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Case Report

A case of omphalocele associated with teratoma in a new born baby: a very rare case report

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ABSTRACT

An omphalocele is a congenital midline abdominal wall defect at the level of the umbilical cord insertion into the fetal abdomen that results in herniation of intra-abdominal contents into a sac comprising of an outer layer of amnion and an inner layer of peritoneum. The most common eviscerated contents are liver and small intestine though virtually any organ can be seen within the omphalocele sac including colon, pancreas, spleen, omentum, stomach, heart or genitalia. We report an unusual case of omphalocele associated with mature teratoma in Tripura medical college and DR. BRAM teaching hospital. Only five cases of teratoma arising in a case of omphalocele are reported in the literature reviewed.

Keywords: Omphalocele, Teratoma, Children

INTRODUCTION

Omphalocele is a congenital malformation of the anterior abdominal wall, which has a central localization and is characterized by the presence of a hernial sac with contents inside. Most often, the contents of the hernial sac are the liver, and loops of the small or large intestine. The incidence of omphalocele varies from 1 in 4,000 to1 in 10,000.[1-3] In comparison with other malformations of the anterior abdominal wall, omphalocele is often combined with other congenital malformations of the cardiovascular, respiratory or gastrointestinal tracts. Approximately 40% of patients with omphalocele have an abnormal karyotype.[4] Teratoma is a germ cell tumor, which is most often localized in the sacrococcygeal region (89%). Teratomas can be cystic, cystic-solid, or mixed. They can also be found in other areas. Congenital teratomas are most common benign formations, although in recent years there have been reports of early malignancy of the formation. Surgical treatment for small and medium-sized omphalocele without viscero-abdominal imbalance consists of a one-stage reconstruction of the anterior abdominal wall. With a large defect and an insufficient volume of the abdominal cavity, treatment consists in a two-stage immersion of the organs as the volume of the abdominal cavity increases. We describe a clinical case of successful surgical treatment of umbilical cord teratoma in a case of omphalocele major containing liver, small and large intestine as content in Tripura medical college and DR. BRAM teaching hospital.

CASE REPORT

A full term neonate of uneventful antenatal history presented with two large swellings in umbilical region. There was no other complaint. On physical examination the neonate was pink, active and weighed 2.5 kg. Systemic examination was unremarkable. On local examination, there was a large swelling measuring 8×7 cm covered partly by skin and amnion both. A clinical diagnosis of exomphalos major was made. Another swelling measuring 5×4 cm covered fully by skin with tuft of hair in it. The surface of the swelling was irregular

and the consistency is variegated. A clinical diagnosis of umbilical cord teratoma was made. Apart from routine investigations, X-ray chest and echocardiography were done all of which were within normal limits. Surgical treatment was performed 15 hours after birth following stabilization of the new born's condition and correction of water electrolyte balance and impaired coagulation. The tumour was surgically removed without disturbing the omphalocele major component. The tumor-like formation was on the external surface of the omphalocele, 5×4 cm in size with irregular margins along with tuft of hairs and no invasion into adjacent tissues. Inside, a structure of bony-cartilaginous consistency was palpated. Since there was insufficient volume of the abdominal cavity, one-stage plastic surgery of the anterior abdominal wall was not performed. The postoperative period was uneventful. On the third post-operative day there were no signs of inflammation and dehiscence of the surgical wound and it eventually healed by primary intention. The child was discharged on the 11th day after surgery.

