The Turkish Journal of Pediatrics 28:123 - 128, 1986

HEAD AND NECK TERATOMAS*

Two cases with rare localizations

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Key words: germ cell tumor, head and neck, teratoma, thyroid gland

Germ cell tumors are neoplasms that develop from primordial germ cells of the human embryo which are normally destined to produce sperms or ova. It is the totipotent nature of the primordial germ cell which determines the histologic characteristics of various tumors of germ cell origin.

These tumors have various localizations, and are to be found, for instance, in the pineal, mediastinal, retroperitoneal and sacrococcygeal regions; it has been explained that these neoplasms originated from primordial germ cells deposited along the route of migration during embryonic life[1].

Both benign and malignant tumors may originate from primordial germ cells. Mature teratomas show benign features whereas embryonic carcinoma, choriocarcinoma, teratocarcinoma, endodermal sinus tumor (yolk sac tumor), dysgerminoma and ones with mixed germ cells make up the malignant group.

It has been reported that two percent of the malignant neoplasms occurring in the under-fifteen age group are gonadal and germ cell ones, and 0.4 percent of them are malignant teratomas[2].

Of the two cases of teratoma described here, one is benign with a thyroid localization and the other is malignant and originating in the oral cavity; they are presented because these types of tumor are rarely seen during the childhood period.

Case Report

Case 1

K.B., a three-day-old girl baby was admitted due to a mass in her neck. She was born at 40 weeks gestation to a 24-year-old multigravida mother after an

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uneventful pregnancy. The patient was asphyctic and had the mass at the time of birth.

On physical examination the patient appeared hypoactive. She had a large cervical mass that measured 8 × 10 × 15 cm and extended from the right mandibular area to the sterno-clavicular area. The lower portion of the tumor was smooth and rubbery and the proximal portion showed lobulation. Neither the right mandible nor a separate thyroid gland could be palpated. Other system findings were unremarkable (Figure 1).

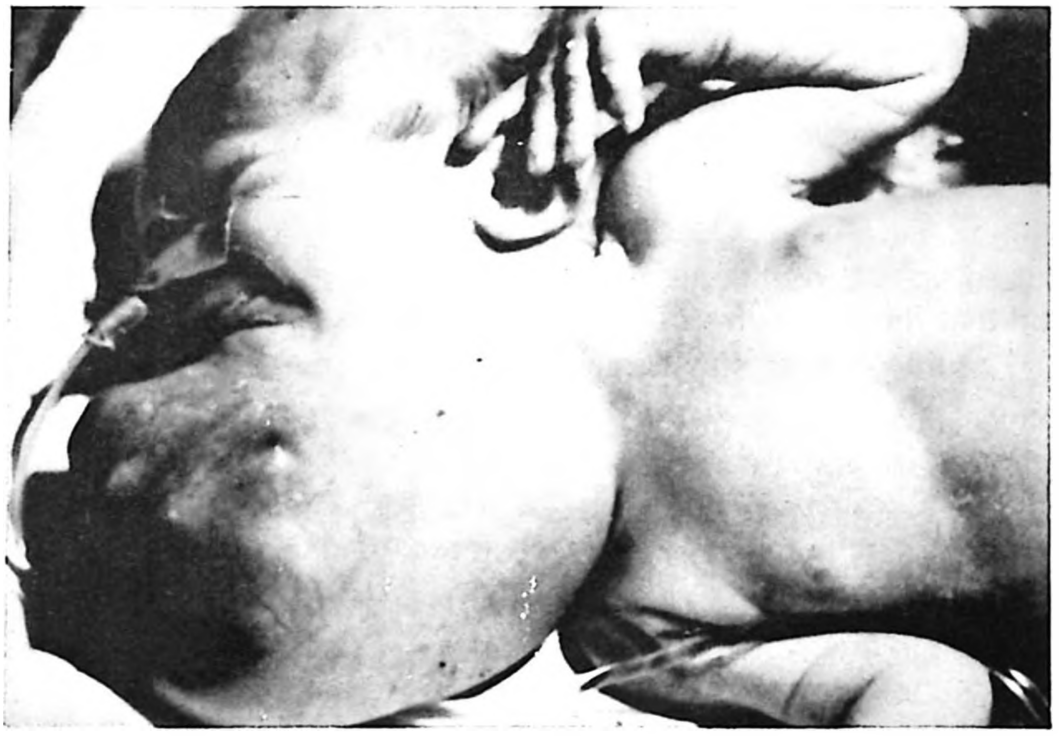

Figure 1
Case 1, benign thyroidal teratoma in the neck.

Laboratory studies including complete blood count, urine analysis, blood electrolytes, BUN, blood sugar, TORCH complex, serum T_3 and T_4 concentrations, and blood and urine aminoacids were all normal.

The tumor mass was excised with the capsula under general anesthesia. Histopathologic examination showed the surgical specimen was a well encapsulated and irregularly lobulated mass measuring, at maximum lengths, 12 × 8 × 6 cm. The cut surface showed lobulated, pale white areas intermixed with distinctly mucoid nodules. Microscopically, in haphazard arrangements, some mature somatic tissues such as striated muscle tissue, fibrous connective tissue, adult fat tissue, hyaline cartilage, squamous cell islands and cysts lined with mucous cells were observed. The histopathologic diagnosis was benign cystic thyroidal teratoma (Figure 2).

