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Hypertrophy of the Placenta and Sacrococcygeal Teratoma

Report of Two Cases

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Summary. This report describes two pregnancies complicated by hydramnios, fetal death and sacrococcygeal teratoma in the second trimester. In both cases the placenta was remarkably enlarged. It is postulated that the tumors behaved in the manner of A–V fistulas and produced forward failure in the fetus similar to what is occasionally seen with large chorangiomas.

Key words: Hypertrophy of placenta – Sacrococcygeal teratoma – Intrauterine fetal death.

Introduction

There are several conditions causing the placental enlargement, however, its precise pathogenesis is still unsettled [1, 4, 7, 8]. We encountered two cases of huge placenta with the fetus having a large sacrococcygeal teratoma. This report describes the autopsy findings of these cases and discusses the pathogenesis of the placental enlargement.

Report of Cases

The mother of *case 1* was a 29-year-old Japanese gravida 5, para 3. She had 3 normal children, 1 artificial miscarriage and 1 stillbirth. Her prenatal course during this pregnancy was normal until the sixth month. At that time she developed rapid increase in body weight and abdominal circumference and began to have abdominal pains. A twin pregnancy was suspected, however, the ultrasonography demonstrated a large singleton placenta and presence of hydramnios. A fetal anomaly was suspected. On admission fetal heart sounds could not be heard. She was delivered of a 2,040 g stillborn female infant after 28 weeks gestation using the drip infusion of Atonin and treatment with metreurynter. The laboratory test for syphilis was negative and the maternal blood type was A and Rh(+). Blood pressure was within normal range (130/70–138/88 mmHg).

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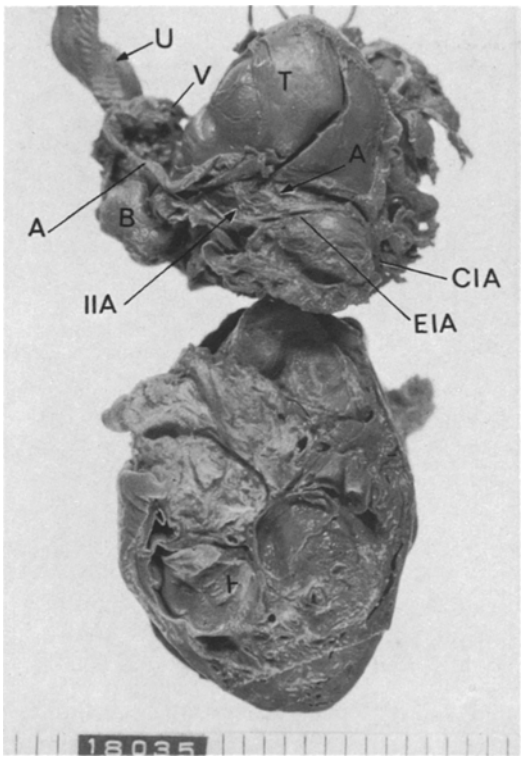

Fig. 1. Teratoma of case 1. Upper half of this figure shows intracecal teratoma (*T*), urinary bladder (*B*), left umbilical artery (*A*), cut end of umbilical vein (*V*), umbilical cord (*U*), left external iliac artery (*EIA*), branches of internal iliac artery (*IIA*) and left common iliac artery (*CIA*). Lower half shows sagittal cut surface of the external teratoma

The infant showed marked maceration and a tumor of over fist size ,was observed on her sacral area. Amniotic fluid was turbid with chocolate color and measured 1,600 ml. Autopsy was performed on this infant about 4 days after the disappearance of heart sounds.

At autopsy, maceration was conspicuous. The sacrococcygeal tumor measured $12 \times 12 \times 8$ cm and partially occupied the pelvic cavity. A large portion of the tumor was situated extracecally. The external genital organ and anus were observed on the frontal surface of the tumor. Sigmoid colon and rectum were embedded in the tumor and lower portion of abdominal aorta, a part of iliac arteries and both umbilical arteries were severely compressed by the tumor. The umbilical vein was dilated. Topographical relation between the tumor and umbilical vessels is shown in Fig. 1. There were hypoplasia of the left kidney and a patent foramen ovale and ductus arteriosus. Both lungs were atelectatic. The other visceral organs, including spleen and liver showed no remarkable changes. The following weights were obtained at the autopsy, with normal weights appearing in the parenthesis: heart 12 g (9.8); lungs 9.5 g (29); liver 75 g (47); kidneys 5.5 g (13); adrenals 2.0 g (4.1); spleen 4.5 g (3.8).

