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Gigantic Immature Sacrococcygeal Teratoma in an Abortus

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ABSTRACT. An autopsy case of giant immature sacroccygeal teratoma seen in an abortus is presented here. It was unusual for such a teratoma to weigh more than the fetus. Histology of the tumor was characterized by a large amount of immature and neuroepithelial elements in addition to other endodermal and mesodermal elements. Although α-fetoprotein was high in the amniotic fluid, no germ cells were identified. No invasion or metastasis was noted. We considered the bad prognosis in fetal cases may be due not to the amount of immature tissue but rather to the obstetric complication resulting from the large tumor size.

Key words : immature — sacrococcygeal teratoma — α-fetoprotein — hydramnios — abortus

Teratomas are tumors of three germ layers, and account for 3.1% of both benign and malignant tumors encountered in children[1]. In infants, the sacrococcygeal area seems to be the most common location for this type of tumor. Sacrococcygeal teratomas occur in one out of 40,000 births[2]. Approximately, three fourths of the patients are girls[3]. Over 90% of the patients are the product of full-term gestation[3]. Waldhausen[4] reported that only 7% of 56 teratomas excised from patients less than 4 months of age were malignant, but the incidence was 42% of 31 tumors in children from 4 months to 5 years of age. The criteria of malignancy varied among authors. Many factors seem to affect the prognosis.

Recently, we experienced a large sacrococcygeal teratoma in a female abortus of 21-weeks gestation. The tumor was larger than the weight of the abortus. Giant size over 20% of total infant weight seems exceedingly rare, and, in fact, a fetal teratoma reaching the weight of the fetus has not been reported to date[5]. Histologically, immature neuroepithelial tissue predominated. Distant metastases were not identified. Although α-fetoprotein in the amniotic fluid was elevated, extensive histological examination failed to reveal germ cell elements. We had some problem in determining whether or not this tumor was malignant or benign. It is generally accepted that immature neuroepithelial components in ovarian teratomas are an indicator of malignancy[6,7]. In reviews of sacrococcygeal teratomas, however, no clear-cut histological criteria of malignancy of sacrococcygeal teratomas have been entertained. The term "malignant"

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has been utilized too liberally in the past to even describe sacrococcygeal teratomas with foci of immature tissue. Thousands of sacrococcygeal teratoma have been recorded, but the prognostic implication of immature tissue in the tumor remains to be elucidated.

For these reasons, we present here a unique case of a giant immature sacrococcygeal teratoma seen in an abortus and review and discuss prognostic factors of such tumors.

CASE REPORT

A 30 year-old woman, gravida 1, para 1, was admitted at 21 weeks of gestation because of suspected hydramnios. Her past history was unremarkable. Ultrasonography revealed an abnormal mass at the caudal end of the fetal trunk, which was generally solid with bizarre internal echographic appearance. The rest of the mass appeared multicystic. On the 10th day after admission, amniocentesis with amniofetography was performed and 150 ml of cloudy and blood-tinged amniotic fluid was withdrawn. α-Fetoprotein in the amniotic fluid was 7.7×10^6 ng/ml. The simultaneously examined α-fetoprotein level in the maternal serum was 3.5×10^3 ng/ml. The next day, she spontaneously delivered a female abortus.

PATHOLOGICAL FINDINGS

An autopsy was done one hour after death. The abortus, excluding the mass, weighed 320 g and had a crown-heel length of 20.5 cm (Fig. 1). The mass, which was located in the post sacral area, was well-circumscribed and clearly separated from the sacral bone by thin connective tissue. The tumor itself measured $12 \times 14 \times 4.5$ cm and weighed 380 g (Fig. 1). The outer surface was almost entirely covered by smooth and translucent membranous tissue which was stretched skin extending from the buttock. Other portions were covered by friable necrotic and hemorrhagic tissue. It was generally soft to resilient in consistency. Overall, cut surfaces (Fig. 2) were solid and yellow-tan in color. Mucinous and cystic areas, measuring up to 0.2-2.0 cm in diameter, were scattered throughout. Some of the cysts contained mucinous fluid, and others had serous fluid. The spinal cord and intrapelvic space were intact. Except for this mass, no other gross external anomalies were identified.

