Intrauterine rupture of sacrococcygeal teratoma : a case report

Ksia. A [1], Ben Saad. M [1], Hadhri. R [2], Ben fredj. M [1], Sfar. S [1], Ben Youssef. S [1], kechiche. N [1], Mosbahi. S [1], Belhassen. S [1], Laamiri. R [1] Sahnoun. L [1], Mekki. M [1], Belghith. M [1], Zakhama. A [2], Nouri. A [1]

[1] Department of Pediatric Surgery Fattouma Bourguiba Hospital; Monastir Tunisia
[2] Department of Anatomo-pathology Fattouma Bourguiba Hospital; Monastir Tunisia

ABSTRACT

Background :

Sacrococcygeal teratoma (SCT) is the most common congenital neoplasm. Intrauterine rupture of SCT has rarely been reported. Prenatal ultrasonography and planned delivery can avoid severe complications such as the tumor's rupture and improve perinatal outcome.

Case report :

We report a case of SCT's rupture during labor. Prenatal sonography was not performed because the mother did not attend antenatal care. An emergency Cesarean section was performed. Hemoglobin concentration of the neonate was 9 g/dL. The patient received a blood transfusion. Surgical resection of the tumor was performed on the second day of life. Posterior sacral approach was used for the excision of the tumor. A wound infection complicated the postoperative course. Follow-up at one year revealed no evidence of recurrence.

Conclusion :

The cornerstone in the management of SCTs is prenatal diagnosis. Delay in diagnosis and treatment may lead to rupture, bleeding and infection.

Key words : sacrococcygeal teratoma, rupture, cesarean section.

INTRODUCTION

Sacrococcygeal teratoma (SCT) is the most common congenital neoplasm with an incidence of 1 in 35,000 to 40,000 live births (1, 2). It is more common in females, with a male: female ratio of 1 :4.
Fetal SCT is being diagnosed increasingly during the early prenatal period by ultrasound. This examination makes it possible to monitor in utero foetal wellbeing and to detect tumor related complications. Planned delivery prevents dystocia and tumor's rupture. This case highlights the clinical presentation and management of SCT's rupture in a neonate.

CASE REPORT

A 45-year-old woman, gravida 5, para 4 was admitted with spontaneous labor at 40 weeks' gestation. Prenatal sonography was not performed because the mother did not attend antenatal care. Fetal distress occured and vaginal delivery was not possible. An emergency Cesarean section was performed. After delivery, the SCT was already ruptured and bleeding. The baby was immediately referred to our department. Physical examination showed a 19 cm in diameter teratoma in the the sacral-buttock area. The mass was irregulate, multi lobulated with some cystic areas. It was firm in consistency, non-pulsatile and non-compressible. The covering skin was redundant, wrinkled and showed some zones of bluish discoloration. Hemoglobin concentration was 9 g/dL. Serum levels of the fetal oncogenes alpha fetoprotein (AFP) and human chorionic gonadotrophin were elevated. Computed tomography revealed a 17 cm non-enhancing pelvic mass without intrapelvic components; a diagnosis of purely external (type I variety of SCT). It seemed to originate from the tip of the coccyx.

Auteur correspondant :

Dr Ksia Amine
Phone : 216 97 523 763
Mail : amineks@yahoo.fr

The neonate received a blood transfusion, and surgical resection of the tumor was performed on the second day of life. Posterior sacral approach was used for the excision of tumor (fig 1). Tumor was excised completely along with coccyx (fig 2 et 3).

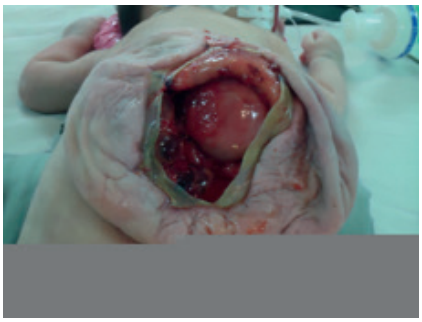

Figure 1 : Preoperative findings: A large ruptured teratoma protruding from the sacrococcygeal region.

Figure 2 : Tumor bed showing divided coccyx.

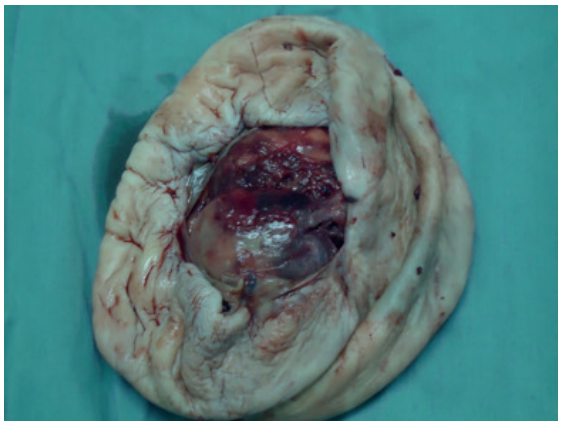

Figure 3 : Macroscopic examination showed a ruptured tumor measuring 19 cm in diameter covered with wrinkled skin.

Soft tissues and neurovascular structures were well preserved. Microscopically, it was a mature benign teratoma. There was a large variety of tissues from all germ layers but no immature elements or malignancies were seen. A wound infection complicated the postoperative course. The isolated germ was Klebsiella and the infection was successfully treated by a combination of Imipenem and Amikacin. Follow-up at one year using clinical, biochemical and radiological assessment revealed no evidence of relapse.