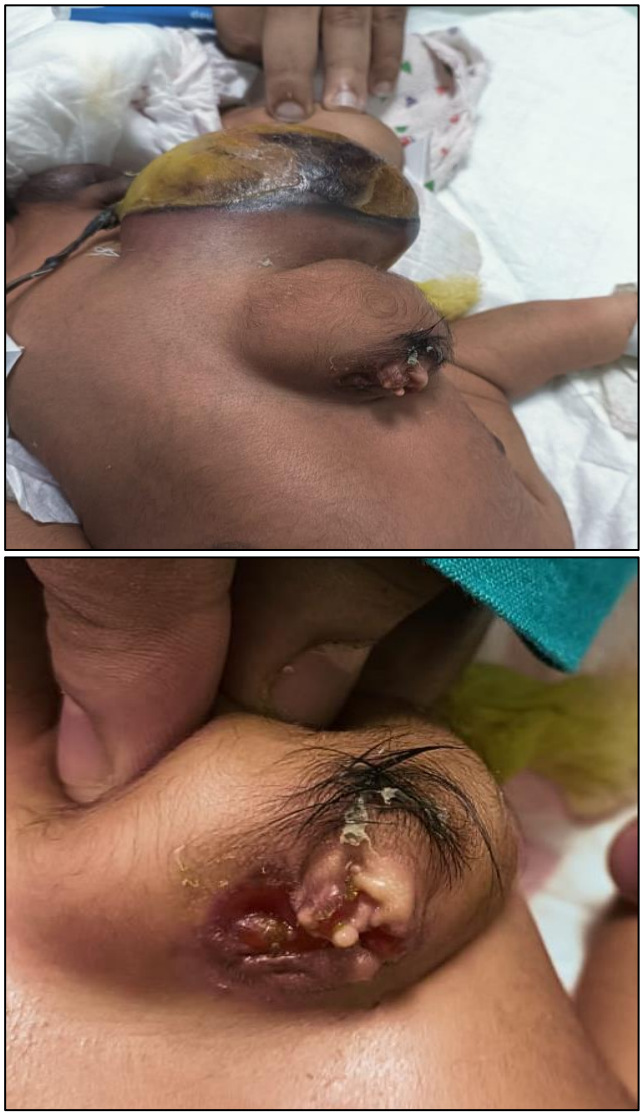

Figure 1: Pre-operative picture of a case of omphalocele major with the umbilical teratoma.

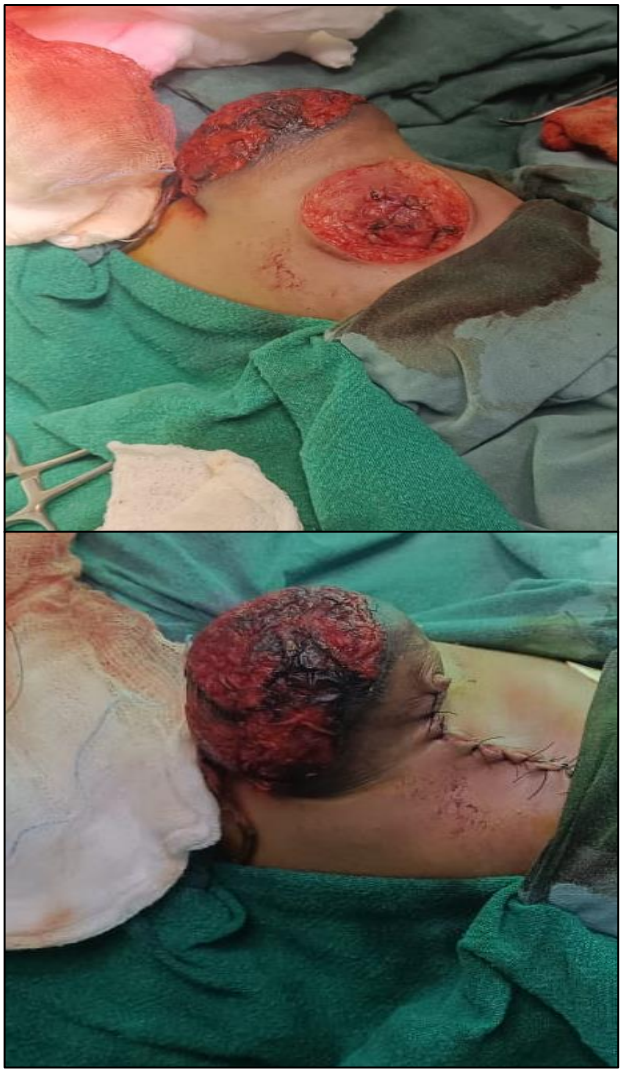

Figure 2: Post operative picture of wide local excision of teratoma with treatment of omphalocele major with 2% mercurochrome solution.

