Sacrococcygeal Mass in a Newborn: A Quiz

Yannick MUKENDI-NKESU[1,2], Marie-Christine MACHET[3], Émilième ÉDÉE[2], Aurélien BINET[2,4] and Annabel MARUANI[1,2]*
*[1]Department of Dermatology, Unit of Pediatric Dermatology, [2]Reference Center for rare Vascular Skin Diseases (MAGEC-Tours), [3]Department of Pathology, [4]Department of Visceral and Plastic Pediatric Surgery, CHRU Tours, FR-37044 Tours Cedex 9, and [5]Universities of Nantes and Tours, INSERM 1246-SPHERE, Tours, France .*E-mail: annabel.maruani@univ-tours.fr*

A 7-month-old girl, with no notable family history, was referred to our multidisciplinary consultation for vascular anomalies for a congenital sacrococcygeal mass discovered during antenatal life. Ultrasonography at month 4 of pregnancy showed a tissular and cystic sacral mass extending to the pelvis. Magnetic resonance imaging (MRI) performed on day 6 of life revealed a poorly vascularized multi-cystic mass, 30×13 mm, extending from subcutaneous tissue to the pelvis, that suggested a cystic lymphatic malformation. Physical examination showed a completely painless cutaneous protrusion of soft consistency on the intergluteal fold, underlying a fine down. There were no cutaneous lymph vesicles and no angioma stains (Fig. 1A). Physical examination was otherwise normal.

A new MRI performed at 9 months of life, without any contrast enhancement, showed clusters of cystic lesions in the subcutaneous tissues hanging on the coccyx (Fig. 1B). Results were normal for blood count and liver and kidney function, but blood alpha-foetoprotein (AFP) level was slightly increased, 17.8 ng/ml (normally <10.0 ng/ml).

At age 10 months, the entire lesion was surgically removed (Fig. 1C).

What is your diagnosis? See next page for answer.

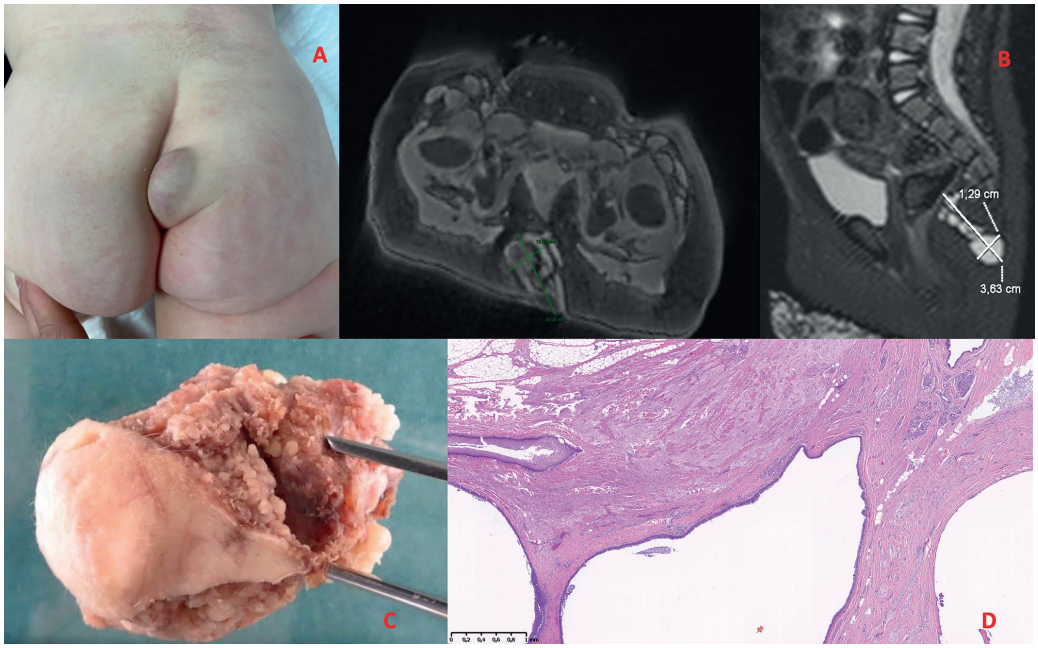

Fig 1. (A) Clinical presentation of a gluteal cutaneous protrusion underlying normal skin in a 7-month-old girl. (B) Magnetic resonance imaging (MRI) at 9 months of life showing multicystic mass attached to the pelvis. (C) Mature teratoma surgically removed. (D) Microscopy (HES) of mature ectopic tissues (glial tissue).

ANSWERS TO QUIZ

Sacrococcygeal Mass in a Newborn: A Commentary

Acta Derm Venereol 2022; 102: adv00782.
DOI: 10.2340/actadv.v102.3934

Diagnosis: **Sacrococcygeal mature teratoma**

Teratomas are rare germ-cell tumours that contain tissues derived from 3 different germ-cell layers of various differentiation staging (1). They have heterogeneous presentations. Teratomas might be located in different sites, the most frequent overall being ovarian (approximatively two-thirds), then intracranial/intraspinal and sacrococcygeal (<10% each) (2). Teratomas occur predominately in children and young adults. Sacrococcygeal teratomas are the most frequent congenital teratomas and can be voluminous and impressive (3).

Treatment of teratomas consists in surgical removal of the entire mass, including the underlying bone if involved. Microscopy shows the tissues involved (glial, vascular, cartilaginous, etc.) underlying the subnormal skin (Fig. 1D) (1, 2, 4). It allows for distinguishing mature from immature teratomas (i.e. teratocarcinoma) according to histological grading, which is important because the latter teratomas can lead to metastasis and a fatal prognosis (4).

Assessment of AFP level might help in the diagnosis. This protein, physiologically produced by the foetus (liver and yolk sac), can be detected in the plasma of neonates, but progressively decreases in level. Its level can be slightly increased in mature teratomas, but when greatly increased, is a marker of teratocarcinoma or other cancers, such as hepatocellular carcinoma (AFP level >100 mg/ml) (4, 5).

The main differential diagnoses of neonatal sacrococcygeal mature teratomas include teratocarcinoma, other malignant tumours, dermoid cysts and cystic lymphatic malformations. Foetal ultrasonography has good diagnostic sensitivity when associated with plasma AFP assessment and MRI (1, 6). Treatment consists of complete surgery with pathology examination.

REFERENCES

1. Lakhoo K. Neonatal teratomas. Early Hum Dev 2010; 86: 643–647.
2. Varma AV, Malpani G, Agrawal P, Malukani K, Dosi S. Clinicopathological spectrum of teratomas: an 8-year retrospective study from a tertiary care institute Indian J Cancer 2017; 54: 576–579.
3. Penny SM. Sacrococcygeal teratoma: a literature review. Radiol Technol 2012; 84: 11–17.
4. Terenziani M, D'Angelo P, Inserra A, Boldrini R, Bisogno G, Babbo GL, et al. Mature and immature teratoma: a report from the second Italian pediatric study Pediatr Blood Cancer 2015; 62: 1202–1208.
5. Guzman G, Alagiozian-Angelova V, Layden-Almer JE, Layden TJ, Testa G, Benedetti E, et al. p53, Ki-67, and serum alpha feto-protein as predictors of hepatocellular carcinoma recurrence in liver transplant patients. Mod Pathol 2005; 18: 1498–1503.
6. Arisoy R, Erdogdu E, Kumru P, Demirci O, Ergin N, Pekin O, et al. Prenatal diagnosis and outcomes of fetal teratomas. J Clin Ultrasound 2016; 44: 118–125.