Teratoma is a very rare tumor of the placenta. The importance of placental teratoma is that it should be distinguished from fetus acardius amorphous. The presence of an umbilical cord and the existence of a degree of skeletal organization are claimed to be the main criteria for distinction. However, in certain instances it is difficult to separate the two entities. We encountered such a case and reviewed the literature for similar cases. The ongoing controversy over the subject is highlighted.

CASE
A 40-year-old (gravida 4, para 4+15) woman presented with vaginal bleeding at 7 weeks of gestation. The ultrasound examination showed a single viable fetus and a subchorionic hematoma. Regular follow-up at different stages of pregnancy (12, 18, 27 and 36 weeks) showed normal development. The subchorionic hematoma disappeared. A review of ultrasound findings was not possible since no records were available. We felt that the mass could be misinterpreted by the ultrasonographer as part of the viable fetus, or not observed from the beginning. The patient was started on insulin in view of the high blood sugar levels. At 38 weeks, labor was induced and a baby girl was delivered by normal vaginal delivery and the placenta followed. The placenta was unremarkable apart from a round mass attached to the membranes. The baby weighed 3.16 kg with Apgar scores of 9 and 10. No congenital anomalies were seen. Two months after delivery the mother and the baby were well.

Gross examination showed a singleton placental disc measuring 20×19×3 cm and weighing 395 g, with a centrally inserted trivascular umbilical cord measuring 29×1.5 cm. Within the membranes there was a round-ovoid mass measuring 7.0×5.0×4.0 cm and weighing 25 grams (Figure 1). The mass was 3.5 cm from the margin of the placenta. The mass was covered by a skin surface with occasional hair-shafts and a few polypoid finger-like projections (Figure 1). There was no axial organization in the form of head, limbs or cranial-caudal poles. The mass was attached to the fetal surface of the placenta.

Figure 1. A) The singleton placental disc with the centrally inserted umbilical cord and the attached mass at the periphery. B) The cut-surface of the mass showing a central core of bone and cartilage surrounded by adipose tissue, with the cystic area seen at one end. Note the vascular pedicle is continuous with the fetal vessels overlying the fetal surface of the placenta.
Figure 2. A) Skin surface with underlying skin appendages and fibroadipose core. (H&E, original magnification ×40). B) Mature cartilage with areas of ossification (H&E, original magnification ×100).

Figure 3. A) Cross-section of the vascular pedicle of the placental mass showing two main blood vessels surrounded by Wharton’s jelly (H&E, original magnification ×40). B) The nerve bundles as highlighted by S-100 immunostain, seen adjacent to the smaller blood vessels in the vascular pedicle of the placental mass (LSAB-Dakocytomation S-100 polyclonal immunostain, original magnification, ×100). C) The vascular pedicle of an ovarian cystic teratoma (unpublished case, illustrated for the sake of comparison), showing two major blood vessels surrounded by Wharton’s jelly with a covering of epithelium (H&E, original magnification ×40). D) The nerve bundles as highlighted by S-100 immunostain, seen adjacent to smaller blood vessels in the vascular pedicle of the case illustrated in 3C. (LSAB-Dakocytomation polyclonal S-100 immunostain, original magnification ×100).
centa by a vascular pedicle measuring 4.0×0.8 cm. The pedicle contained two major blood vessels in continuity with the fetal vessels (Figure 1). The mass was predominantly solid and composed mostly of adipose tissue. A small cyst measuring 1.2 cm filled with clear fluid was also seen at the end of the mass, with the overlying skin covered by a tuft of hair. The center of the mass showed a disorganized collection of bone, cartilage and muscle tissue (Figure 1). Microscopic examination showed an intact skin surface with underlying skin appendages and adipose tissue (Figure 2). The mass was composed mostly of mature adipose tissue arranged in lobules. Foci of neuroglial tissue and ganglions were seen.

