Immature malignant sacrococcygeal teratoma: case report and review of the literature

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Summary

Immature malignant sacrococcygeal teratoma (SCT) is a rare tumor, deriving from the three germinal layers and is found in the sacrococcygeal region. It is the most frequent site of teratomas in the fetus. A nut-brown, solid tumor with cystic areas with a ten-cm diameter is reported in the sacrococcygeal region of a female fetus of 23 weeks and with a weight of 308 g. The ultrasound and pathology evaluations revealed characteristics of an immature malignant SCT. The incidence of this tumor type is one in 35,000 to 40,000 live births and females are four times more likely to be affected than males. Sacrococcygeal and cervical teratomas can be diagnosed by prenatal ultrasound and magnetic resonance imaging (MRI). Teratomas are considered an interesting field for research.

Key words: Teratoma; Fetus; Malignant; Sacrococcygeal.

Introduction

Teratomas are tumors that consist of multiple organ and tissue components resembling the derivatives of all three germinal layers of the embryonic disk. In rare cases, not all the three germinal layers can be determined [1]. The components of a teratoma may be quite different from near tissues and may be dissimilar [2].

The most common germ cell tumor diagnosed in neonates and fetuses [3] belongs to a class of tumors known as non-seminomatous germ cell tumors (NSGCT). All these tumors originate from the abnormal development of embryonal and germ cells. The type of pluripotent cell defines the location of the teratoma in the body [4].

Teratomas are observed in many sites at birth, more frequently in the sacrococcygeal and presacral regions and in the neck and are most commonly diagnosed in the fetus [3]. It is also reported that such tumors can be found in the pericardium and oropharynx. They can cause uterine enlargement, polyhydramnios, hydrops fetalis, and tumor mass [5].

There are IV types of sacrococcygeal tumors: a) type I: predominantly external, b) type II: tumors have significant external and intrapelvic components, c) type III: small external component with the majority of the lesions extending intrapelvically and intra-abdominally, d) type IV: tumor occupy the presacral space and have no external component.

Case Report

The authors present a case of a female fetus 23w that was born in the Department of Obstetrics and Gynecology of the University General Hospital of Alexandroupolis and received by the Laboratory of Histology-Embryology of Medical School of Democritus University of Thrace for further examinations. Its weight was 308 g and the measurements of the basic embryonic growth parameters fell within the normal range for the respective age of gestation.

Ultrasound examination showed a large hypervascular exophytic mass with cystic and solid components in the sacrococcygeal region. Gross examination showed a nut-brown solid tumor with cystic areas of maximum diameter of ten cm (Figure 1). Microscopically, the tumor had the histological characteristics of an immature malignant sacrococcygeal teratoma (SCT) and it was constituted of a heterogeneous collection of elements of the three germ cell layers, not arranged in organoid fashion. This variant included islands of poorly-formed cartilage, loose mesenchyme (muscle bundles), clusters of squamous epithelium, neuroblasts (neural tissue, brain substance), structures reminiscent of thyroid gland, bronchial epithelium, and bits of intestinal wall, all embedded within a fibrous gray-white solid matrix (Figures 2 and 3). All the elements were undifferentiated.

Furthermore, the molecular detection for human parvovirus B19-possible factor for hydrops fetalis by polymerase chain reaction (PCR) was negative (Figure 4). The method was performed using the PCR kit Parvovirus B19 according to the manufacturer's instructions.

Interestingly, this stillborn derived from a twin pregnancy of which the other embryo survived.

Discussion

In the present article, the authors report a case of an immature malignant SCT. In general, a mature teratoma is benign and is usually found in females, while an immature teratoma is typically malignant and is commonly found in males. Although, it is reported that in some cases, tumor immaturity does not appear to be a measurement for malignancy in congenital teratomas [5].

Factors usually associated with benign lesions include: early (neonatal) presentation, female patient, cystic composition, and the presence of large areas of calcification or ossification. Factors associated with malignant lesions in-

