Teratoma is a germ cell neoplasm which has any combination of ectodermal, mesodermal and endodermal elements. The frequency of teratoma is 1 in 4,000 live births and 1-2% of the cases seen in the head and neck region. Approximately 75% of this tumor group are mature teratomas, and about 12% of the cases are malignant and lethal. Head and neck teratomas are most commonly seen at the cervical region, oropharynx is the second most common location. Congenital cervical teratoma is approximately 3% of the teratomas seen in childhood/infancy. Since head and neck teratomas frequently presents with respiratory distress, surgeons should be aware of the clinical presentation and pathology of the teratoma.

Choroid plexus papillomas (CPP) are benign and slow-growing tumors derived from choroid plexus epithelium. They are predominantly seen in children, and in the lateral ventricle, while they are seen in the fourth ventricle in adult. Many different localizations of the tumor have been documented. The incidence of CPP is 0.3-0.6% of all brain tumors; 2-4% of the cases occur under the age of 15, and 10-20% of the cases occur within the first year of life. Teratoma is an unusual location for CPP. Four cases of CPP located in teratoma have been published in the literature so far. All these cases were found in ovarian teratoma. Unlike the other cases, our case was at the cervical region.

Up to 90% of childhood teratomas contain derivatives from all three embryonic germ layers. Though teratomas develop from the three germ cell layers, some reports showed that they may arise from a single germ cell. The present teratoma included totally mature neuroglial tissue.

Case Report

A female neonate born with cesarean section at 39 weeks of age weighting 3350 grams had a mass...
on her neck at birth. The voluminous cervical mass was detected prenatally. It was followed up by a gynecologist until birth. She was operated on two weeks after birth. On postnatal magnetic resonance imaging (MRI) a 9 cm diameter mass with cystic and solid components was observed on the right side of the neck. It extended from the parapharyngeal region to the subcutaneous soft tissue. Teratoma, rhabdomyosarcoma and neuroblastoma were considered in the differential diagnosis. Tumor markers were within normal level. The lesion was removed by protecting vital organs and skin. There was no complication during the postoperative period. Macroscopically; the mass had cystic and solid areas. Microscopically; a CPP was found in the cyst. The diameter of the lesion was 0.6 cm. It was arranged of papillary structures with fibrovascular cores. Papillary structures showed complexity and there were acinar structures in some areas (Fig. 1). Epithelium of the CPP showed variably pseudostratification, surface epithelium of it was flat, and nuclei were oval and elongated (Fig. 2). Necrosis, mitosis and solid pattern was not observed. The other areas consisted of mature neuroglial tissue entirely (Fig. 3). Neuroglial tissue was hypercellular in some areas, but non-neoplastic. Immunohistochemical study showed that neuroglial tissue was positive for Alpha Thalassemia/Mental Retardation Syndrome X-Linked (ATRX), Glial fibrillary acidic protein (GFAP), Oligodendrocyte Transcription Factor 2 (OLIG2), and negative for The p53 gene (P53) and Isocitrate dehydrogenase 1 (IDH1). Antigen KI-67 (Ki67) index was very low.

Written informed consent was obtained from the patient’s parents for publication.

Discussion

The teratoma of the head and neck region can shows unpredictable behavior and dramatic clinical presentation. Because of the potential for rapid growth of teratomas and as a result airway compression, the treatment of cervical and nasopharyngeal teratomas is immediate excision. Cervical teratoma is not common. In the literature 217 cases of cervical teratoma have been reported until 2009, and 90% of the cases were diagnosed in the pediatric population.7

**Fig. 1.** HE X 200: Choroid plexus papilloma; papillary structures with fibrovascular cores and acinar structures in some areas.
The present case included CPP in teratoma of the head and neck region.

Choroid plexus in teratoma is commonly seen. It is important to separate a teratoma with choroid plexus from a teratoma with choroid plexus tumor. CPP contains more complex papillary architecture than non-neoplastic choroid plexus, and epithelium is flat in comparison with normal cobblestone-like surface. Only

**Fig. 2.** HE X 400: Choroid plexus papilloma; variable pseudostratification of epithelium, flat surface epithelium, oval and elongated nuclei.

