

CLINICOPATHOLOGICAL FEATURES OF IMMATURE NECK TERATOMA

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Teratomas are complex tumors composed of a collection of heterogeneous cells and/or organoid structures reminiscent of normal derivatives of all three germ cells.^{1,2} They occur in genital systems and other organs along the midline of the body, with similar morphology and classification. Ovarian teratomas constitute the major fraction and make up 15%-20% of all ovarian tumors,¹ yet teratomas are very rare in other locations, especially the neck.³⁻¹⁷ Benign mature teratomas have well-differentiated mature tissue representatives of all three germ cell layers, including neuroectodermal,¹⁶ while immature malignant teratomas have less differentiated elements from all or any of the three germinal layers. Histologically immature elements that are prognostically important include embryonal neuroepithelial structures that resemble medulloepithelioma, neuroblastoma, retinoblastoma or ependymoblastoma.¹⁶

Case Report

A female infant was born to a 21-year-old primigravida after a 37-week uneventful pregnancy. The delivery was by an elective cesarean section due to polyhydramnios and a large neck mass detected by an antenatal ultrasound. The mass was described as of mixed nature and suspected to be a cystic hygroma or teratoma.

The infant was born flat, bradycardic and was not breathing. A 3.0 size ETT was inserted with difficulty. Apgar scores were 3 at 1, 5 at 5 and 8 at 10 minutes. Her birth weight was 2.850 kg. An MRI was done, and suggested an extensive cystic hygroma involving both sides of the neck. The mass extended superiorly to the floor of the mouth and the base of the skull, laterally to the mandibles and carotid sheaths, and inferiorly to the upper margin of the innominate vessels. It caused marked compromise of the oropharynx and the upper airway. The mass was excised, and the gross specimen consisted of an irregular, encapsulated, multiple lobulated cystic mass

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measuring 10.5 x 7.5 x 5 cm in maximum dimension. The cut surface revealed mainly cystic to spongy lesions filled with straw-colored fluid. A calcified area was also identified.

Microscopically, sections revealed tumor composed of tissue derived from all three germ cell layers. A variety of immature tissue was recognized, such as fibrous tissue, immature cartilage, bone, muscles and undifferentiated mesenchymal tissue (Figure 1). The ectodermal component was represented by glial, immature neuroblastic and immature neuroepithelial tissue. Ependymal tissue (ependymal rosettes), choroid plexus and pigmented retinal epithelium were also seen (Figure 2). Most of the tissue was immature and had a haphazard arrangement. No other germ cell elements were seen. The large amount of immature neuroepithelial tissue was seen in all sections (more than three foci per low-power field in each slide), making the tumor immature teratoma grade 3.

Postoperatively the patient was weaned off ventilatory support and successfully extubated. At the time of discharge, the patient required 0.5 L oxygen via head box. She was maintaining oxygen saturation above 95% with mild subcostal recessions and an audible stridor. Her respiratory rate ranged between 55-65/minute. She was on full feeds via a nasogastric tube. A barium study was reported as normal. The plan was to slowly shift her to oral feeds as the respiratory distress resolved. Other than left microphthalmia, her system exam was unremarkable. She was on antibiotics for *Staphylococcus epidermidis* isolated from blood culture and *Pseudomonas* isolated from tracheal aspirate. Tumor markers were done: alpha fetoprotein was 12,473 IU/mL (normal 0-5.8) and BHCG was 1.36 IU/mL (normal <5). A repeat sample taken after two weeks showed alpha fetoprotein to be 1081 IU/mL. There was no further clinical follow-up of the patient.

Discussion

Teratomas arising in the neck are very rare.³⁻¹⁷ Upon reviewing the cases of teratomas of infancy and childhood reported in four major studies,^{5-7,13} only 16 of 335 teratomas (4.7%) were in the neck. Hajdu and associates reported the first well-documented case of a cervical teratoma to Hess, in 1854.¹⁴ A century later, fewer than 100 cases had been reported.^{3,4,12}

FIGURE 1. Immature teratoma showing mesodermal (cartilage), ectodermal (choroidal plexus-like tissue) and immature brain tissue on top.

FIGURE 2. Immature teratoma showing pigmented retinal epithelium, Homer-Wright rosettes (on left side) and small thyroid follicle (on the right side).

The term "neck teratoma" generally incorporates lesions arising in the anterior and posterior triangles of the neck and excludes those arising from the base of skull or from the spine. In the literature, attempts that have been made to subclassify teratomas of the neck into thyroid teratomas, cervical, or extrathyroid teratomas,¹² on the basis of their blood supply, have not been acceptable.¹⁷ Moreover, as in our case and most cases in the literature it was difficult to determine the origin of the main feeding vessels. In our case the tumor was huge, extending from the base of skull to the mediastinum. So the exact location in the neck, i.e., anterior or posterior triangle of the neck, was not possible to pinpoint. In the literature most of the teratomas originating anteriorly in the neck had some relation and connection to the thyroid. The tumor in our case was located posteriorly and did not involve the thyroid gland. This relationship to the thyroid gland was so constant that Roediger and associates have been able to argue persuasively that all teratomas presenting in the anterior neck region arise from embryonic cells in the primitive analogue of the thyroid gland.¹⁰ In the rare instances in which muscle and soft tissue are interposed, this may be due, in their opinion, to mechanical dislocation of embryonic teratogenic cells in a manner analogous to the dislocation of normal thyroid tissue that comes to lie adjacent to, and at some distance from, the normal gland.^{5,12} Other authors do not share this opinion and regard the relationship with the gland as entirely fortuitous,⁸ in view of the extreme rarity of intrathyroid origin.

There is no satisfactory explanation for the development of these tumors. Colton and co-workers reviewed the literature on teratomas of the neck in adults in 1978, and found a total of 10 cases.⁸

Most of the cases in the literature present clinically as a large oval mass in the neck and the consistency of the mass usually appears partly cystic and partly solid. There is no predilection for left, right, or median regions of the neck.¹⁷ All races and both sexes are affected with approximately equal frequency.^{12,13} The largest tumor reported in the literature had extended to the mastoid process superiorly and to the clavicles inferiorly. In our case, the tumor was even larger, extending more inferiorly in the mediastinum even up to the aortic arch.

Most neck teratomas are reported as mature, and malignant immature teratomas have been said to occur in approximately 5 percent of teratomas of the neck.⁹ The prognosis of neck teratomas is poor, with a large proportion of patients with cervical teratoma (more than 25% of those reviewed by Silberman and Mendelson) dying prior to surgery.¹² Hence, unless surgery is instituted without delay, the prognosis of cervical teratoma can be serious. This grave outlook seems to depend largely upon obstruction of upper respiratory passages. Tracheostomy may be very difficult without removal of the tumor in patients presenting with very large masses. Often these were so large and situated as to preclude access to the trachea. With

less than complete laryngotracheal occlusion, the degree of interference with deglutition may be severe. In prenatal life this causes polyhydramnios,¹¹ postnatally, it may lead to aspiration of secretions, atelectasis, and bronchopneumonia.

The operative mortality has been reported at between 9%¹² and 17%.^{14,15} Removal of the tumor is the treatment of choice for this condition. There is no evidence that radiation is beneficial, and it carries potential risks.¹⁷ Further studies are required to define the proper line of management and therapy for these patients.

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