

Antenatal Diagnosis of a Sacrococcygeal Teratoma in Its Cystic Form: A Case Report

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Abstract: Although rare, sacrococcygeal teratoma is the most frequent congenital tumor and often benign, detectable before birth by obstetrical ultrasound during pregnancy follow-up. We report the case of a patient with a poorly followed pregnancy in whom the ultrasound scan during an etiological workup of hydramnios revealed the presence of a sacro-coccygeal cystic lesion suggestive of a type I teratoma that was managed by pediatric surgeons after birth. The prognosis of this tumor depends on the precocity of the diagnosis and treatment, which should always be considered in case of associated hydramnios.

Keywords: Sacro-coccygeal teratoma, benign tumor, hydramnios, Antenatal diagnosis, cystic form

Introduction:

Fetal tumors are relatively rare, and early prenatal diagnosis can predict potential complications and decide on appropriate treatment. Sacrococcygeal teratoma is the most common tumor (1 in 35,000-40,000 newborns) and occurs most often in female infants [2,3,4]. It is a mostly benign tumor, detectable before birth. We report a case of prenatal diagnosis of a sacrococcygeal teratoma in its cystic form, ALTMAN type II, which was managed by pediatric surgeons with good outcome. This paper deals with the role of ultrasonography in the early prenatal diagnosis of sacrococcygeal teratoma.

Case report:

A 35-year-old primigravida, admitted to the obstetric emergency room for increased uterine height in relation to gestational age, in whom the examination found a patient stable on the hemodynamic and respiratory level with a gynecological examination: uterine height to 34 cm in a patient out of labor. Obstetric ultrasound was done objectifying the presence of a cystic sacro-coccygeal lesion partitioned with multiple septates and extension mainly extra pelvic making 100 mm of major axis, with large amniotic cistern to 11 cm (fig 1), the systolic velocity of the middle cerebral artery was normal, with signs of maturity that are complete, a delivery by high route was programmed within 48 hours after notifying the pediatric surgeons and neonatologists, giving birth to a newborn female, birth weight 3500g, pink, tonic and reactive with the presence of a sacro-coccygeal mass of 10 cm of large axis, soft to palpation, polylobed with a bluish coloration in its lower part without anorectal malformations or mass to the abdominal examination (fig2). The baby was hospitalized in the pediatric surgery department where a CT scan of the lumbar spine (fig 3) was performed showing a large sacro coccygeal lesion formation attached to the coccyx, with well-limited fluid density, roughly oval in shape, poly-lobed, measuring 96*62*97 mm in diameter, this lesion formation is of endo and exo-pelvic development, the endo-pelvic portion is significant, of right latero-rectal seat pushing back the uterus and the bladder upwards and forwards the exopelvic portion is predominant, median and right paramedian, exophytic through a wide pedicle of 16 mm, this mass pushes the sacrum medially, without signs of invasion or endo canal extension, no haemorrhagic changes or sacro coccygeal vertebral anomaly, this aspect first evokes a mature sacro coccygeal teratoma in its cystic form, type II according to ALTMAN's classification. An alpha foetoprotein and HCG assay was done and came back negative. A large exeresis of the mass was performed in the operating room after careful dissection from the muscles, sacrum and rectum. The postoperative follow-up was simple.

Histologically, it is a skin tissue with a cystic formation in the dermis bordered by a squamous epithelium with the presence of skin appendages of fibrous and adipose tissue. There is also the presence of a choroid plexus and mature brain tissue with oligodendrocytes and neurons arranged on a fibrillary background. This concludes in a mature and pluritissular teratoma.

Discussion:

Teratomas are germline tumors containing tissue from two or all three embryonic layers (ectoderm, mesoderm and endoderm) and are the most common tumor in the neonatal period. The etiology of teratomas is not yet clear, but may result from the migration of totipotent cells located near the node of Hensen in the embryo. Teratomas, especially those diagnosed in the neonatal period, occur most frequently in the sacrococcygeal region. Nevertheless, they can be found in other locations depending on the location of migration of the totipotent cells [1]. The incidence of sacrococcygeal teratoma is approximately 1/40,000 and 1/35,000 live births [2]. There is a clear female predominance with a sex ratio of 1/3 to 4 [3,4]. The appearance of these tumors is highly variable. The most typical presentation is that of a mass discovered at birth. The size and weight

are variable; it may be a small sacral mass, or a huge mass comprising almost half the baby's weight. Lesions may be cystic, solid, or mixed: - Solid teratomas are the rarest, but they have a high potential for malignancy. - Cystic teratomas are usually benign and have a good prognosis. The origin of the fluid is thought to be the functional choroid plexuses within the mass that produce cerebrospinal fluid. - Mixed teratomas are the most common [5].

The overlying skin may be normal in appearance, taut and shiny, rough and wrinkled, hemangiomatous, or ulcerated. Tumor growth may be in the medial direction, producing a pendulous mass, or may extend preferentially to one side, presenting as a distinct gluteal mass or simply as a minor gluteal asymmetry [6].

