Sacroccygeal teratoma with intradural extension: case report

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Intradural sacroccygeal teratoma (SCT) is a rare entity that has been reported in only a few cases previously. The authors present the case of a 2-week-old, otherwise healthy neonate with a mass in the buttock. The imaging findings and the high level of serum alpha-fetoprotein were highly suggestive of SCT. On operation the authors found intradural extension of the teratoma. The lesion was managed successfully without any remaining sequelae. The authors briefly review the currently proposed etiology regarding teratoma formation and the intradural extension of SCT.

<http://thejns.org/doi/abs/10.3171/2014.10.PEDS1445>

KEY WORDS sacroccygeal teratoma; neonate; intradural; embryology; oncology

TERATOMAS are made up of 3 embryonic layers giving rise to a wide range of tissues.¹¹ Sacroccygeal teratoma (SCT) is the most common type of fetal teratoma, with great diversity in the degrees of maturity and invasiveness.⁷ In most patients with SCT, the tumor is successfully removed along with coccyx. Intradural invasion or extension of SCT is not common, and only a few cases have been reported previously (Table 1). Most patients with intradural teratoma have a pure teratoma without an associated sacroccygeal lesion.^{10,13–16} Permanent neurological deficits can be found in patients with intradural lipoma or intraspinal metastases of malignant SCTs. Sometimes early presentation of these kinds of tumors consists of deficits such as paraplegia and quadriplegia.^{10,13}

We present the intradural extension of SCT in a neonate, briefly review the currently proposed etiology regarding teratoma formation, and discuss the possible embryological basis causing this kind of teratoma.

Case Report

History and Examination

A 2-week-old boy was referred to the neurosurgery department because of swelling in the buttock that had been present since birth. He was the second child of a healthy mother and was born following an uneventful pregnancy

via vaginal delivery. There was no prenatal diagnosis of the lesion. Physical examination revealed a soft-tissue mass in the buttock with a maximum diameter of 7 cm that was covered by intact skin. All neurological examination results were normal. He had regular defecation and urination, with normal anal folds in addition to normal kidney and bladder ultrasound findings. Sacral MRI confirmed a cystic mass of the buttock extending through a widened sacral canal to the S-2 level with anterior displacement of rectum and bladder (Fig. 1). The mass was iso- to hypointense on T1-weighted images and hyperintense on T2-weighted images (Fig. 2). Clinical diagnosis of SCT was considered. Alpha-fetoprotein and beta-human chorionic gonadotropin levels in serum were checked, and alpha-fetoprotein was abnormally high for the patient's age.

Operation

At surgery, the mass was found to be extended from the buttock toward the sacral canal. There was a widely bifid sacrum from S-5 to S-1 into which the stalk of the cystic mass was going intradurally; the mass was noted to be attached to a thick filum (Fig. 3). The capsule of tumor was in continuity with dura mater, through which the tumor entered the sacral canal and attached to filum. The roots inside the sacral canal were freely passing around the in-

ABBREVIATIONS NCC = neural crest cell; SCT = sacroccygeal teratoma.

SUBMITTED February 5, 2014. **ACCEPTED** October 7, 2014.

INCLUDE WHEN CITING Published online January 23, 2015; DOI: 10.3171/2014.10.PEDS1445.

DISCLOSURE The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

TABLE 1. Literature review of SCT cases associated with intradural extension

Authors & Year	Patient Age, Sex	Tumor Type	Region	Deficit	Time of Dx	Explanation
Koen et al., 1998	12 days, F	Mature	Sacroccocygeal	No neuro def, neuro-genic bladder, fecal retention	At birth (no prenatal Dx)	Intradural & extramedullary expansion
Dupin & Sommer, 2012	2 days, M	Immature in 1st op, mature in 2nd op	Sacral	No neuro defs	33 wks of gestation	Intrapelvic & intraspinal expansion, reop for recurrence or residue
Habibi et al., 2007	2 days, F	Mature	Sacral, spinal, pelvic, & abdominal components	Bilat flaccid paralysis of LEs, low motor function of hip	At birth	Intraspinal intradural extension, neurogenic bladder, high levels of α -fetoprotein & β -HCG
Hawryluk et al., 2012	NR, F	Mature	Sacral	No neuro defs	At birth	Extension to spine & sacral roots, wound dehiscence & discharge
Kunisaki et al., 2011	1 yr, M	Mature	Lumbosacral	No neuro defs	1st yr	Colpocephaly, asymmetrical lumbosacral MMC, SCM

Dx = diagnosis; HCG = human chorionic gonadotropin; LE = lower extremity; MMC = myelomeningocele; neuro def = neurological deficit; NR = not reported; SCM = split cord malformation.

tradural extension of the mass without any attachment to the tumor. Dura was opened and en bloc resection of lesion was done after cutting the attachment of the mass to the filum at the S-1 level. Complete closure of dura mater and skin was performed in separate layers. Pathological examination of the mass was diagnostic of mature teratoma (Fig. 4).