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Fig. 1

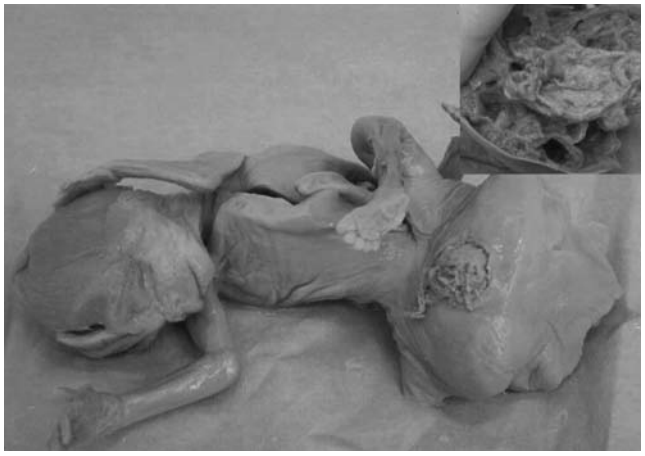

Fig. 2

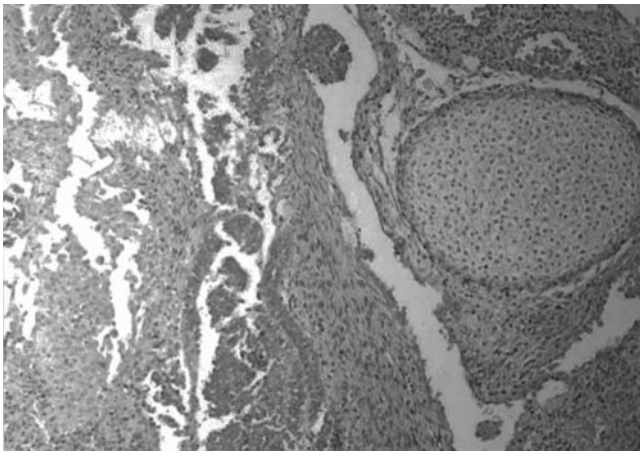

Fig. 3

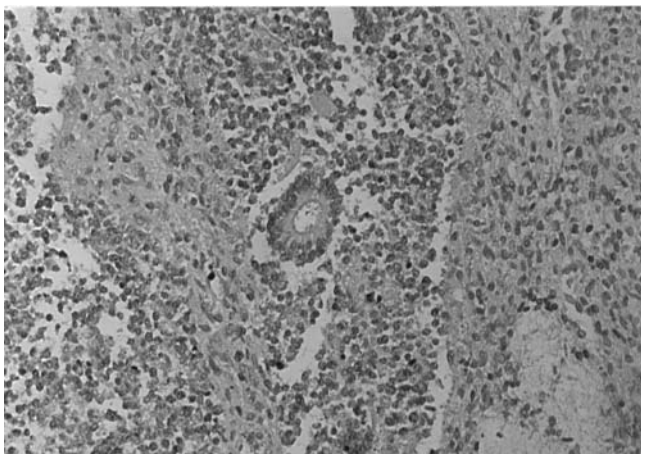

Fig. 4

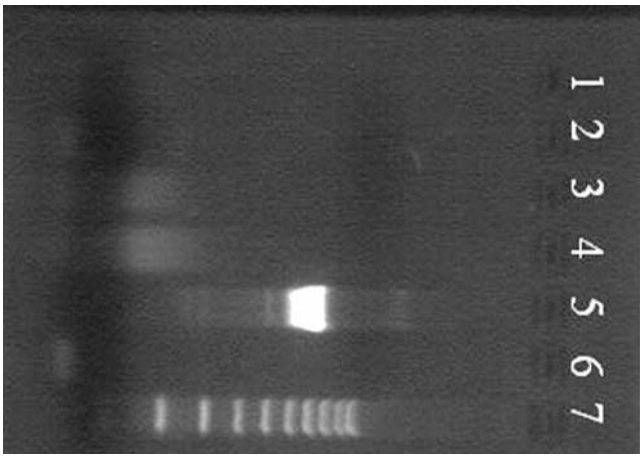

Figure 1. — Gross anatomy of tumor and cut surface with solid and cystic areas (inset).
Figure 2. — Tissue section (H&E X200) showing immature malignant SCT. Islands of immature cartilage and glandular structures are distributed, through a poorly-differentiated stroma.
Figure 3. — Tissue section (H&E X200) showing immature malignant SCT, illustrating predominantly neuroectodermal tissue in the form of rosettes, tubules, and cellular glial foci.
Figure 4. — Molecular detection of human parvovirus B19 in different tissues of the embryo. Only positive control of PCR amplification is visible in line 5. Lines 1, 2, 3, and 4: negative samples, line 6: negative control, line 7: MW marker.

clude: clinical presentation beyond infancy, male patient, presacral location, solid composition (particularly with areas of hemorrhage or necrosis), and lack of calcification.

A clinicopathologic study of 22 fetal and neonatal tumors by Heerema-Mc Kenney *et al.*, reports that in the majority, congenital immature teratomas that require in utero intervention present small foci of conventional yolk sac tumors and are more commonly immature at variance to those that are dissected postnatally. Furthermore, they may present hepatic differentiation and immature endodermal glands, as it is known that teratomas are composed of recognizable tissues of ectodermal, mesodermal, and endodermal origin, in any combination [5].

Germ cell line teratomas are usually present in adult men and women, fetuses, and children. The embryonal ones are most often found in fetuses, in babies at birth, and in young children. Teratomas may also be found in non-humans as reported by Catone *et al.* [6].

SCT is diagnosed in one out of 40,000 fetuses. According to the current world population birthrate, this means 1,800 per year. Adding to these, the number of teratomas diagnosed in other locations of the body and SCT that are diagnosed later in life increase the incidence rate to 10,000 cases per year [7].

An immature teratoma that is benign has a much higher risk to become malignant and requires closer follow-up [8]. These cases may be difficult to correctly diagnose. As sacrococcygeal and cervical teratomas spread from the fetal body into the surrounding amniotic fluid, they can be detected by prenatal ultrasound examination. Additional diagnostic methods may include prenatal magnetic resonance imaging (MRI) of the uterus which is more informative for those teratomas within the fetal body that are less clearly seen with ultrasound [9]. Tumors, however, affecting fetuses and newborns differ from those found in older children and adults, which means that the therapeutic and diagnostic challenges are different, including surgical resection and chemotherapy.

Generally, teratomas comprise the most common fetal tumors and cause high morbidity because they can also develop hydrops fetalis or premature delivery [5]. In the case of the present reported embryo, the PCR amplification for human parvovirus B19, which is a common agent causing hydrops fetalis [10], was negative.

In association with other reported cases, the authors agree that teratomas should be specifically assessed, aiming for a prenatal prognosis, diagnosis, and treatment that increases the chance for survive of the infected embryos.

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