The grossly described cyst was, in fact, an area of cystic degeneration surrounded by neuroglial tissue without epithelial or ependymal lining. The core of the mass showed mature bone trabeculae with normal bone marrow tissue surrounded by mature hyaline cartilage and skeletal muscle tissue. The hyaline cartilage showed adjacent foci of ossification (Figure 2). No evidence of organization in the form of a vertebral body was seen. No evidence of gastrointestinal, respiratory or urogenital tract was seen. No immature tissue component was seen. The vascular pedicle showed two blood vessels surrounded by Wharton’s jelly and covered by amniotic membrane (Figure 3). In addition, a few nerve bundles were observed adjacent to smaller blood vessels (Figure 3). The placenta, membranes and the umbilical cord were unremarkable.

Our primary diagnosis was teratoma of the placenta, based on the negative clinical history of early multiple pregnancy, and the absence of skeletal and/or internal organ organization on gross and microscopic examination. However, the differential diagnosis of fetus acardius amorphous was considered on the basis of the presence of a rudimentary umbilical cord.

**DISCUSSION**

Placental teratoma is a very rare non-trophoblastic tumor of the placenta, with only 27 cases reported so far in the literature (Table 1). The main differential diagnosis is fetus acardius amorphous. The main distinguishing criteria proposed by Fox are the following: (a) placental teratomas do not have an umbilical cord attachment to the placenta; they may have a vascular pedicle but without the features of an umbilical cord, i.e., Wharton’s jelly and amniotic membrane, while fetus acardius usually has an umbilical cord which is usually bivascular (with one artery and one vein) even though it can be rudimentary, and (b) fetus acardius amorphous usually has some degree of axial organization, either as cranial-caudal poles, central skeletal development with vertebral column formation, internal organs formation, or recognizable limbs formation, while teratomas usually have a disorganized collection of mature tissue including bone and cartilage.

A review of the literature revealed three cases similar to our case, i.e., with a vascular pedicle attachment and a central core of bone and cartilage, and yet they were called teratomas. Each author described a mass connected to the fetal surface of the placenta by a vascular pedicle with the central core of the mass showing a collection of bone and cartilage. One author described the vascular pedicle as covered by fetal membranes without Wharton’s jelly.

Most of the reported cases were solid with a few exceptions in which cystic teratoma was described. Even though most of the cases were described as lying between the amnion and the chorion and not within the placental substance, one case was reported as lying below the fetal membrane within the placental tissue. Some authors found that those cases that were called fetus amorphous were in fact examples of teratomas, while some cases which were considered as teratomas were actually cases of fetus amorphous. A review of the literature showed that opinions are divided into two groups. One group favors separate entities (splitters). Others prefer a single entity (lumpers). Few authors consider the two entities as an “anatomical continuum” and so the distinction between the two is “meaningless.” Stephens divided fetus amorphous into two subtypes: (a) the more standard type with recognizable organization and (b) the teratoma-like type with no recognizable organization. In their study, they found that some cases of fetus acardius did not have an umbilical cord, while approximately half of the cases have disorganized bone fragments, i.e., no skeletal organization. So, they came to the conclusion that the proposed criteria by Fox are “fragile” and are only suggestions and therefore need further evaluation.

Homunculus dermoid cyst is a well known entity with a high degree of organization. We came across one case of an ovarian mature cystic teratoma (unpublished case) with a central solid ovoid mass with primitive cranio-caudal poles and primitive extremities formation. The cranial pole contained primary brain with meningeal coverings and central ventricular cavity. This mass was connected to the wall of the dermoid cyst by a vascular pedicle with two blood vessels surrounded by Wharton’s jelly and covered by an amniotic membrane-like epithelium (Figure 3). In addition, a few nerve bundles were seen adjacent to smaller blood vessels (Figure 3). This may suggest that placental teratomas can have a high degree of organization and a
vascular pedicle that looks like an umbilical cord, i.e., mimicking acardius amorphous.

Placental teratomas are usually discovered incidentally and are usually not associated with complications, with few exceptions, in which fetal malformations were described. They usually do not show geographical predominance nor familial clustering and tend to be distributed sporadically in different populations (Table 1).

Several theories have been proposed to explain the occurrence of teratomas in the placenta. The favored hypothesis is that proposed by Fox, i.e., aberrant germ cell migration. The primordial germ cells may migrate out through the evaginated gut into the loose connective tissue of the umbilical cord, or, if they continue “astray”, may reach the loose tissue of the fetal surface of the placenta, or even into the fetal membranes. This is supported by the occurrence of teratomas along this line of migration.

