

# Ruptured occipitocervical teratoma mimicking an upper cervical myelomeningocele

## Case report

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✓ Occipitocervical teratoma is an extremely rare condition, and only a few cases have been reported. The authors report on a neonate who harbored a ruptured posterior midline occipitocervical lesion that mimicked an upper cervical myelomeningocele, although the pathological findings were diagnostic of teratoma.

**KEY WORDS** • occipitocervical teratoma • myelomeningocele • pediatric neurosurgery

**T**ERATOMAS contain lines of cells derived from more than one germ layer that are often misplaced in remote areas of the body.<sup>1,4</sup> They often occur in the midline, most commonly in the sacrococcygeal region. Teratomas of the head and neck constitute only 1 to 3.5% of all cases.<sup>7</sup> The majority of neck teratomas are located in the anterior and lateral regions, whereas the occipitocervical location is exceedingly rare.<sup>4</sup> We report on a newborn with a ruptured posterior midline occipitocervical teratoma, although the lesion mimicked an upper cervical myelomeningocele.

### Case Report

*Examination.* This 2-day-old girl born by cesarean delivery was the first child of young, unrelated parents who had an 8-year history of infertility. The mother had taken clomiphene citrate to achieve this pregnancy, which was uneventful. The patient was admitted because of a 7 × 5-cm, nonmobile, ruptured mass that was observed just after birth (Fig. 1). The red-to-purple mass was fleshy, had a watery discharge, and lacked normal skin coverage. The infant had a short, webbed neck and limited neck movement. Motor and sensory examinations were nondiagnostic. Her head circumference was 36 cm, and the fontanelle was wide and bulging. The child otherwise appeared normal.

Because emergency magnetic resonance imaging was not available in our hospital, cervical imaging was not performed at the time of admission. Ultrasonography of the brain confirmed ventriculomegaly without Chiari malformation Type II. An ultrasonographic study of the cervical

mass was not performed because of the absence of skin and the potential risk of infection. Ultrasonography focusing on the kidney revealed unilateral hydronephrosis, which was demonstrated to be Grade III vesicoureteral reflux based on results of voiding cystourethrography that was performed later. The cervical mass could not be evaluated using cervical radiography because of the child's short neck and the shadow caused by her face and shoulders, which obscured the images.

*Operation.* We believed that the lesion was a ruptured cervical myelomeningocele based on the lesion's appearance, its leakage, associated congenital hydrocephalus, and vesicoureteral reflux (a hallmark of neurogenic bladder). On the 2nd day of hospitalization, surgery was performed to prevent meningitis. A horizontal, elliptical skin incision was made to reach the base of the lesion, and a well-encapsulated cervical mass was found that had not penetrated the dura mater. The lesion was located at C-2 and C-3; both levels were bifid and had loose, mobile posterior element remnants around the capsule of the lesion, which were removed. Microscopic evaluation showed mature derivatives of all three germ layers, including skin and its appendages, cartilage, muscle, lung, colon, and brain tissue.

*Postoperative Course.* The postoperative course was uneventful. At 6 months' follow up, there is no recurrence and her ventriculomegaly is stable; thus, there was no need for shunt placement.

### Discussion

The incidence of teratoma is approximately one in



FIG. 1. Photograph showing the ruptured solid mass and the patient's short, webbed neck.

13,000 live births.<sup>1</sup> Although the lesion's appearance in the head and neck is unusual, many have been reported in the lateral and anterior regions. Very few cases of teratoma involve the occipitocervical area. A comprehensive Medline search of articles encompassing the period between 1966 and 2005 revealed fewer than 10 cases of teratoma in the posterior midline area of the neck.<sup>1,2,4-8</sup> Most of them featured a well-formed appendage similar to an accessory lower extremity or an arm. Debate exists as to whether this appendage is a twin who underwent incomplete cleavage, a fetus in fetu, a parasitic fetus, a hamartoma lacking neoplastic elements,<sup>3</sup> or a teratoma. Indeed, the classification of teratomas remains difficult. Overall, a midline lesion having an appendage or firm projections is suggestive of a teratoma.<sup>1</sup> Some posterior cervical teratomas that have no appendage such as the one in a case reported by Naor, et al.,<sup>4</sup> or have a projection such as the one in our case should be differentiated from other masses in this region, including encephalocele, myelomeningocele, hygroma of the neck, lymphangioma, lipoma, and dermal cyst. Clinical, radiological, and histological studies are necessary to establish the correct diagnosis.

The interesting aspect of our case is that by nature of its having a watery discharge and associated abnormalities (that is, vesicoureteral reflux and hydrocephalus) the ruptured mass was presumed to be an upper cervical myelomeningocele. On the other hand, it is odd for myelomeningocele to rupture during a cesarean section in which the delivery was nontraumatic. Therefore, teratoma should have been considered as a differential diagnosis in this case. Magnetic resonance imaging could have been helpful for making a definite diagnosis, but because of the critical condition of the patient, it was not possible.

### Conclusions

Because of the risk of malignant change in this type of tumor, early surgical removal of lesions suspected of being teratomas is important, regardless of the lesion size or the age of the patient (even premature infants should undergo surgery).<sup>4</sup> The necessity of early surgical treatment has been noted particularly in cases of sacrococcygeal teratomas, in which the occurrence of malignancy appears to be related to age. A delay in diagnosis in neonates has been reported to result in a 33% malignancy rate; thus, the higher malignancy rate in older infants is caused by a delay in diagnosis of less apparent lesions.<sup>7</sup>

We therefore recommend that every posterior midline mass having an appendage or odd projection, every mass indicative of myelomeningocele, every myelomeningocele whose rupture seems unreasonable, and all masses suspected of being teratomas undergo early resection.

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