Histologically, the tumor was composed of tissue of all three germ layers with a preponderance of neuroectodermal derivatives. Ectodermal derivatives were of a primitive neural tube-like structure (Fig. 3 & 4) and squamoid nest (Fig. 5). Some of the squamoid nests were reminiscent of an optic cup with melanin pigments (Fig. 6). Occasionally observed were cuboidal epithelium with papillary projections into the cystic cavity similar to a choroid plexus although capillaries in the papillary stroma were not prominent (Fig. 7). Occasional clusters of mature ganglion cells were seen in the immature neural tissue. Endodermal elements included intestinal epithelium with mucin production

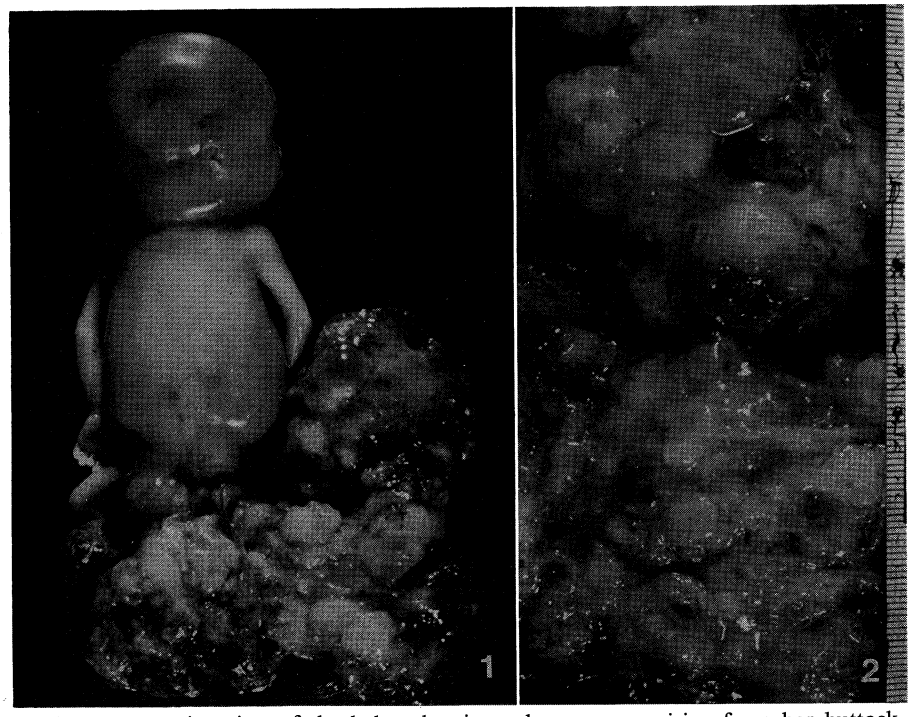
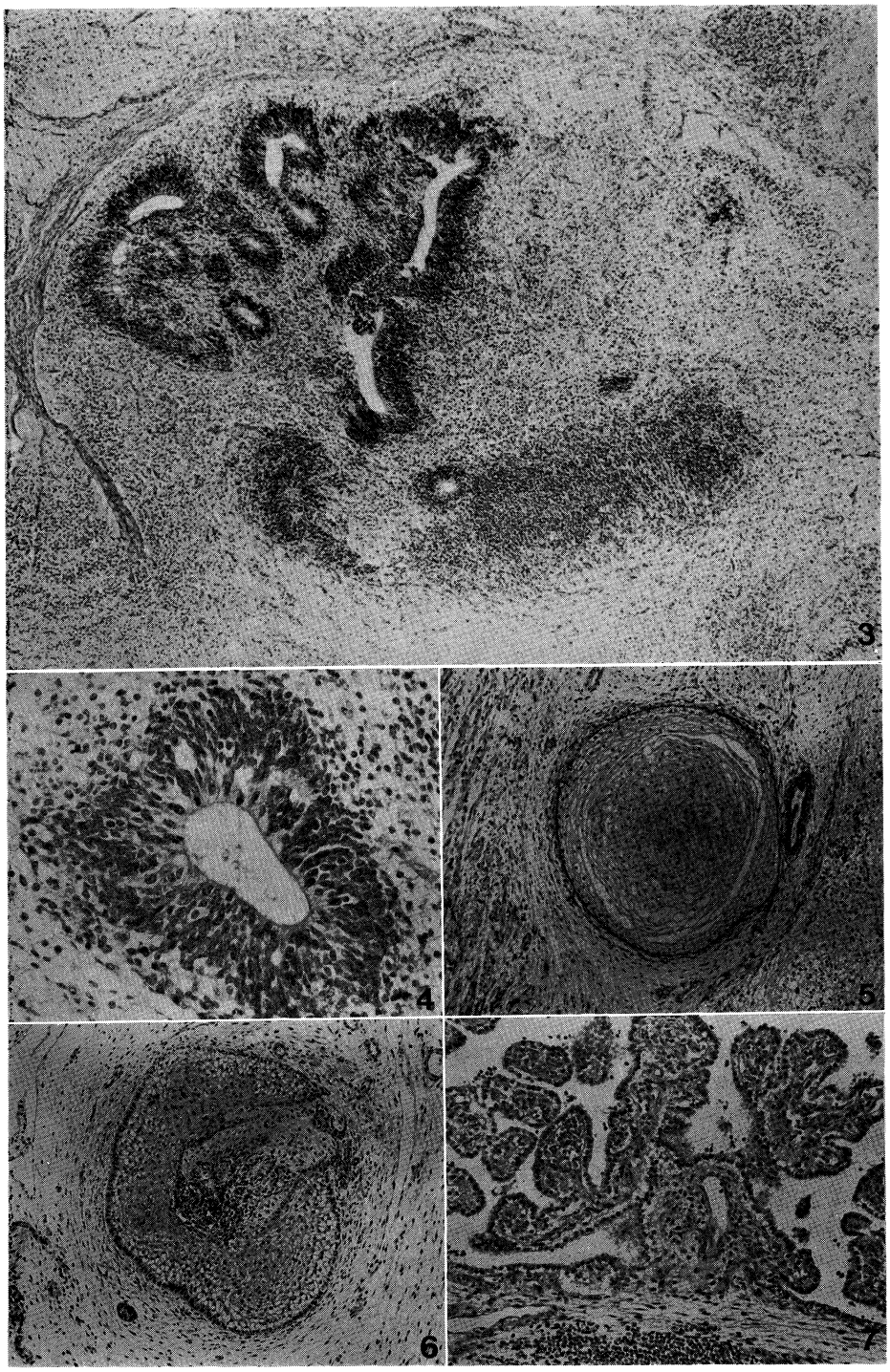


