Where Do These Neck Masses, Fistulae and Sinuses Come From?

Yoav Parag M.D., Kalliopi A Petropoulou M.D., Charles R Fitz M.D.
Children’s Hospital of Pittsburgh, University of Pittsburgh Medical Center, Pittsburgh, PA

Introduction

During the 3rd week of gestation the human embryo has formed 3 cell lineages (ectoderm, mesoderm and endoderm) that will form all adult tissues and organs. The embryo at this stage can be likened to a hollow tube with a central cavity that represents the primitive gut. The external surface of the embryonic tube is composed of ectoderm, the internal surface is composed of endoderm and there is an intermediary layer of mesodermal cells. These arches are defined externally by ectodermal infoldings known as the pharyngeal clefts and internally by invaginations known as the pharyngeal pouches. After further differentiation 5 arches continue to develop with 5 complimentary pairs of clefts and pouches.

The Pharyngeal Clefts

The first pharyngeal (branchial) cleft and pouch participate in formation of the ear. The cleft elongates in a medial direction and the pouch invaginates laterally. These structures eventually come to lie in close apposition. The first pharyngeal cleft will become the external auditory canal and the interface with the pouch will become the tympanic membrane. Rarely a duplication of the first pharyngeal cleft occurs, resulting in formation of the first branchial cleft cyst. These cysts are typically located inferior or ventral to the external acoustic meatus and may become apparent as periocular swelling. They may also drain externally through a fistulous tract, usually connecting to the external auditory canal.

The other pharyngeal clefts do not form adult structures. During the fourth and fifth weeks of life the second pharyngeal arch rapidly expands caudally and covers the other arches and clefts, effectively enveloping the remaining clefts in a transient, ectoderm-lined cavity called the lateral cervical sinus. In normal development this cavity disappears, however, if it persists it will become a lateral cervical cyst. If only one of the clefts persists as opposed to the whole sinus, it will be referred to as a branchial cleft cyst.

The Arches

Each arch contains a core of mesenchymal tissue that will give rise to the future muscles, bones and cartilages of the face and neck. Each arch is supplied by a solitary artery and innervated by a solitary nerve. These neurovascular associations continue throughout the development of the head and neck.

The Pouches

The pharyngeal pouches give rise to important structures in the adult.

The first pouch forms the tubotympanic recess, eventually forming the adult middle ear space and mastoid air cells. The second pouch gives rise to the parathyroids.

The first pharyngeal pouch forms the external auditory meatus and the tympanic membrane. The second pharyngeal pouch gives rise to the stapedius muscle.

The first pouch contains cells that differentiate into the calcitonin producing thyroid C cells. Structures of the third pouch:

During the 3rd week of gestation the human embryo has external endoderm) that will form all adult tissues and organs. Each arch contains a core of mesenchymal tissue that will give rise to the future musculature of its respective region. The thymic primordia elongate inferomedially as thyopharygeal tubes. They detach from the pouches and migrate to their resting position in the superior mediastinum. These tubes eventually involute. However, if they do not, thymic rests and cysts will be seen along the path of migration of the thymic primordia. (Fig 10a, b, c)

These tubes eventually involute. However, if they do not, thymic rests and cysts will be seen along the path of migration of the thymic primordia. (Fig 10a, b, c)

Internal opening

External opening

The thymic primordia elongate inferomedially as thyopharygeal tubes. They detach from the pouches and migrate to their resting position in the superior mediastinum. These tubes eventually involute. However, if they do not, thymic rests and cysts will be seen along the path of migration of the thymic primordia. (Fig 10a, b, c)

Clinical presentation may be pits or palpable masses.

If a mass lesion is found the images should be scrutinized for less conspicuous sinuses or fistulae.

Take Home Points

Figure 1: Schematic drawing of the embryonic tube at around 4 weeks of gestation (a). The pharyngeal arches are marked by the external clefts and internal pouches. A coronal section through the embryonic tube (b) shows the mesenchymal core of each arch. Note the central prominence at the site of the future tongue primordium.

Figure 2: 3D ultrasound is not only performed during the second and third trimesters but also in the neonatal period. (a) Normal thyroglossal duct with no cystic or solid extension. (b) Thymopharyngeal diverticulum extending from the base of the tongue. (c) Thyroglossal cyst with solid extension.

Figure 3: The thyroglossal duct can be found in several anatomic locations. During the development of the thyroid gland the thyroglossal duct persists at one of two sites within the fetal neck. The most common location is the isthmus of the thyroid gland just inferior to the hyoid bone. Less commonly, it may be found in the floor of the mouth or base of the tongue. (a) Posterior view of normal right thyroglossal duct anatomy. (b) Coronal view of normal right thyroglossal duct anatomy. (c) Sagittal view of normal right thyroglossal duct anatomy. (d) Sagittal view of normal right thyroglossal duct anatomy.

Figure 4: The cystic thyroglossal duct may appear as a cystic mass midline or parathyroid gland cyst. The cystic thyroglossal duct may also contain a solid component. The cystic thyroglossal duct may also contain a solid component. The cystic thyroglossal duct may also contain a solid component. The cystic thyroglossal duct may also contain a solid component. The cystic thyroglossal duct may also contain a solid component. The cystic thyroglossal duct may also contain a solid component. The cystic thyroglossal duct may also contain a solid component.

Figure 5: Schematic representation of the adult neck showing the typical location of branchial cleft cysts and the typical draining locations of cervical sinuses.

Figure 6: Schematic representation of the adult neck showing the typical location of branchial cleft cysts and the typical draining locations of cervical sinuses.

Figure 7: Schematic representation of the adult neck showing the typical location of branchial cleft cysts and the typical draining locations of cervical sinuses.

Figure 8: Schematic representation of the adult neck showing the typical location of branchial cleft cysts and the typical draining locations of cervical sinuses.

Figure 9: Schematic representation of the adult neck showing the typical location of branchial cleft cysts and the typical draining locations of cervical sinuses.

Figure 10: a, b, c. 9 year old male with a newly found right neck mass. Sagittal and axial FSET2W sequences reveal cystic masses in both carotid spaces bilaterally (Fig 8a, b arrows). The right mass extends down to the carotid bifurcation. The left mass extends down to the dissection plane. (Fig 10a Fig 10b Fig 10c)

Figure 11: Schematic representation of the adult neck showing the typical location of branchial cleft cysts and the typical draining locations of cervical sinuses.

References


Figures 1, 3, 6, 7 and 9 were adapted from KL Morre, TVN Persaud. The Developing Human; Clinically Oriented Embryology 6th Edition 1998 W.B. Saunders