

A Craniocervical Teratoma with an Encephalocele-Like Appearance

Ensefalosel Görünümlü Kranioservikal Teratom

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ABSTRACT

Teratomas are rare congenital tumors. Teratomas frequently occur in adult ovaries, but can be located primarily in the sacrococcygeal zone and intracranial compartment in newborns. Teratomas are rare in the head and neck region. Teratomas arise following a sequence of cells derived from more than one germ layer at different regions of the body due to a change in location of germ cells, and contain ectodermic, endodermic, and mesodermic tissues. Herein we detail the follow-up and treatment of a newborn with a mass that was located at the back of the head and was referred to our Neurosurgery Clinic with a pathologic diagnosis of a grade II-III immature teratoma resembling an encephalocele.

KEYWORDS: Craniocervical teratoma, Encephalocele, Newborn

ÖZ

Teratomlar nadir görülen konjenital tümörlerdendir. Yetişkinlerde overde sık görülürken yenidoğanda en çok sakrokoksigeal lokalizasyonda ve daha sonra da intrakranial kompartmanda görülür. Baş ve boyun bölgesinde ise oldukça nadir görülürler. Teratomlar dorsal orta hat konjenital tümörlerinin en yaygınıdır. Teratomlar germ hücrelerinin yer değiştirmesi sonucunda vücudun farklı yerlerinde birden fazla germ yaprağından türeyen hücrelerin sıralanması ile oluşur ve ektodermal, endodermal ve mesodermal dokular içerirler. Bu makalede, Nöroşirürji kliniğine başının arkasında kitle şikayeti ile başvuran ensefalosele benzeyen fakat patolojik tanısı grade 2-3 immatür teratom olan yenidoğanın takip ve tedavisini sunduk.

ANAHTAR SÖZCÜKLER: Kranioservikal teratom, Ensefalosel, Yenidoğan

INTRODUCTION

Teratomas are rare congenital tumors. Teratomas frequently occur in adult ovaries, but can be located primarily in the sacrococcygeal zone and intracranial compartment in newborns (1,6). Teratomas are very rare tumors with a prevalence of approximately 1/13000 (7). Teratomas are considered the most common congenital tumors located on the dorsal midline. Teratomas arise following a sequence of cells derived from more than one germ layer at different regions of the body due to a change in location of germ cells, and contain ectodermic, endodermic, and mesodermic tissues (3,4,7). Embryonal teratomas most commonly occur in the sacrococcygeal region and are designated as sacrococcygeal teratomas. Indeed, sacrococcygeal teratomas are the single most common tumor found in the midline of the sacrococcygeal region of newborns. Teratomas located in the head and neck region are very rare. We present a patient with a teratoma of the head and the treatment provided.

CASE REPORT

A 1-day-old newborn was referred to our clinic for evaluation of a mass located on the back of the head. The general condition of the newborn was good, and the newborn was alert with an interest in the environment. There was a soft tissue mass

which resembled a cystic structure, 11 x 8 x 7.6 cm in size, that appeared to be an encephalocele in the left occipitotemporal region (Figure 1). No neurological deficits were noted during the neurological assessment; the lower and upper extremities were active. A brain computer tomogram showed a mass lesion located in the left frontoparieto-occipital region that extended from the subdermis to the left half of the neck after including the posterior part inside the left shoulder, and a mass lesion was reported within the calcified fatty cystic and solid areas, 11 x 8 x 7.6 cm in size, with an exophytic extension (Figure 2). The patient had an electrolyte imbalance pre-operatively, which was corrected. During surgery, the patient was placed in the prone position on the operating room table. A vertical skin incision was made over the mass and the contents of the sac were excised by dissection from the temporal and parietal regions. The cervical component contained fatty structures. The soft tissue of the mass had no connections with the brain tissue, whereas the content of the soft tissue of the mass contained hairy and finger-like structures and a tissue that resembled a shell that covered these structures and cystic components (Figure 3). After hemostasis, the skin was primarily sutured. Antibiotic therapy was initiated after surgery and wound care was performed. During follow-up, the wound healed well and the patient was



Figure 1: Preoperative presentation.