Fetus acardius amorphous is also an unusual finding in the placenta. Several cases were also described in animals. Acardius amorphous is considered a result of a blighted fetus from multiple pregnancies, due to either overpowering of the heart of the acardius due to reversed circulation or primary agenesis of the heart of the acardius. Both scenarios can result in undernourishment and reduced development of the acardius fetus.

The conclusion that can be drawn from these hypotheses is that if a twin pregnancy is detected early, then the presence of a mass will support the idea that it is a true fetus acardius rather than a teratoma. Most of the reported cases failed to mention the early findings of a single or twin pregnancy.

Is our case a presentation of fetus acardius amorphous of the teratoma-like type, as cited by Stephens, or is it a true teratoma i.e., a neoplasm of the placenta? Our case shows overlapping features of both teratoma and fetus amorphous, i.e., the presence of an umbilical cord on one side and the absence of axial organization or internal skeletal organization on the other side. However, we feel that our case represents a true teratoma of the placenta. The absence of a multiple pregnancy, the absence of skeletal and/or internal organ organization, and the fact that teratomas can have a high degree of organization, i.e., the homunculus type, will support our primary diagnosis of a placental teratoma. In addition, our observation of the nerve bundles in the vascular pedicle may suggest that this pedicle is a well-organized neurovascular continuation of the teratoma rather than a true umbilical cord. We did not come across any comment on this observation in the previously reported cases. We do not know the exact significance of this observation in the distinction between fetus acardius and placental teratoma. We feel it should be validated by retrospective review of the previously reported cases in the literature.

We feel that separating placental teratomas and acardius amorphous into two different categories is of importance to the parents. The parents have the right to know if the mass was a teratoma i.e.; a true tumor, or if it was a fetus acardius, i.e., a dead twin fetus. This will of course have significant psychological and emotional implications for the parents.

In summary, whether placental teratomas present as true tumors of the placenta or an extreme form of fetus amorphous is not yet resolved, since in certain cases there is an overlap between the two entities. One part of the problem is that the distinguishing criteria are not always helpful. The criteria are helpful in those cases in which the distinction between the two entities is easy. However, in certain cases in which the distinction is difficult, these criteria are not helpful in separating these “gray-zone” cases. Whether more, or totally different criteria are needed to assist in differentiating between the two entities, or to group the two under one pathology with different stages of development and differentiation will certainly await more studies.

We feel that including the clinical information about early evidence of multiple pregnancies may help in dividing between the two entities. The other part of the problem is the rarity of the cases, which precludes an

---

Table 1: Reported cases of placental teratoma.

<table>
<thead>
<tr>
<th>Morville (1925) - France</th>
<th>Akimov (1991) - Russia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kuster (1928) - Germany</td>
<td>Block (1991) - USA</td>
</tr>
<tr>
<td>Perez (1939) - Argentina</td>
<td>Sironi (1994) - Italy</td>
</tr>
<tr>
<td>Fox (1964) - UK</td>
<td>Gayer (1994) - Australia</td>
</tr>
<tr>
<td>Fujikura (1964) - USA</td>
<td>Jaswal (1995) - India</td>
</tr>
<tr>
<td>Joseph (1975) - USA</td>
<td>Wang (1995) - China</td>
</tr>
<tr>
<td>Kobos (1982) - Germany</td>
<td>Koumastakis (1996) - Greece</td>
</tr>
<tr>
<td>Smith (1982) - Australia</td>
<td>Shimojo (1996) - Japan</td>
</tr>
<tr>
<td>Calame (1985) - Holland</td>
<td>Elagos (1998) - Turkey</td>
</tr>
<tr>
<td>Fernandez (1989) - Spain</td>
<td>Chandy (2002) - India</td>
</tr>
<tr>
<td>Unger (1989) - USA</td>
<td></td>
</tr>
</tbody>
</table>
appropriate study, as well as the absence of a genetic study. Therefore, more accumulative and comparative studies supplemented with genetic studies, may help in resolving the issue in the future.

Acknowledgement: We would like to thank Dr. Lakshmiah Ganapathy from the Histopathology Department, Dubai Hospital, for his contribution of the macrophotographs.

REFERENCES