Neck Masses of Congenital Origin

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Palpation of the neck of infants and children is an important part of the pediatric physical examination. The general pediatrician is usually searching for lymph nodes associated with upper respiratory infections or infections of the face or scalp; the pediatric endocrinologist is evaluating the thyroid gland; the pediatric oncologist is looking for tumor, either primary or metastatic; the pediatric cardiologist is feeling for carotid pulsations; and so forth, down the line of subspecialities. Any or all may come across the three major congenital masses in the neck, and while these are not serious problems in childhood, they can be diagnostically confusing. A knowledge of the embryology and anatomy of these masses is helpful in making an accurate diagnosis. The three lesions to be discussed are thyroglossal duct cysts, branchial cleft cysts and cystic hygroma. Of the benign congenital masses in the neck, 72 per cent will be cysts of the thyroglossal duct, 24 per cent will be branchial cleft cysts, and 4 per cent will be cystic hygroma.

Thyroglossal Duct Cysts

In the third week of embryonic life the thyroid anlage begins to develop on the floor of the pharynx at the base of the tongue. As the embryo grows, the developing thyroid gland descends in the neck, maintaining its connection to the tongue by a narrow canal, which is the thyroglossal duct. At the tongue end of the duct, the epithelial cells differentiate into tongue mucosa and persist as the foramen cecum of the tongue. At the thyroid end, the duct epithelium is induced to form thyroid tissue and, in some individuals, a pyramidal lobe of the thyroid gland. The mid-portion of the duct persists as a microscopic thread of undifferentiated epithelial cells tracking from the foramen cecum, through the hyoid bone periosteum or the bone itself, to the thyroid gland. For unknown reasons, this undifferentiated thread of cells may become active later in life and differentiate into columnar, ciliated, or squamous epithelium or into glandular tissue (sebaceous, salivary, or thyroid). When the cells are activated they produce a midline mass which is almost always cystic and contains mucoid material. Occasionally the cyst or another part of the duct will create a sinus tract to the skin at or just lateral to the midline. Anatomically these masses may be found anywhere from the foramen cecum to the thyroid gland, but most are adjacent to the hyoid bone.

Clinically the mass is usually observed by the mother, who may report that it is intermittently present. In fact, these cysts are best visualized when the neck is hyperextended and the usual small ones are not visible when the head is in neutral position. The majority of these are 1 to 2 cm in diameter, but some are 5 to 6 cm in diameter on presentation. Thyroglossal cysts are smoothly rounded with well defined margins and are slightly movable. Because of the thyroglossal duct attachments, the cyst will rise with tongue protrusion and move with swallowing. The cysts are nontender unless there is inflammation of the cyst or an associated sinus tract. They do not transilluminate well because of the overlying fascia of the neck. The sinus openings may be anywhere from the suprasternal notch to the hyoid bone. Careful palpation may enable the tract of the duct to be felt from the opening in the skin to the hyoid bone. The opening may show heaped up edges secondary to low grade chronic

inflammation. Drops of clear or cloudy mucus may escape from the sinus. If the sinus is present, the diagnosis is clear. If there is only the mass of the thyroglossal cyst, observation over several weeks will differentiate it from an inflammatory lesion. Other tumors that may be found in this region are lipomas and dermoid cysts. Lipomas are more lobulated and softer than thyroglossal duct cysts, whereas dermoids are attached to the skin and move with it. Rarely the thyroid gland itself will not descend to its normal position and will be found in the region of the hyoid bone. The absence of a thyroid gland in the normal position should suggest that the upper mass is the thyroid gland.

Treatment is probably not necessary for cysts less than 6 mm in diameter because they are asymptomatic and have no morbidity. The large ones and/or those that have been infected should be surgically excided. Sistrunk, in 1920, described the definitive surgical procedure. This includes excision of the cyst, the central 1 cm of the hyoid bone, and the tract of the duct up to the foramen cecum. Leaving behind remnants of the duct invites recurrence of either the cyst or the sinus tract.