Fig. 1. Posterior view of the baby showing a large mass arising from her buttock.
Fig. 2. Cut surface of the tumor shows gray tan, solid areas. Cystic spaces are also present.

(Fig. 8). Tissue of mesodermal origin was represented by small foci of cartilage, bone (Fig. 9) and smooth and skeletal muscles (Fig. 10). Nowhere in the tissue examined among numerous sections were there germ cell tumor components. With the peroxidase-antiperoxidase technique for α-fetoprotein detection we did not identify any source of α-fetoprotein in the tumor tissue.

DISCUSSION

The presence of a mass in the sacrum is the major sign of a sacrococcygeal teratoma. Growth may be gigantic in comparison to a small baby. To our knowledge, the largest tumor reported so far was that of Salaymeh[5]. In his case, the tumor, which was attached to a full-term baby weighing 4,200 g, weighed 1,700 g and included 1,500 ml of serous fluid. It is known that the weight of such a tumor may reach or surpass half the weight of the baby[3,5]. No reports have described a tumor weighing more than the baby as in our case. Large tumors may result in difficult births and obstetrical accidents. Hydramnios has sometimes been associated with this tumor[8]. Protein sequestration by the tumor, disproportionate increase in the vascular territory and loss of surface



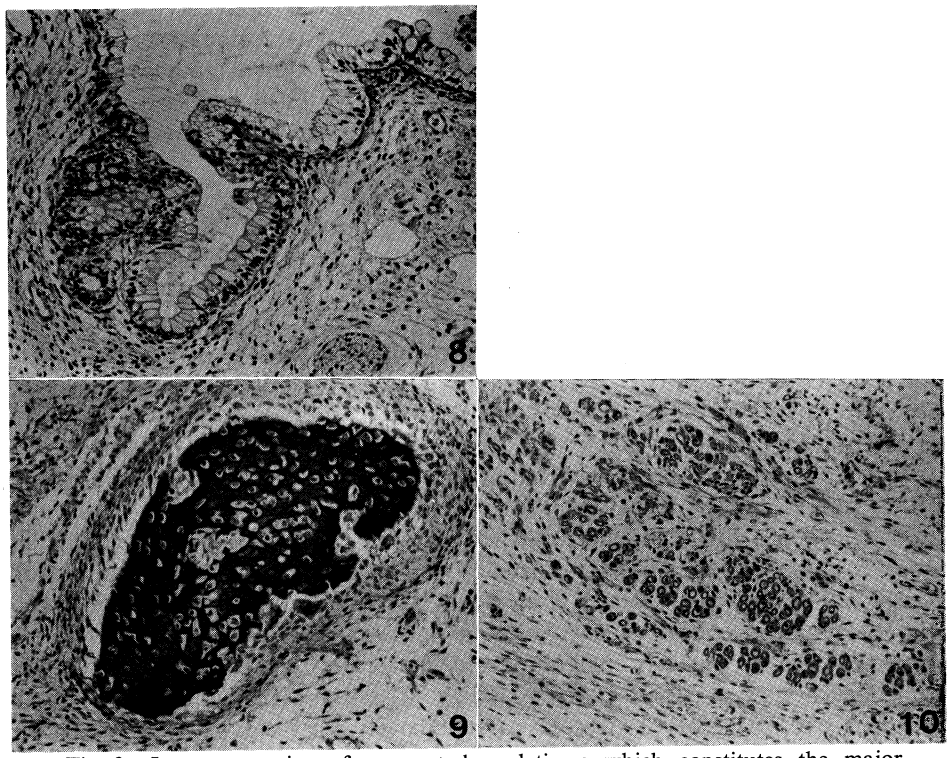


Fig. 3. Low power view of neuroectodermal tissue which constitutes the major component of this tumor. (H-E, ×100)
Fig. 4. A primitive neural tube-like structure. (H-E, ×400)
Fig. 5. A nest of well differentiated stratified squamous epithelium. (H-E, ×250)
Fig. 6. Cup-shaped structure of squamous epithelium with melanin pigments suggestive of differentiation toward an optic cup. (H-E, ×250)
Fig. 7. Papillary proliferation of cuboidal epithelia, with reminiscent of a choroid plexus. (H-E, ×300)
Fig. 8. Columnar epithelium with mucin production. This appears to represent differentiation toward intestine. (H-E, ×300)
Fig. 9. Bone formation. (H-E, ×300)
Fig. 10. Muscle fibers mimicking myotube, which are characteristic of primitive skeletal muscles, are seen. (H-E, ×250)

covering were speculated to be responsible for the hydramnios. Compression effects on the anus may also have been responsible in our case.

As to the benign or malignant nature of sacrococcygeal teratomas, no clear-cut criteria have been presented to date. According to the literature, the size of the tumor seems to have no bearing on prognosis[3,9], yet there are not enough neonatal cases with a large teratoma to conclude so. In our case, it is certain that the large size of the tumor was somehow responsible for the adverse result.