In 1974, ALTMAN et al [7]. Proposed a topographic classification of sacrococcygeal teratomas in their report to the American Academy of Pediatric Surgery (Figure 4): Type I: Type I teratomas are almost exclusively external with a minimal pelvic component. Type II: Type II teratomas have a significant pelvic component. Type III: the intra-abdominal and intra-pelvic component is much greater than the external component. Type IV: They are exclusively pre-sacral with no external component, this is the most difficult form to diagnose both clinically and on ante- or post-natal ultrasound.

Sacrococcygeal teratoma is considered a surgical emergency because of its potential for malignant transformation from the 4th month of life [8], the tumor removal must be radical, complete and associated with a coccygectomy, to avoid recurrence, which is more likely to occur in a malignant form, even if the tumor was initially benign. The anatomopathological study confirms the diagnosis. It shows a tumor made up of somatic tissues, derived from the three ecto-, endo- and mesodermal layers. According to their degree of differentiation, immature teratomas are distinguished from mature teratomas. A therapeutic complement by chemotherapy is essential in the malignant forms [9].

Serum tumor markers play an important role in the management of MGT, both for diagnosis and for monitoring the efficacy of the treatment or for the detection of possible relapse. These markers are represented by AFP, a marker of vitelline tumors, hCG, a marker of CC and to a lesser extent of germinoma [10].

Interpretation of AFP remains difficult because it is physiologically elevated during fetal development and at birth. Tsuchida et al [11] proposed a graph for the interpretation of AFP levels by designating normal values in 61 normal newborns, aged 0-300 days (fig5)

The prognosis is generally excellent in the neonatal period but becomes progressively poorer as the child ages. In addition to the histological type of the tumor, which is benign in 90% of cases, other prognostic factors of CST are: its size as well as its extension, the degree of prematurity and finally the complete resection of the tumor [12,13]. Until recent years, CSTs were discovered incidentally at the time of delivery and, depending on their size, could sometimes pose serious obstetrical problems. However, the advent of ultrasound has allowed the antenatal diagnosis of CST [14], and thus better obstetrical and neonatal management of patients.

Postoperative clinical, biological and radiological monitoring should be performed regularly to look for functional sequelae and early detection of recurrence [15].

Conclusion

Early prenatal diagnosis is important. Once sacrococcygeal teratoma is diagnosed, clinicians should be aware of maternal and fetal complications. The expectant parents should be informed by the multidisciplinary team management and prognosis of the baby. Prompt surgical excision of the sacrococcygeal teratoma after birth should be suggested.