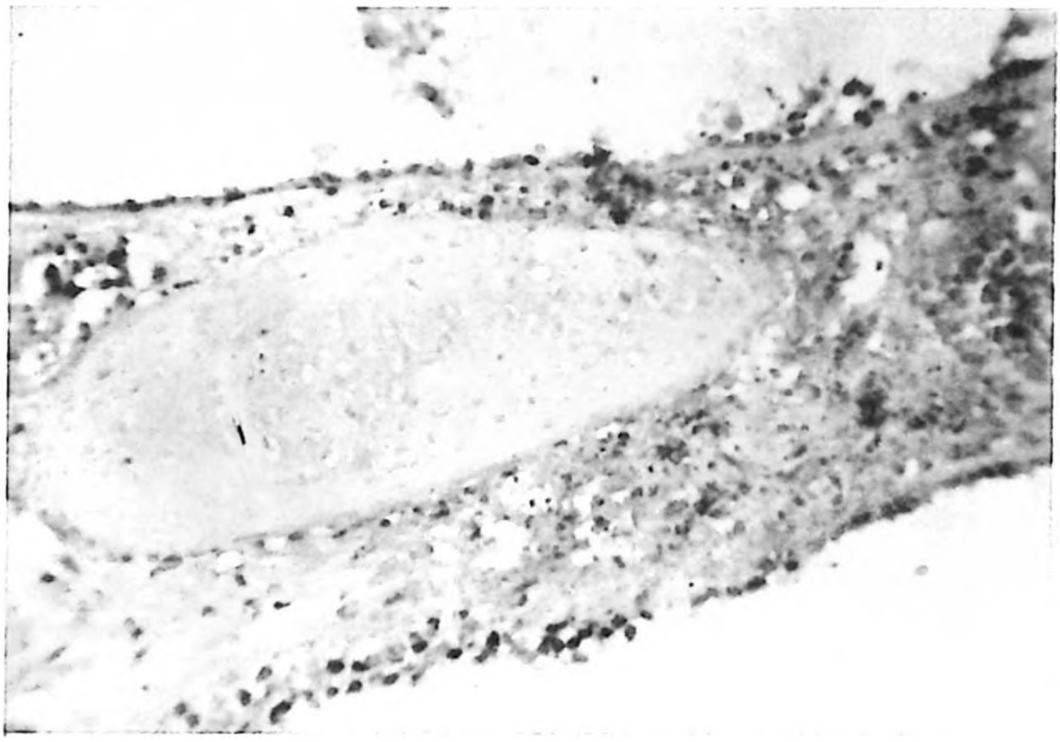

Figure 2

Case 1, hyaline cartilage and two cysts lined with mucous epithelium.

Case 2

G.B., a 13-hour-old girl baby was admitted due to a mass situated in the oral cavity. She was born at 38 weeks gestation to a 21-year-old mother. This was the second pregnancy. The mother had used some drugs to prevent nausea 40 days prior to the birth. Family history revealed the parents were first cousins.

On physical examination the patient was active in appearance. There was a 6 × 6 cm sized mass originating from the base of mouth and filling the oral cavity completely. It did not have definite limits and it pushed the mandible outside. Other system findings were within normal limits (Figure 3).

Laboratory studies including complete blood count, urine analysis, blood electrolytes, BUN, blood sugar, TORCH complex, serum T_3 and T_4 levels, blood and urine aminoacids, and chest X-ray were all normal.

The mass was excised totally including the right mandibular portion inside, under general anesthesia. Histopathologic examination showed that the surgical specimen was a well circumscribed but non-encapsulated mass with a piece of mandible on its pole. The cut surface of this lesion had multiple hemorrhagic, necrotic and cystic areas. Microscopically, some immature tissues such as neuroepithelium and immature epithelial island and some mature tissues such as

