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A Hairy Congenital Back Mass: Case Report and Literature Review

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Abstract

We present a female neonate born with an unusually located hairy back mass. There were no further anomalies on physical examination. The mass was not discovered on routine prenatal sonography. Complete surgical removal was achieved without complications and histological examination was consistent with mature teratoma.

The differential diagnosis of back masses in the newborn and the classification and work-up of neonatal teratomas are reviewed.

This case suggests that even though most teratomas are located in the sacroccocygeal region, prenatal screening should not overlook the possibility of additional uncommon locations.

Keywords: Neonatal teratoma; Posterior thoracic mass; Congenital anomalies

Abbreviations

SCT: Sacrococcygeal Teratoma

Introduction

Congenital back masses are relatively common in the newborn period and are most commonly located in the sacrococcygeal area in a midline position [1]. These masses may represent a congenital anomaly of the spinal tract or a congenital tumor. When a back mass occurs in a location other than the sacrococcygeal area, especially if it is not in a midline position, it may be overlooked because less attention may be given to areas where masses are less likely to occur.

We present a case with an unusually located back mass and the diagnostic evaluation and subsequent management.

Case Presentation

A female neonate was born at 37 weeks gestation, the second child of a healthy 44 year old woman. Family history was unremarkable and there was no history of exposure to medications or of substance abuse. Pregnancy follow-up was uneventful, with the exception of a growing uterine myoma that required Cesarean delivery. Apgar scores at birth were 9 and 10 at 1 and 5 minutes, respectively. After delivery a posterior mass was noted and the child was transferred to the Neonatal Intensive Care Unit for further evaluation.

Initial physical examination showed a posterior thoracic mass, covered with hair (Figure 1).Vital signs were normal, no additional congenital abnormalities were noted and the neurological exam was normal. Close examination of the mass showed that it was mostly located on the left side of the thorax, but extended medially close to the midline. The mass was of a mixed solid and cystic consistency and did not show any signs of inflammation. Chest X-ray showed a solid mass with ossification at the level of T3-T6. Ultrasound showed a heterogenic mass with solid, cystic and calcified areas and suggested possible invasion of the spinal cavity. Additional imaging including head and abdominal ultrasound scans and a thoracic spine X-ray were normal. Blood chemistry and complete blood count were normal as were β-human chorionic gonadotropin and alpha fetoprotein. A magnetic resonance imaging scan (Figure 2) showed a 2.1 x 3.5 x 1.7 cm mass, located in the

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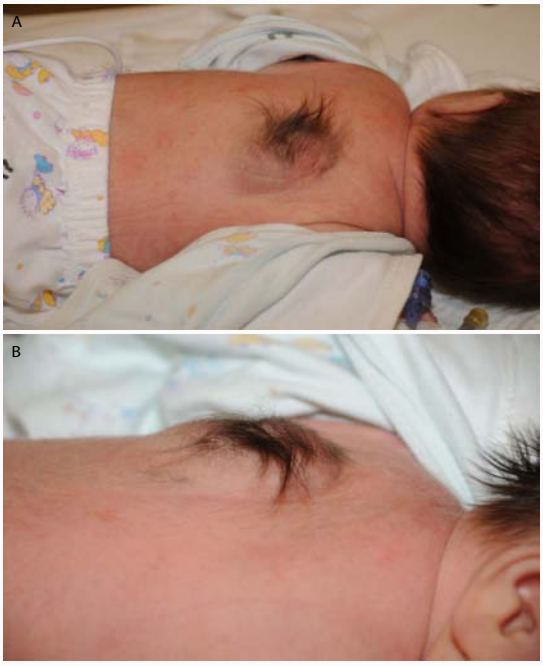

Figure 1: Hairy back mass.

soft tissues of the back with solid and cystic consistency, extensive calcifications and some fat tissue. It did not involve the muscles of the upper back nor did it invade the spinal cavity. The spinal cord had a normal signal and structure. The magnetic resonance scan concluded that the mass was most likely a teratoma.

Surgery excision was performed on day 12 of life with no complications. Histological exam was consistent with a mature teratoma (Figure 3).

The patient was discharged from hospital 4 days after the procedure. Follow-up at 2 months showed normal development and a normal neurological examination.

Discussion

Back masses in the newborn period are commonly located in the sacrococcygeal area. These masses often represent neural tube defects, or a sacrococcygeal teratoma (SCT). However, thoracic back masses occurring in a non-midline position are most likely not related to the spinal tract and have a different differential diagnosis.

The differential diagnosis of chest wall masses includes a wide range of pathologies [1,2]. Midline masses may represent congenital anomalies of the spinal cord. Benign lesions include lipoma, neurofibroma, lymphangioma, tufted angioma, hemangioma, ganglioneuromaand mesenchymal hamartoma. Malignant lesions include neuroblastoma, rhabdomyosarcoma and primitive neuroectodermal tumors. Masses may also represent systemic diseases such as leukemia, lymphoma and Langerhans cell histiocytosis. The main clinical differential diagnoses considered in the case presented were teratoma, congenital melanocytic nevus, meningocele, spinal fusion abnormality with hemangioma or lipoma, plexiform neurofibroma and ganglioneuroma. When evaluating the origin of a congenital back mass its position, consistency, size, color, additional skin findings, evolution with time and imaging findings should all be considered. Imaging is diagnostic for spinal abnormalities and often for teratomas, however for most of the above mentioned lesions there are no diagnostic imaging characteristics. The mass identified in our infant initiated close to the midline and extended laterally, thus a neural tube defect and a teratoma were considered less likely. The consistency of the mass was solid and cystic suggesting the possibility of a teratoma. The color of the mass was somewhat bluish suggesting a vascular component such as a hemangioma. The hair on the mass was compatible with a teratoma or a neurofibroma [3].