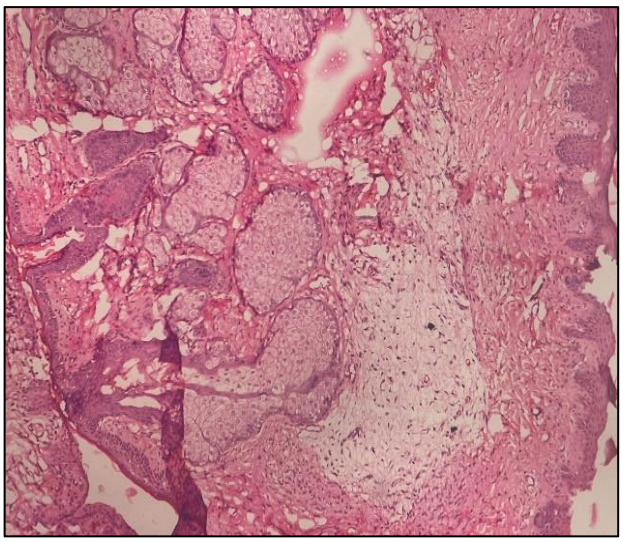

Figure 3: HPE showing hair follicle and the neural tissue.

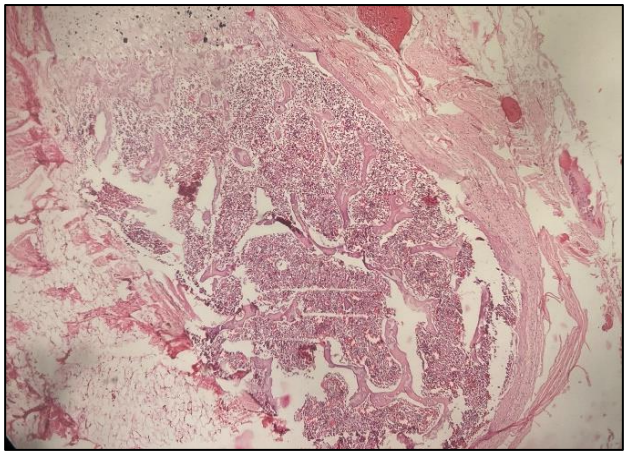

Figure 4: HPE of bone and marrow.

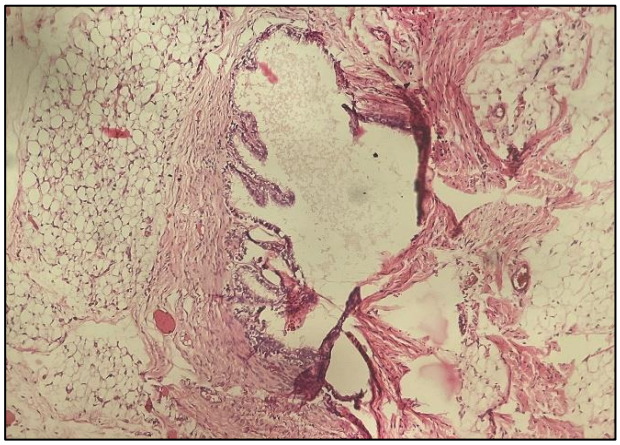

Figure 5: HPE of GI mucosa.

DISCUSSION

The medical literature describes rare cases of congenital umbilical cord teratomas.[5] In our clinical case, the child had a combination of a mature teratoma with a congenital malformation of the anterior abdominal wall-omphalocele. We share our experience of successful surgical treatment of a newborn with this extremely rare combination. One-third of newborns with omphalocele detected in the antenatal period have related developmental abnormalities.[1] The frequency of malformation ranges from 1:5,000-1:10,000 live-born children.[6] Congenital teratomas of the umbilical cord are extremely rare and only 15 cases have been reported in the literature. Five cases included a combination of umbilical cord teratoma and omphalocele, with two early abortions due to chromosomal abnormalities.[7] Histologically, teratomas are often constituted of embryonic cells of all three germ layers. In this case, omphalocele and teratoma are two different pathologies in the same baby at the same time. Since there was significant viscero-abdominal disproportion, surgical treatment was performed to deal with umbilical teratoma by removal of the teratoma with simultaneous regular dressing of the omphalocele with 2% mercurochrome solution. Congenital omphalocele needs to be differentiated from gastroschisis, which is also a congenital malformation of the anterior abdominal wall. Unlike omphalocele, the defect in gastroschisis is most often located to the right of the normally formed umbilical cord, and the hernial sac is absent in most cases. Despite the pronounced changes in the internal organs in gastroschisis and the presence of viscero-abdominal imbalance, the prognosis following treatment is most often favourable. In conclusion, combination of the omphalocele and the teratoma that we have described is extremely rare and difficult to diagnose antenatally. This malformation is correctable and does not constitute an indication for the termination of pregnancy.

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Ethical approval: Not required

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