Microscopically, the tumor was a teratoma. Fairly well differentiated elements of all germ layers, including gastrointestinal mucosa, columnar epithelium, smooth muscle, striated muscle, cartilage, bone with marrow, fat, neuroglial tissue and nerve bundles were present. Malignant cells were not observed. The visceral organs, e.g., liver, spleen, pancreas, kidneys, adrenals and gastro-intestinal tracts showed marked postmortem changes. Several patchy calcified lesions were found in the liver and left hypoplastic kidney.

The placenta weighed 1,680 g. The membranes were complete, but partially lacerated. The fetal surface was smooth and gray in color (Fig. 2). The maternal surface was intact, pale yellow gray, normally fissured and fairly friable. The

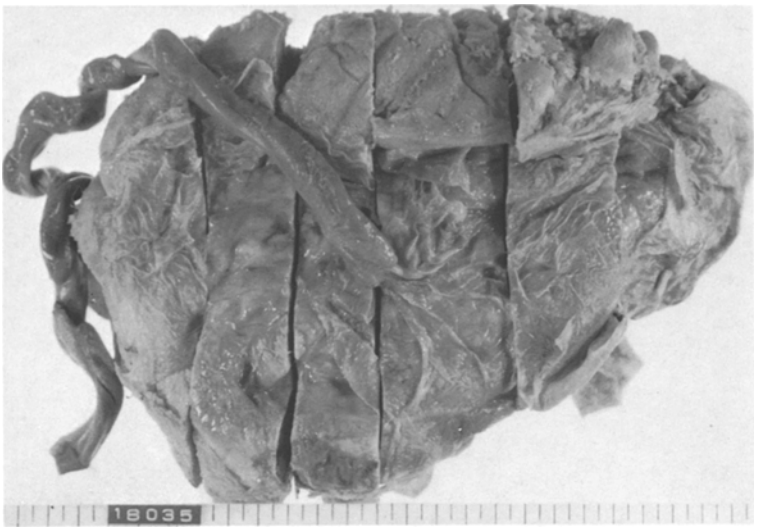

Fig. 2. Huge placenta and umbilical cord of case 1. Fetal surface is smooth and gray in color and partially lacerated

Fig. 3. Umbilical cord of case 1. Umbilical cord, 15 cm from umbilicus, shows dilatation of umbilical vein. ×7

umbilical cord showed no remarkable changes, but the umbilical vein was dilated (Fig. 3). The placenta cut with usual resistance and the cut surface was pale reddish-yellow and partly yellowish-white with firm consistency. No definite thrombosis or necrotic foci were observed.

Histologic examination showed normal configuration of the chorionic villi but they were edematous and rich in vasculature (Fig. 4). Congestion of vessels,

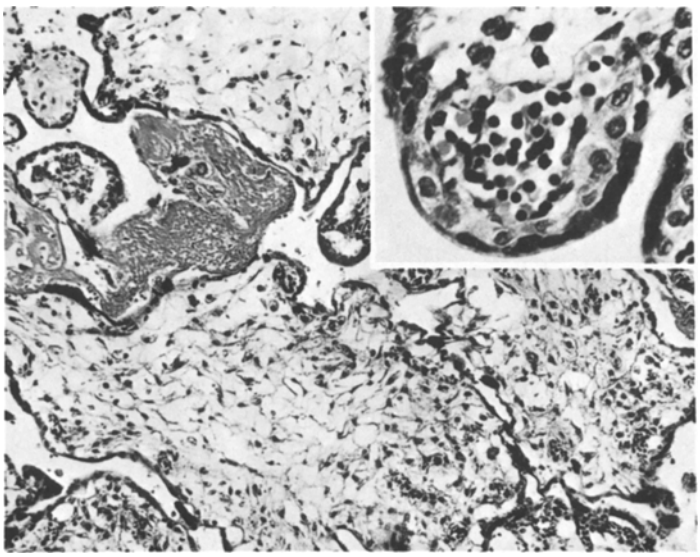

Fig. 4. Chorionic villi with edema and congestion in case 1. Edema and congestion are observed in the villi. ×100. Insert: Nucleated erythrocytes are found in the capillaries. The Langhans' layer is fairly well preserved in this area. ×200

containing a large number of nucleated erythrocytes (Inset in Fig. 4) both in the capillaries and larger vascular spaces was noted. The Langhans's layer was fairly well preserved (Inset in Fig. 4) and a normal number of Hofbauer cells were also present. Syncytial cells were normal in size. Fine granular calcium deposition was seen at the peripheral area of the villi. Fibrin deposition was found scattered in the intervillous spaces. The decidual cells show no remarkable changes.

