponderance.

Associated anomalies

Branchial fistulae are sometimes associated with facial or cervical anomalies. Preauricular sinuses, accessory auricles, (auricular tubercles), hemithyroid or unilateral sternocleidomastoid are examples of some of these remote malformations.

Aberrant or accessory glandular vestiges

Vesicular, canalicular or acinar gland tissues often seen normally attached to the foetal thymus or parathyroid may sometimes persist in the neonate. These vesicular structures may distend resembling a branchial cyst. The persistent tissue is said to represent secondary buds or ducts from foetal thymus or parathyroid.

Accessory parathyroids have been described as intra – parathyroidal, intrathyroidic or even in the anterior mediastinum. Mediastinal parathyroid must be suspected when the usual parathyroids are of normal dimension in hyperparathyroidics.

The thymus may in the part or completely remain undescended. A normal thymus may show an accessory cervical portion or even aberrant thymic nodules. Intrathyroidic thymic tissue has also been recorded.

Discussion :

It is embryologically understandable that, excluding the first cleft to (ectodermal) and first pouch (endodermal) which form the external auditory meatus and the tubothympanic recess respectively, vestigial remnants are at least theoretically possible from all the other pouches and clefts. Indeed, all known lateral cervical vestiges can be easily correlated to defective derivations there from⁴.

One school of embryologists however, believes that all brachial cysts are remains of non-regressed epibranchial vesicles of epibranchial placodes which are thickened ectodermal patches overlying the dorsal extreme of the arches. They play a role in the formation of the cranial nerve ganglia, especially the facial nerve in man and the glossopharyngeal and vagal ganglia in lower vertebrates.

In frequency, the first arch remnants form only 5% of cases seen while the remaining 95% are defects from the 2nd arch. The remaining arches rarely produce any recognizable developmental defects.^{5,6}

Whatever the rationale and theory surrounding the appearance of the congenital lateral cervical vestiges, the specificity of their location, distinct histopathological features and definable anatomical relations are giveaways that every clinician should be familiar with, especially those from specialties that most often and most intimately comes into contact with the age groups that manifests the branchial lateral cervical developmental anomalies

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