DISCUSSION

SCTs are congenital tumors that develop embryologically from multipotent cells in Hensen's node and enlarge as pre and post sacral masses. They have a 2-5% risk of malignancy (3). Approximately 75% of the affected infants are females. All SCTs involve the coccyx. They may be solid, cystic or a combination of both types like in our case. Altman and al. classified these tumors according to the degree of exterior component or intrapelvic extension (4).

Altman Type I tumors (46.7%) are completely external. Type II tumors (34.1%) are present externally but have significant internal components. Type III tumors (8.8%) are apparent externally but they lie mostly internally. Type IV tumors (9.8%) are completely internal with no external presentation. CT determines the intrapelvic extension of SCT and thus classifies the tumor. During pregnancy, SCT's prognosis is mainly determined by acute and chronic hydramnios, high-output cardiac failure, placentomegaly and hydrops fetalis.

Tumor rupture may be caused by uncontrolled labor or complications during delivery (5). This complication increases the morbi-mortality because of acute bleeding and anemia (6,7,8). During labor, severe dystocia may occur in 6% to 12% of the cases, and this is usually encountered in cases of infants with large tumors delivered vaginally. In our case, the rupture happened during labour. Bleeding, anemia and infection were the consequences of the tumor's rupture.

Prenatal diagnosis of SCT is often made during antental period by ultrasounds which can also predict the onset of complications. Early diagnosis enhances coordinated and well-planned management of the patients. When the tumor diameter is greater than 5 cm of biparietal diameter of the fetus head, elective caesarean delivery is indicated (8). This mode of delivery will forestall possible tumor rupture.

Fetal surgical interventional procedures could be performed when the diagnosis is made early in pregnancy (9, 10). Three-dimensional sonography now allows prenatal diagnosis of SCT even in the first trimester (11, 12). At birth, excisional surgery must be performed as early as possible to prevent infection, haemorragic shock and malignant transformation.Various surgical approaches have been defined. The coccyx should always be resected with the tumor, to avoid local recurrence. Chemotherapy is indicated in malignant cases (13). The prognosis of prenatally diagnosed SCT differs from postnatally diagnosed SCT.

Prenatal assessment of the fetus is critical for reducing the morbi-mortality of these tumors especially that now open and minimal access techniques of fetal intervention have been shown to be feasible (14).

CONCLUSION

The cornerstone in the management of SCTs is prenatal diagnosis. Delay in diagnosis and treatment may lead to rupture,bleeding and infection.

Conflict of interest : none.

REFERENCES

[1] Backer D, Erpicum P, Philippe P, Demarche M, Otte JB, Schwagten K, et al. Sacrococcygeal teratoma: Results of a retrospective multicentric study in Belgium and Luxembourg. Eur J Pediatr Surg.2001;11:182–185.

[2] Pantanowitz L, Jamieson T, Beavon I. Pathology of sacrococcygeal teratomas. S Afr J Surg.2001;39:56–62.

[3] Dehner LP. Soft tissue, peritoneum and retroperitoneum.Pediatric Surgical Pathology. 2nd ed. Baltimore (MD):Williams and Wilkins, 1987.

[4] Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma : American Academy of Pediatrics Surgical Section Survey-1973. J Pediatr Surg 1974;9:389-98.

[5] Yamaguchi Y, Tsukimori K, Hojo S, Nakanami N, Nozaki M, Masumoto K, Taguchi T, Wake N.: Spontaneous rupture of sacrococcygeal teratoma associated with acute fetal anemia.Ultrasound Obstet Gynecol. 2006 Oct;28(5):720-2.

[6] Schmidt B, Haberlik A, Uray E, Ratschek M, Lackner H, Höllwarth ME.Sacrococcygeal teratoma: clinical course and prognosis with a special view to long-term functional results.Pediatr Surg Int. 1999;15(8):573-6.

[7] Hoehn T, Krause MF, Wilhelm C, Lattermann U, Rueckauer KD. Fatal rupture of a sacrococcygeal teratoma during delivery. J Perinatol. 1999 Dec;19(8 Pt 1):596-8.

[8] Kay S, Khalife S, Laberge JM, Shaw K, Morin L, Flageole H.Prenatal percutaneous needle drainage of cystic sacrococcygeal teratomas.J Pediatr Surg. 1999 Jul;34(7):1148-51.

[9] Hedrick HL, Flake AW, Crombleholme TM, Howell LJ, Johnson MP, Wilson RD, Adzick NS. Sacrococcygeal teratoma: Prenatal assessment, fetal intervention and outcome. J Peadiatr Surg.2004;39:430–438.

[10] Sy ED, Lee H, Ball R, Farrell J, Poder L, Nobuhara KK, Farmer DL, Harrison MR. Spontaneous rupture of fetal sacrococcygeal teratoma. Fetal Diagn Ther. 2006;21(5):424-7 .

[11] Legbo JN, Opara WE, Legbo JF. Mature sacrococcygeal teratoma: case report. Afr Health Sci. 2008 Mar;8(1):54-7.

[12] Ruano R, Duarte S, Zugaib M. Percutaneous laser ablation of sacrococcygeal teratoma in a hydropic fetus with severe heart failure--too late for a surgical procedure? Fetal Diagn Ther. 2009;25(1):26-30.

[13] Match M, Arya NC. Sacrococcygeal teratoma : Two case reports and a review. Obstetrics and Gynaecological Communications. 2000;2:34–36.

[14] Hirose S, Farmer DL. Fetal surgery for sacrococcygeal teratoma. Clin Perinatol. 2003 Sep;30(3):493-506.

Acknowledgment : this work was supported by the Tunisian ministry of scientific research (scientific laboratory LR 12 SP 13).