The mother of *case 2* was a 25-year-old, white primigravida who was first seen at 22 weeks with sudden onset of massive and generalized edema, abdominal pain and systolic hypertension (140–170 mmHg). She had gained 8,000 g in the preceding two weeks, had 1+ proteinuria and normal BUN and creatinine. No abnormality of renal function was detected. The edema disappeared with bed rest. The uterus was of term size and fetal heart tones were normal. Ultrasonography revealed an extremely large anterior placenta, a hydropic fetus and an additional large intrauterine mass. Two days after admission the heart sounds ceased and a macerated fetus was delivered, the mass having detached during delivery.

The female fetus weighed 500 g, measured 15 cm crown-to-rump and was severely edematous and macerated. A 320 g ovoid, macerated teratoma from the sacral area was separated (Fig. 5). The placenta weighed 1,000 g. Upon dissection, the abdominal and pleural cavities contained large quantities of fluid. The following weights were obtained after fixation, with normal weights appearing in parenthesis: heart 4.2 g (4.6); lungs 3.8 g (15); liver 25 g (31); kidneys 2.4 g (5.3); adrenals 1.3 g (2.5). The lungs were markedly hypoplastic, perhaps secondary to the large quantity of pleural fluid. There was a 7 cm defect in the sacral area where the tumor had detached. No excessive amounts of extramedullary hematopoiesis were present in the organs on histologic study. The uterus and ovaries were normal. The lungs contained inspissated debris in its lumina, apparently aspirated dead cells. Myocardial fibers showed clear evidence of hypertrophy. The definitive cortex of the adrenal was much more developed than is normal at this fetal age. The teratoma was

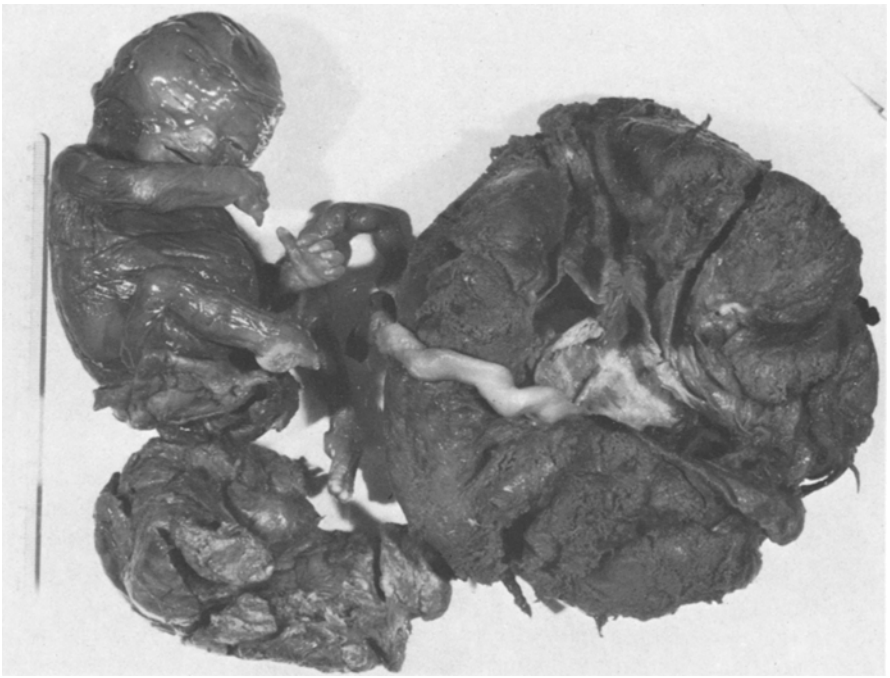

Fig. 5. Macerated fetus of case 2 with detached sacrococcygeal teratoma below and huge placenta at right

predominantly composed of neural tissue but also contained cartilage and cysts of epithelial nature, including pigmentation of possible retinal origin.