Postoperative Course

The postoperative period was unremarkable. No wound problem or CSF leakage occurred postoperatively. One year after surgery the patient's developmental status was within normal limits, with no neurological deficit. Serum alpha-fetoprotein was undetectable and the results of pelvic ultrasound studies were normal.

Discussion

Sacroccocygeal teratoma is the most common teratoma occurring in early life.⁷ Intradural teratoma is less common, but intradural extension or invasion of an SCT is much more infrequent.^{10,14} Tumors with intradural invasion are not associated with a higher probability of malignancy or recurrence.¹⁰ Early presentation of this disease in some patients can be with severe neurological deficits like paraplegia. Therefore, early diagnosis and management of these lesions is important for preventing neurological sequelae.^{10,13}

Although different theories have been proposed, the exact etiology of teratomas is unclear. The classic model of teratoma formation is based on the misplacement of primordial germ cells from the primitive yolk sac.^{5,6,11} Many

FIG. 1. Sagittal T1-weighted MRI studies of the lumbosacral area, confirming an iso- to hypointense mass of the buttock that seems to be extending to the sacral canal.

FIG. 2. Sagittal T2-weighted MRI studies showing a hyperintense mass of the buttock with probable intradural extension; note the cystic mass inside the sacral canal.

FIG. 3. Intraoperative photograph confirming the buttock mass that is in continuity with sacral dura mater and is extended to the sacral canal and attached to a thick filum. Figure is available in color online only.

cases of intradural spinal teratoma associated with dysraphic features such as split cord malformation, tethered cord, spina bifida, and myelomeningocele have been described. Despite the fact that not all spinal teratomas are associated with dysraphism, this association can lead to the dysembryogenic nature of teratoma formation.^{3,5}

It has been suggested that mesenchymal progenitor cells in the caudal cell mass, which are remnants of the primitive streak and Hensen's node, have a pluripotent nature and are able to differentiate into the tissues derived from all 3 germ layers and cause teratoma formation.^{5,6}

Supposing that mesenchymal cells are the cause does not explain the association between teratomas and dysraphisms. Instead, we suggest neural crest cells (NCCs) as the cause of these defects. Neural crest cells develop at the end of the 1st month of pregnancy. During and after folding of the neural tube, NCCs separate from the ectoderm at the boundary between neural and nonneural epithelia and form a mass dorsal to the neural tube, and then they start migration and differentiation to other tissues such as melanocytes, meninges, and sympathetic and dorsal ganglia.^{4,8}

On the other hand, NCCs can easily differentiate to mesenchymal tissue,^{2,12} and detection of mesenchyme does not always mean caudal cell mass. Another important characteristic for teratoma formation is the ability to produce different tissues. It has been demonstrated that NCCs are pluripotent, show stem cell markers, and are able to self-renew.^{2,8,12} It is strongly supported that environmental factors play a crucial role in deciding the final fate of NCCs.^{2,9} As an example, the interplay of Wnt and BMP regulates the migratory and self-renewal capacity of NCCs. As these cells arrive at their targets, sensitivity and response to these factors decreases and the EGF/small Rho GTPase cascade became the main signaling pathway.^{1,2} Whereas Shh is a factor mediating migration through suppression of cell adhesion molecule integrin, RA inhibits NCC migration by stimulating the neural outgrowth, re-

FIG. 4. Photomicrographs showing central neural tissue accompanied by glandular structure with cuboidal epithelium and an area of squamous epithelium associated with normal smooth muscle and vessels found inside the sacrococcygeal mass. H & E, original magnification $\times 100$. Figure is available in color online only.

sulting in specification.⁴ Despite this, it has been shown that even after the completion of the migration, cells on the target location have multiple differentiation options.¹ Therefore, any disturbance in the process of migration can end up with dislocation of these cells from their main target, causing a high potential for inducing teratoma.

Some authors have asserted that the midline position or dorsal origin of teratomas are a proof for germinal or mesenchymal models,^{5,6,11} but both of these properties are satisfied by assuming that NCCs are the responsible cells.

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