**Fig. 3.** HE X 200: Neuroglial tissues in different regions of teratoma (hypercellular and hypocellular areas).
four CPP in teratoma cases have been reported so far (Table I). In the present case, the diameter of the papilloma was very small, and was only visible at microscopic examination. In our case there was a delicate fibrovascular stalk covered by a single layer of cuboidal epithelial cells, arranged in a papillary configuration with fingerlike projections of tissue in the teratoma. The nucleus of the epithelium was slightly elongated.

Monodermal teratoma which includes only neuroglial tissue is uncommon in the literature. Akbulut et al. described an ovarian teratoma that mainly consists of neurogenic elements including glial tissue, cuboidal cells lining the lumen of the cyst resembling ventricular ependyma, melanotic cells, ganglion cells, peripheral nerve and choroid plexus. Sharma et al. reported a teratoma case that was predominantly composed of neurogenic tissue in the retroperitoneal region. Williams et al. presented a monodermal teratoma which was named neurogenic ovarian cyst. It included neuroglial tissue with astrocytes, occasional Rosenthal fibers, oligodendrocytes, ependymal cells, scattered neurons with an overlying arachnoid layer and cerebellar tissue components. Our case consisted of only neuroglial tissue which included astrocyte, oligodendrocyte and choroid plexus. But it was not a monodermal teratoma because of the presence of choroid plexus. The choroid plexus has dual embryonic origin, with the choroid plexus epithelium originating from the ectoderm and the central stroma from the mesoderm.

Our case was unique with its neuroglial component that includes CPP. CPP should be considered in the differential diagnosis with other papillary neoplasms which could be primary or metastatic such as serous, clear cell or endometroid gynecologic tumours, mesothelioma, papillary thyroid carcinoma, and lung adenocarcinoma.

Immunohistochemically, choroid plexus epithelium-derived tumors react with pancytokeratin and epithelial membrane antigen (EMA), sometimes with vimentin, S-100 protein and synaptophysin. Focal GFAP reaction can be seen in epidermal differentiated choroid plexus tumor. But none of them are used in the differentiation of primary choroid plexus tumor from metastasis.

Two specific markers of choroid plexus tumor have been recently described. Kir7.1, a potassium inwardly-rectifying channel family member that may play a role in the transepithelial transport

| Case and reference | Year | Age | Localization | Diagnosis | Teratomatous elements | Size | Treatment |
|--------------------|------|-----|--------------|-----------|-----------------------|------|-----------|
| I                  | 2006 | 14  | Ovary        | CPP       | Lungparenchyma, neuropil, skin withadnexal structures, neuroglial elements, adiposetissue | ND   | ND        |
| II                 | 2011 | 26  | Ovary        | ACPP      | Skeletalmuscle, Adiposetissue | 9 × 7 × 7 cm | Cystectomy |
| III                | 2013 | 32  | Ovary        | CPP       | Bone, cartilage, skin withadnexal structures, respiratorytype epithelium, ependyma | 8 × 3 cm | Oophorectomy |
| IV                 | 2015 | 23  | Ovary        | CPP       | | 22.5 × 20.5 cm | Salpingo-oophorectomy |
| Present case       | 2018 | Newborn | Neck     | CPP       | Neuroglialelements | 10 × 9.5 × 8.5 | Resection |

ACPP: atypical choroid plexus papilloma, CPP: choroid plexus papilloma, ND: not described.
of potassium. Stanniocalcin-1, a glycoprotein normally expressed in human choroid plexus that might participate in the regulation of cerebrospinal fluid calcium levels. Both normal choroid plexus and choroid plexus neoplasm react for these two immunohistochemical markers. Therefore, Kir7.1 and Stanniocalcin-1 are sensitive and specific diagnostic markers for choroid plexus tumor. They are used in the differentiation of metastasis from choroid plexus tumor.

A choroid plexus papilloma located in a teratoma at neck region was presented. Pathologists need to be aware of this entity in the distinction from other papillary neoplasms that may be primary or metastatic.

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