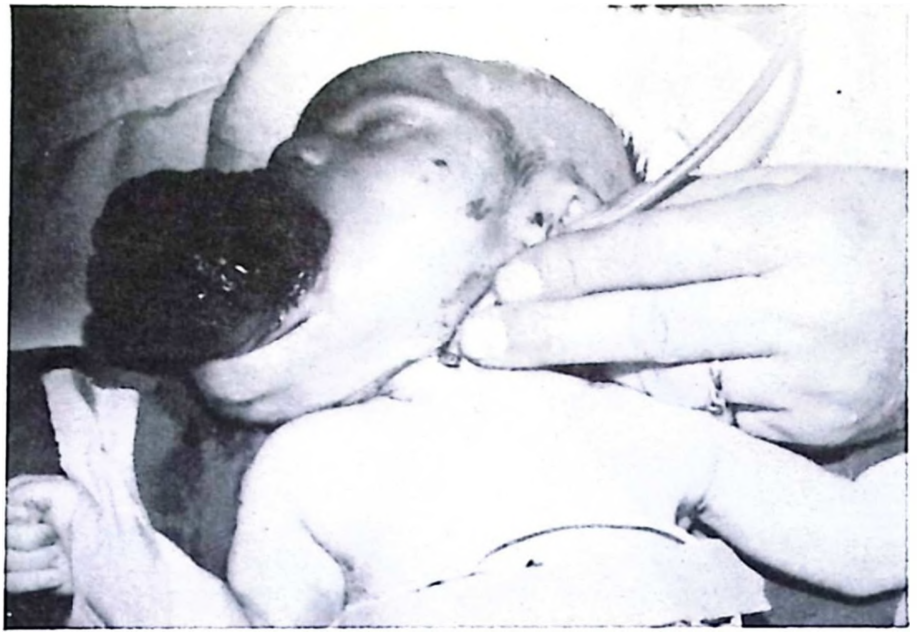

Figure 3

Case 2, malignant teratoma protruding from the mouth.

hyaline cartilage and squamous cell islands were observed in a fibromatous stroma. The histopathologic diagnosis was malignant teratoma of the oral cavity (Figure 4).

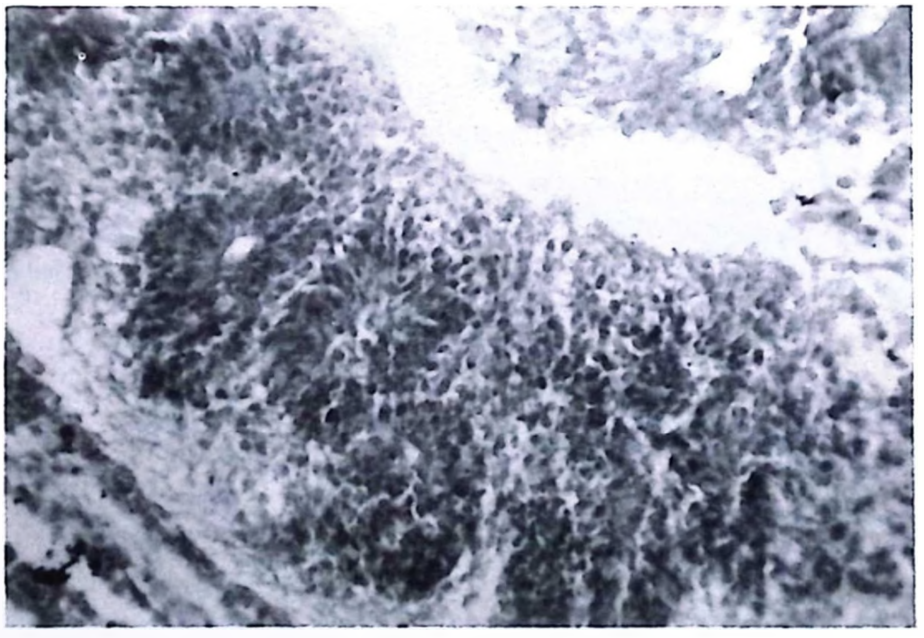

Figure 4

Case 2, immature epithelial tissue.

Discussion

Teratomas are neoplasms that originate from primordial germ cells differentiating in a way similar to that of an embryo. They may have a benign or a malign character and occur in various localizations. It has been reported, based on the data of a study of 526 subjects, that 42 percent of childhood germ cell tumors are situated in the sacrococcygeal region, three percent of them in the neck, and two percent of them in the head (excluding the CNS)[3]. Tınaztepe et al reported that three percent of 110 cases showed a cervical localization (including the thyroid gland)[4]. In the central nervous system, teratomas occur in the pineal region or replace a portion of the cerebral hemisphere[5]. The nasopharynx, oral cavity, orbit and cervicothyroidal region are the sites of development of extracranial head-neck teratomas. With a few exceptions, these tumors are recognized at or shortly after birth[6]. Cases of teratomas of the tongue and tonsils in newborns and infants have been reported[7].

The epignatus is a clinically impressive lesion that manifests as a solid and cystic mass protruding from the mouth with an attachment to the basicranial region through a midline palatal defect[8].

Newstedt and Shirkey[9] in a review of the literature found 82 cases of teratoma in infants and four in adults. Of the 82 teratomas in infants, 81 were clinically benign and one was apparently malignant. With the adults, however, three of the four teratomas were malignant. The case of an adolescent girl with a malignant thyroid teratoma has also been reported in the literature[10].

The first of our cases was a benign thyroid teratoma recognized as a large mass in neck. The second was a malignant teratoma in which the tumor originated from the oral cavity and protruded from the mouth. We have not encountered any articles in the literature reporting malignant teratomas originating in the oral cavity. Therefore, we believe that our case might be interesting due both to its localization, which is unusual, and to its histopathologic features.

Summary

Childhood teratomas with head-neck localization are rare and the tumors located in these regions have seldom a malignant character. Here two cases of teratoma, one benign and originating in the thyroid gland, the other malignant and developed in the oral cavity are presented and relevant literature is reviewed.

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