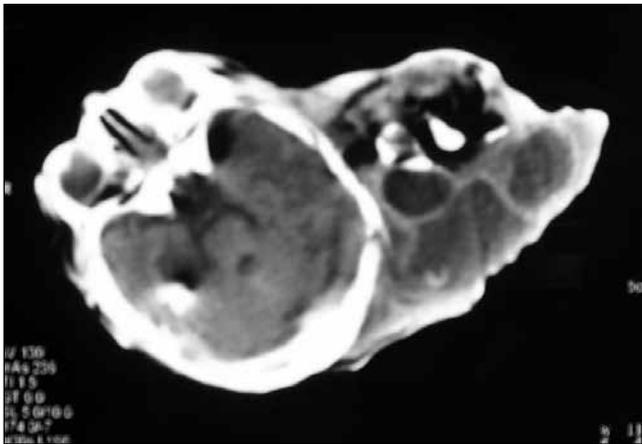


Figure 2: CT image of soft tissue mass.



Figure 3: Total occipitotemporal mass extracted.

discharged from the hospital to be followed in the outpatient clinic. The pathologic study of the excised mass was reported as a grade II-III immature teratoma (Figure 4). The patient has been followed for 1 year without a recurrence (Figure 5).

DISCUSSION

Even though teratomas are rare tumors, they are the most frequent congenital tumors located in the dorsal midline, and



Figure 4: Histopathological image of soft tissue mass.



Figure 5: Postoperative presentation.

occur in 1 in 13000 live births (7,8). The migration of primordial germ cells during the early stage of embryogenesis into the midline structures of the yolk sac can be blocked, and germ cell tumors, including teratomas, may subsequently develop (8). Teratomas are differentiated from other tumors by the presence of three germ folia (3). Russell and Rubinstein et al. (9) described teratomas as tumors that contain ectodermic, mesodermic, and endodermic structures.

Teratomas are typically divided into three groups (mature, immature, and malignant). Mature teratomas contain well-differentiated cells, while immature teratomas contain primitive structures that are not adequately differentiated. Teratomas that contain a malignant component are classified as a malignant teratoma (3,7).

Teratomas commonly occur in the sacrococcygeal region in children. Teratomas may also occur in the retroperitoneal,

anterior mediastinum, and thymus regions, including the pharynx, base of the head, brain, orbit, pelvis and spinal regions, gonads, and subcutaneous tissues (3,8).

Head and neck teratomas are very rare conditions and teratomas occur in these locations in nearly 1%-3.5% of all cases. Head and neck teratomas are usually located anteriorly and laterally; however, there have been a few cases in which the teratoma is located in the occipitocervical region (4,8). We present a case which is very rare in the literature, pathologically diagnosed as an immature teratoma resembling an encephalocele with cervical extension and frontoparieto-occipital location.

The literature has demonstrated that teratomas located on the head and neck are very rare conditions. Mahour et al. (5) reported 1 of 81 patients with teratomas to have an extracranial teratoma. Bale et al. (2) reported that teratomas were located at an extracranial site in 6 of 107 patients with teratomas. In a comprehensive review conducted between 1966 and 2005, 10 teratoma patients were identified who had a midline teratoma on the back of the neck (4,8).

It may be difficult to differentiate teratomas located on the head and neck regions from encephaloceles. Clinical, radiological, and pathology studies can differentiate an encephalocele and a teratoma.

Magnetic resonance imaging could have been helpful for making a definite diagnosis, but it was not possible because of the critical condition of the patient. CT imaging was therefore used.

Fetal pathologies can be diagnosed very easily with intrauterine imaging techniques. These techniques are USG and fetal MR. Even using these imaging methods, definitive diagnosis can be confirmed by surgery.

Our patient was first thought to have an encephalocele. However, during surgery the contents of the mass included hairy and finger-like structures and the mass was covered by tissues that resembled a shell and cystic components, and therefore the encephalocele diagnosis was revised. There were no neural tissues and intracranial extensions inside the mass. There were also no bony defects. In encephaloceles, neural tissues and CSF are herniated inside the sac, together with bony defects. The differential diagnosis of teratomas of the head and neck may include hemangiomas, cystic hygromas, lymphangiomas, lipomas, dermal cysts, vascular malformations, and cutaneous cysts (4,8).

Treatment of teratomas involves early and complete resection because these tumors are prone to grow rapidly, and surgical excision may prevent transformation to a malignant form (10). In our case, an early and complete resection was carried out and no relapses were observed during a 1-year follow-up.

Teratomas on the head and neck regions of the newborn are rare and can be easily confused with an encephalocele. The diagnosis must be supported with clinical, radiological, and pathology studies, and early surgical intervention must be offered. Indeed, complete resection may avoid recurrence of the teratoma.

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