Branchial Cleft Cysts

In the early embryo the branchial arches, branchial clefts, and pharyngeal pouches are the primitive structures from which the muscles, bones, and ligaments of the face and neck and their nerve and blood supplies will develop. Embryologists and surgeons have determined that the source of most cysts of branchial origin arise from remnants of the second branchial cleft. This cleft is really a groove separating the second and third branchial arches. Remnants of this cleft would lie anterior to the sternocleidomastoid muscle, between the internal and external carotid arteries and on a line between the tonsillar fossa and the junction of the middle and lower thirds of the sternocleidomastoid. In fact, when fistulas are found these course from the sternocleidomastoid to the posterior tonsillar pillar passing between the internal and external carotids. The cysts are lined by squamous or columnar epithelium and usually have lymphoid tissue adjacent to them. The sinuses are lined by stratified squamous, ciliated, or columnar epithelium and these tracts may have lymphoid tissue in the walls.

Clinically, the branchial sinuses present as a 2 to 3 mm slit anterior to the lower third of the sternocleidomastoid. They can be identified in the newborn infant. The importance of identifying the sinus is obvious as it is a portal of entry for pathogenic organisms. The cysts of branchial origin can appear at any age and usually are opposite the middle third of the sternocleidomastoid. The commonest age at presentation is in the early school age child. The presence of a rounded mass in the position described, which is nontender and only slightly movable, makes branchial cleft cyst a good possibility. If the patient is seen in the context of an acute upper respiratory infection, the mass may be mistaken for an enlarged lymph node. Indeed because of adjacent lymphoid tissue, the cyst may enlarge concomitant with the lymph nodes. Observation that the lymph nodes decrease in size after the infection, while the mass persists, should differentiate this lesion.

Treatment is surgical removal of all the remnant structures. In the newborn infant with a sinus tract, it is not advisable to explore the neck until the structures are large enouogh for appropriate dissection. The parents should be instructed in good skin care to prevent infection of the tract, and any infection that occurs should be promptly treated. In the older child with a cyst, the surgery is elective and should be done in those with visible masses.

Cystic Hygroma

These masses of lymphatic tissue appear to arise from the lymphatic sacs, which develop either as a direct bud from the jugular vein, or from mesenchymal tissue which coalesces and ramifies and then acquires venous connections. These lymphatic sacs appear to develop outpouchings that are then pinched off or sequestered to give rise to a cyst. The cysts have been found in the groin and axilla but most commonly occur in the neck. These lymphatic cysts have thin walls with sparse fine vessel vascularity, and are lined with endothelial cells. Narrow outgrowths of endothelial cells from the cyst wall can then insinuate themselves between muscle bundles, nerve fibers, or other tissues, produce lymph-like fluid, and enlarge in that manner.

Most extensions communicate with the main cyst cavity but there can be multiloculated cysts. Cystic hygroma is not a lymphatic malignancy, but it can damage surrounding tissue by pressure and can become large enough to compromise the airway or interfere with swallowing.

Most cystic hygromas (90 per cent) are found in the posterior cervical triangle behind the sternocleidomastoid muscle in the supraclavicular fossa. These can extend toward the midline, up toward the angle of the mandible, or out toward the crest of the shoulder. The mass may be quite large in the first few weeks of life. In Gross's series, 65 per cent were present at birth, 80 per cent discovered in the first year, and 90 per cent were found before the second birthday. The mass is usually soft, compressible, and may be smoothly rounded or slightly lobulated. It transilluminates well which confirms its cystic character. Occasionally any acute upper respiratory infection will trigger a sudden increase in size of the cyst, possibly because the normal lymphatic channels of the region become inflamed and partially obstructed. It appears that this obstruction causes lymph stasis which distends the cyst. Even though anastomotic connections to normal lymph channels are not easily demonstrated pathologically, these cases suggest that they do exist.

In the past, attempts to treat these cysts included aspiration, injection of sclerosing agents, radium therapy, and X-irradiation. Watchful waiting for spontaneous regression has also been recommended. With modern anesthesiology and pediatric surgical techniques, the safest and most satisfactory outcomes are from surgical dissection of the whole lesion. In the large infiltrating masses, the pediatrician should realize that this is not "shelling out" a clear cut mass, but is careful dissection through planes of the neck to get all the ramifications of the cyst. Occasionally, these procedures need to be staged when the mass is very large.