Age has a rather dramatic influence on the prognosis, and the progressively increasing incidence of malignancy in sacrococcygeal teratomas excised at later periods after birth has been repeatedly emphasized. Donnellan and Swenson[10] showed that 10% of the tumors discovered between birth and two months of age were malignant, while of teratomas discovered after two months of age, 90% were malignant. In their study, malignancy was defined as the presence of actual invasion and/or metastases and not solely as the presence of histologically immature tissue. Anatomically, the American Academy of Pediatrics divided the cases into four groups[3], namely : Type I, tumors predominantly external with only a minimal presacral component ; Type II, tumors existing externally but with a significant intrapelvic extension ; Type III, tumors apparently existing externally with extension into the pelvis and abdomen, and Type IV, presacral with no external presentation. Although malignant cases were more commonly observed in type III cases, some of the other types also had metastases. Our case belonged to Type I and had no metastasis. This classification system does not correlate the clinical course with the histology of the tumor. Gonzalez-Crussi et al.[11] examined sacrococcyeal teratomas histologically and divided them into four grades according to the amount of immature or embryonic tissue, especially neuroepithelial, and the association of germ cell carcinomas. It was concluded that a high rate of recurrence and early development of metastasis may be anticipated when foci of yolk sac carcinoma are discovered in the tumor and furthermore, that immature somatic tissue does not necessarily signify a malignant prognosis, though a few were associated with metastasis. Results from Japanese cases are in good agreement with their study except for the presence of one stillborn case[12]. It should be noted, however, that with an increase of immature tissue in the tumor, the incidence of germ cell tumor component increases. Therefore, it seems prudent to carefully monitor all patients whose teratomas contain abundant amounts of immature or incompletely differentiated tissues.

Recently, the determination of α-fetoprotein in the serum of patients with a sacrococcygeal teratoma has proved to be the laboratory test that most commonly correlates with malignancy[10]. α-Fetoprotein can also be detected in the amniotic fluid obtained by amniocentesis. The elevation of α-fetoprotein is known in cases of yolk sac carcinoma. It is also known that sacrococcygeal teratomas with elevation of serum α-fetoprotein often contain areas of embryonal carcinoma in the same tumor mass[10]. In our case, therefore, there might have been small foci of germ cell carcinomas including yolk sac tumors, which were not detected by the examination of multiple randomly taken sections. In addition, it has been proposed that immature and intestinal tissue in the teratoma may produce α-fetoprotein[13]. In the present case, however, α-fetoprotein was not detected in these immature tissues by the peroxidase-antiperoxidase technique.

Our patient with a giant immature sacrococcygeal teratoma had no invasion or metastasis, but was aborted because of the large tumor size and associated hydramnios. We considered that the bad prognostic implication in fetus may

be attained not by the amount of immature tissue itself but rather by the large tumor size.

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