Operation procedure of sacrococcygeal fetus in fetu

Rochadi

Fetus in fetu is a condition in which a fetiform calcified mass often presents in the abdomen of its host, a newborn. It is an extremely rare condition, estimated once in 500,000 deliveries and has a 2:1 male predominancy, with most patients presenting with an abdominal mass in the first year of life. The term fetus in fetu is used to point out an unequal division of totipotent cells of blastocyst where the result is the inclusion of a small cellular mass in the more mature embryo. It was encapsulated, pedunculated and represents a malformed monozygotic, monochorionic, diamniotic parasitic twin. In 80% cases, fetus in fetu is located retroperitoneally but can be found in unusual location such as in oropharynx, neck, skull, mediastinum, pelvis, iliac mesentery, adrenal gland, sacrococcygeal region and scrotal sac.

We report a case of newborn with giant sacrococcygeal fetus in fetu who underwent operative procedure due to bowel obstruction by the mass.

The case

A 21-day-old boy was hospitalized in the neonate ward of Sardjito Hospital with a sacrococcygeal mass. Its size was 30 x 20 x 15 cm³ and covered by skin. The baby was born at 34 week gestation in Sardjito Hospital from a healthy 20-year-old mother in November 2009 by caesarian section due to mass compression to surrounding organs. His birth weight was 5644 gram. CT scan examination of sacrococcygeal mass revealed calcification appearance of skull, limbs bone and vertebrae, thus diagnosis of fetus in fetu was established. Preparations for surgery were including forming a team who were.

A medical team of surgeons, anesthetists, intensive care specialist and laboratory staffs planned an operative procedure to remove this mass. The operation was done after the condition of the baby has been optimized.

In operative room, rectal tube was inserted through the anus to prevent damage to the rectum and to decompress the colon. “W incision” was made in the mass surroundings to make it easier to remove the mass. The mass was detached from all surrounding tissues and the walls of the bowel were closed using 2 ABS-500 sutures. The total operating time was 70 minutes. The child was successfully discharged in good condition after 7 days from the day of operation.

Figure 1. Mass in sacrococcygeal region.

From the Department of Pediatric Surgery, Medical School, Gadjah Mada University/Sardjito Hospital, Yogyakarta, Indonesia.

Reprint request to: Rochadi, MD, Department of Pediatric Surgery, Medical School, Gadjah Mada University/Sardjito Hospital, Jl. Kesehatan No. 1, Sekip, Yogyakarta, Indonesia. E-mail: bedahunakugm@yahoo.com
Rochadi: Operation procedure of sacrococcygeal fetus in fetu

Figure 2. CT Scan: oval soft tissue mass, cranial bones, vertebral column, long bones.

Figure 3. W Incision.

Figure 4. Head with hair, trunk with viscera and limbs with digits.

Figure 5. Post operative body weight 1750 gram.

To prevent bleeding, hemostatic agent was given and all vessels on the mass were ligated. After incision, we discovered a well encapsulated mass and revealed a yellowish fluid and an incompletely developed fetus covered by vernix caseosa. We cut the connecting pedicle and closed the wound. We faced difficulties during operation when separating the bridging vascular system in pre-sacral region and reconstructing the pelvic floor. There was minimal blood, thus no hemodynamic instability during the operation.

Fifteen minutes after four hours of surgery, the baby moved their limbs and opened his eye. There was no problem in bowel function and feeding started one day after operation. Body weight after operation was 1750 gram. Pathological examination revealed bony cranium, composed of a head with hair, and complete extremities. We also found brain, vertebrae and visceral as internal organs.

Discussion

The exact embryogenesis of the fetus in fetu is still controversial, but Potter and Craig propose that the fetus in fetu occurs from the anomalous embryogenesis in a diamniotic monochorionic twin pregnancy inside the body of its fellow twins, but others consider it to represent a highly organized teratoma. Fetus in fetu is thought to result from unequal division of the totipotent inner cell mass of the developing blastocyst. The controversy is
whether the fetus in fetu is a distinct entity or represents a highly organized teratoma. Potter claims that teratoma is defined as a neoplasm with slight potential for malignancy that is composed of multiple tissue foreign to the part in which they are located. A final important feature that has been used to distinguish between fetus in fetu and teratoma is the presence of a vertebral column. Willis emphasizes that the identification of vertebral column secured the diagnosis of fetus in fetu and differentiates this entity from a teratoma. Identification of the vertebral column indicates that there was fetal development of the included twin.\(^2\)

Abdominal fetus in fetu result in compression to adjacent organ leading to abdominal distension, constipation, vomiting, jaundice, urinary retention and distress respiration but sacrococcygeal fetus in fetu result in compression to the rectum and anus leading to intestinal obstruction. The location of fetus in fetu 80% located retroperitonealy but can arise in other side such as in mediastinum, neck, skull, oropharynx, pelvis, iliac mesentery, adrenal gland, sacrococcygeal region and scrotal sac.\(^1,3,4,10\)

Imaging plays an important role in correctly diagnosis of fetus in fetu. Plain abdominal radiograph may be helpful in diagnosis of fetus in fetu which may show presence of vertebral column and axial skeleton. In reality, one may find cases whose vertebral column is insufficiently calcified and therefore invisible on the plain radiograph.\(^7,11\) In the present case, CT Scan examination showed mass with multiple calcification. Non visualization of the vertebral axis on radiography or CT scan does not exclude the diagnosis of fetus in fetu as it can be seen by pathologist.\(^9\)

Most of fetus in fetu were cases of pedunculated masses within a capsule containing fluid. In our case the vertebral column was detected by CT scan examination. It was therefore in accordance with Willis’ theory that fetus in fetu is a mass containing a vertebral axis often associated with other organs or limbs around this axis. However review of the literature showed that in about 9% of cases fetus in fetu, there was no vertebral column, even on pathologic examination. On the contrary, teratoma is an accumulation of pluripotential cells in which there is neither organogenesis nor vertebral segmentation.\(^7\) When fetus in fetu is discovered in a newborn during physical examination, the differential diagnosis includes all the common masses such as Wilms’ tumor, hydronephrosis and neuroblastoma. Laboratory tests, tumor markers, conventional radiographs, sonogram and CT scan are important to determine the diagnosis. Treatment of fetus in fetu was complete resection of the mass except when it was adherent to the host’s organ.\(^7,9\) In the present case, the location of fetus in fetu is in sacrococcygeal region so the operation with complete resection was not difficult.

References