The placenta and cord had no inflammatory lesions. The villi were remarkably enlarged and corresponded to those of *case 1*. Here also, an excessive number of nucleated red blood cells were found. Erythroblastosis was not formally ruled out, lacking blood grouping studies; however, the small size of the liver and diminutive spleen argue strongly against this possibility.

Discussion

These cases of large placenta and intrauterine fetal death with sacrococcygeal teratoma have several features which prompt a brief review of the pertinent literature. No report has apparently been published of the association of large placentas in cases of sacral teratomatous malformations.

The fetal-placental weight ratio at the end of pregnancy is normally about 7:1 [7], while it was 1.2:1 in *case 1* and 0.8:1 in *case 2*. Hypertrophy of the placenta is primarily associated with erythroblastosis fetalis, fetal cardiac decompensation and occasionally it is seen with syphilis, diabetes mellitus and hormonal changes [1, 5, 7]. Characteristic pathological changes may be found

in the placenta in cases of syphilis and diabetes mellitus but they were lacking in our cases.

Erythroblastosis fetalis is the most important condition correlated with extremely large placenta. The placental enlargement is due to cardiac failure secondary to deficient oxygen supply which results from hemolysis. As to the maternal factor, in 90% of the mothers, erythroblastotic children are Rh(−) [7]. The mother of the present *case 1* was Rh(+) and no definite hepatosplenomegaly was observed in either infant, thus precluding erythroblastosis. In cases of erythroblastosis fetalis, the most characteristic features of the placenta are the persistence of the Langhans' layer of the villi, hematopoiesis, edema and pigment [2, 5, 7, 10]. Almost all the Langhans' cells of normal chorionic villi are thought to disappear for light microscopy by the sixth or seventh month, but they persist until term as flattened cells and are best observed electronmicroscopically [10]. In certain pathologic conditions, such as erythroblastosis and diabetes, these cells become much more evident [7].

In our cases, Langhans' cells were fairly well preserved but Hofbauer cells were not so prominent as found in erythroblastosis. Extramedullary hematopoiesis, though the maceration was marked, was seen scattered in the kidney, liver and placenta, but the degree was not excessive. Moreover, there was no pigment in spleen or chorion.

One possible explanation for the pathogenesis of the placental enlargement in *case 1* is a fetal circulatory disturbance due to the compression of umbilical vessels. The dilation of umbilical vein in *case 1,* the edema and a large number of erythroblasts in the capillaries and vascular spaces in the villi support this speculation. Under this circumstance, the fetal heart will be overloaded resulting in the dilation of umbilical vein followed by congestion and edema of chorionic villi. Then the placenta is thought to become larger possibly to keep up sufficient function.

Another hypothesis relates to the presence of the large teratoma. In such a condition, compared with a normal single pregnancy, much more blood supply would be required by the fetus and also by the large tumor. The teratomatous mass would take away so much circulation that the fetus and placenta were not getting enough and were hypoxic. The fetal heart could not keep up with this high demand and placental edema ensued. This second hypothesis, high output failure, is supported by *case 2* in which no vascular compression was demonstrated. All of the tumor was external, yet the pathologic changes in the placenta and clinical presentation were nearly identical. A similar situation pertaines in large chorangiomas [3, 4]. Particularly hydramnios, but also large placentas have been observed with large chorangiomas and the most likely explanation for this association again is high output failure. In any case, high output failure leads to hypoxia and essentially to a similar placental pathology as the villi would have in erythroblastosis fetalis and chronic fetal bleeding or, even better, in alpha-thalassemia.

At one time during our observations, we speculated that possibly the neural elements of the tumor may secrete vasoactive hormones to affect placental growth but find no precise evidence for this. Various other causes of placental enlargement and hydrops exist. Thus, placental neuroblastoma metastases, ade-

nomatoid malformation of the lungs, alpha-thalassemia, fetal cardiac anomalies, chronic fetal-maternal transfusion, storage diseases and a few other very rare conditions have been described as causes [9]. None of these conditions were identified in the present cases of placental enlargement.

Sacrococcygeal teratomas are now recognized as being of mitotic rather than postmeiotic origin as are ovarian teratomas [6]. They are more akin to monozygotic twinning events but why some such tumors are more organized than others, for instance making limbs, while others are typical tumorous masses as the present cases is unknown. All cytogenetic studies have shown the teratomas to be diploid. Regrettably, in these cases, the degree of maceration prevented cytogenetic study.

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