Teratomas are a common form of Germ Cell Tumor and as all germ layers are represented, its' cells may differentiate into any body tissue including hair, teeth, fat, skin, muscle and others. Teratomas are the most common tumor identified in the perinatal period, with an incidence of about 1/20,000 live births [3].The estimated incidence of SCT, the most common teratoma, is 1 per 35,000 to 40,000 live births [4,5]. SCT's have a female-to-male ratio of 4:1 [4,5]. Locations where teratomas may occur are described in the Kiel Pediatric Tumor Registry that includes 541 teratoma specimens. The most frequent tumor sites are: the sacrococcygeal region (33.8%), the ovaries (31.2%) and the testes (10.5%). Rare localizations include the mediastinum, the retro peritoneum, the head and neck region as well as the central nervous system [6].

Morbidity and mortality are usually low when the teratoma is mature, diagnosed prenatally and with early surgical treatment [7]. Delay in diagnosis and treatment can lead to complications and increased perinatal mortality. Patients with teratomas can have coincident malformations in up to 18% of cases [8]. Follow up is cardinal because the recurrence rate after complete resection of a mature teratoma can be as high as 10-20% of patients [9].The risk of malignancy is about 10% in neonates and increases with age [9].

Teratomas are usually diagnosed on the basis cystic and solid components identified by ultrasonography or MRI, either prenatally

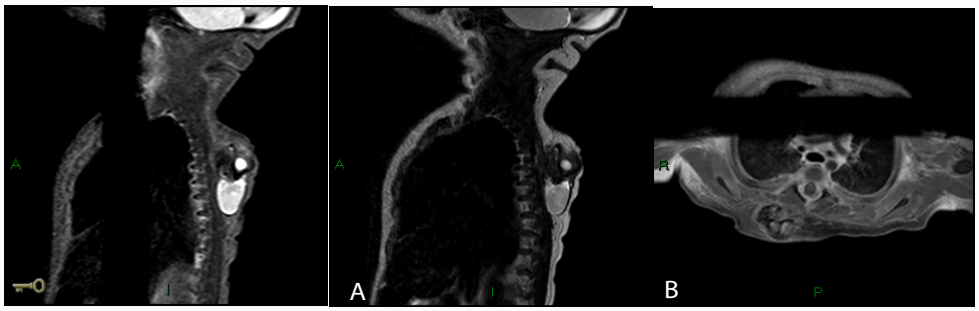

Figure 2: Magnetic resonance imaging of the thorax.
(a) Sagittal T1 WI F. (b) T2 WI of the back showing a complex mass with cystic component and a solid component with fat and calcifications and heterogeneous enhancement following Gadolinium injection.

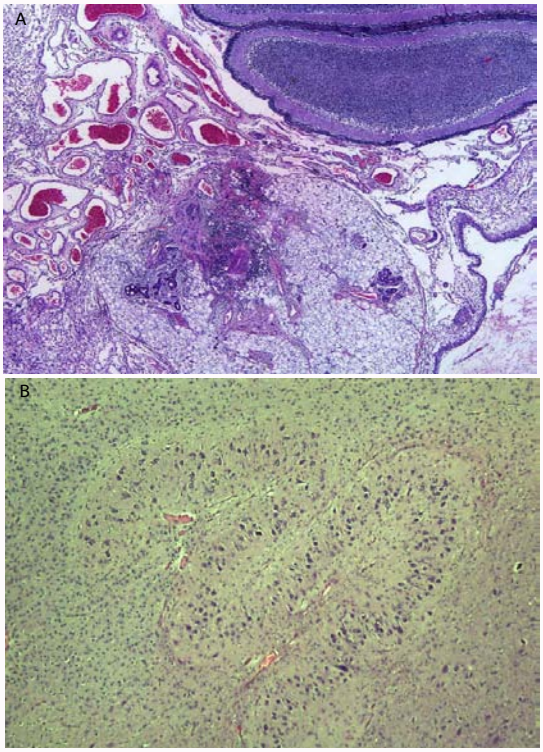

Figure 3: (a) Histology of back mass: the figure shows cerebellar tissue and skin appendages with mature eccrine glands. (b) Glial tissue with basal nucleus-like structure.

[10], often as early as the 1st trimester [10] or postnatally [11]. The present case was preoperatively diagnosed as teratoma according to clinical presentation and imaging studies. The diagnosis was confirmed histologically, and was classified as mature solid and cystic teratoma [12]. The main novelty in the case depicted above is the unusual location of the teratoma. In a summary of 717 cases of teratomas of infancy and childhood by Grosfeld and Billmire [13], there was only one mature teratoma. However, this teratoma was not identified in the neonatal period, was located in the scapular area and not on the back, and was not close to the midline [14]. When a mass compatible with teratoma is identified in utero or after birth in areas not typical of teratoma, this diagnosis should still be taken into account. The hairy nature of the mass should suggest the diagnosis of a teratoma. The potential complications associated with birth of an infant with a large posterior mass were avoided in the present report as a cesarean delivery was performed.

We conclude that while performing prenatal screening, the examiner must not overlook the possibility of a teratoma that is not located in the sacral area. Hairy masses suggest a possible teratoma.

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