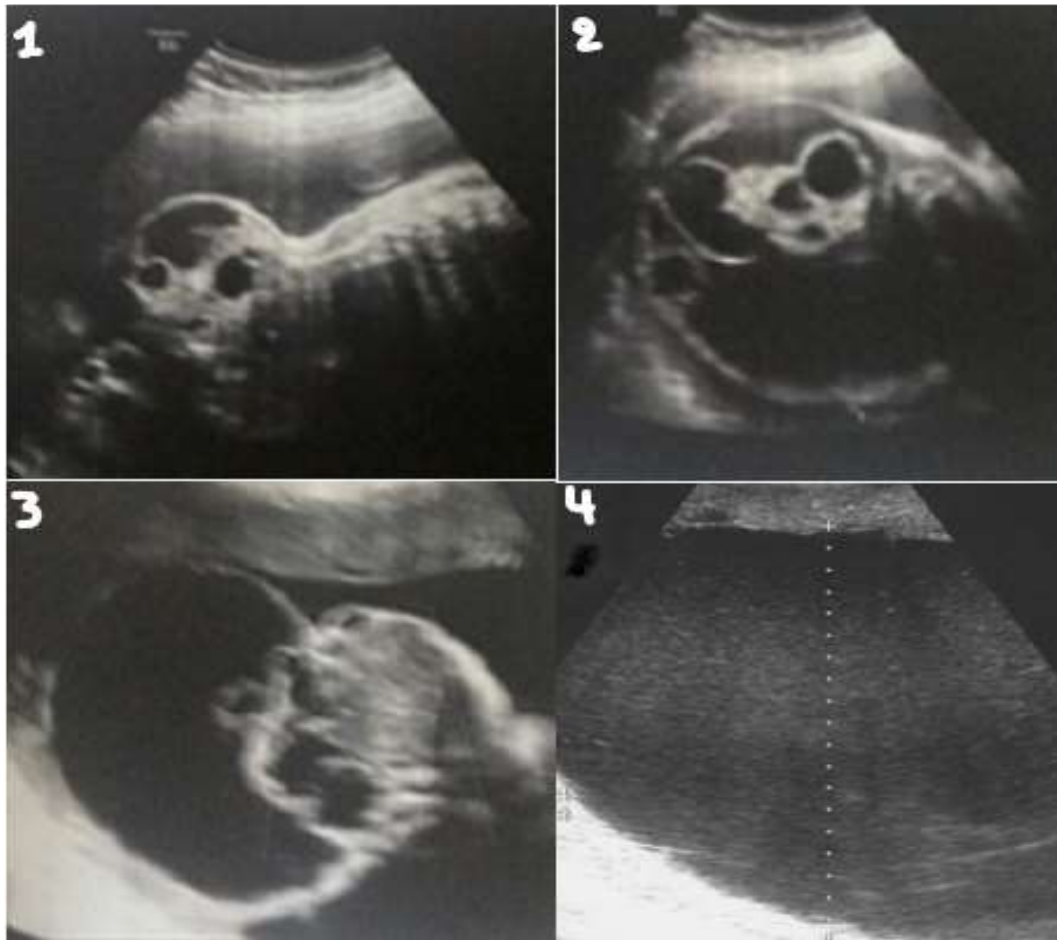


Figure1 : Images échographiques montrant une masse kystique sacro-coccygienne avec de multiples septas associée à un hydramnios



Figure 2 : photos montrant une masse sacro-coccygienne polylo bée avec aspect bleuâtre

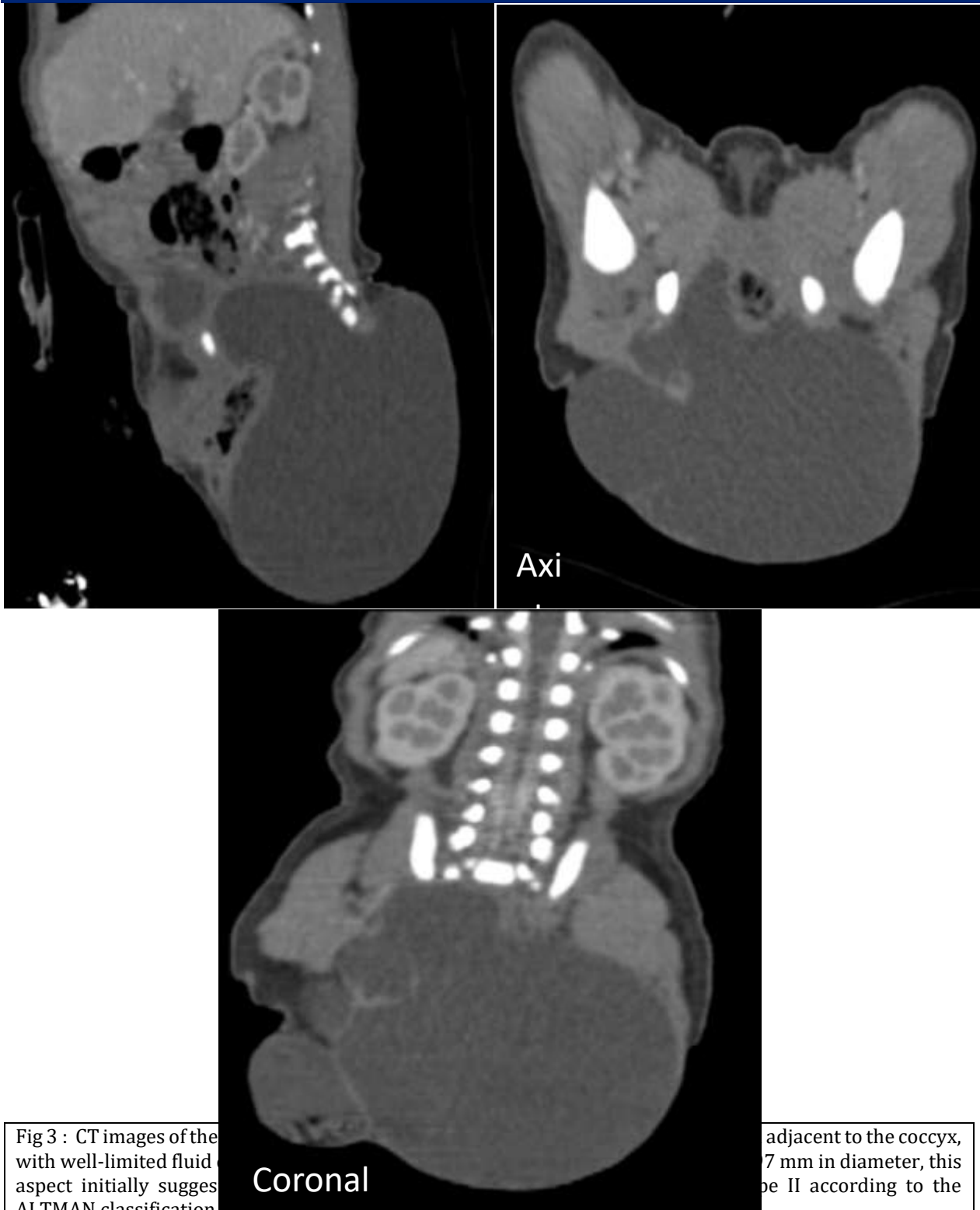


Fig 3 : CT images of the pelvis showing a well-limited fluid-filled mass. This aspect initially suggests a cystic lesion according to the ALTMAN classification.

adjacent to the coccyx, 7 mm in diameter, this is classified as a Type II according to the

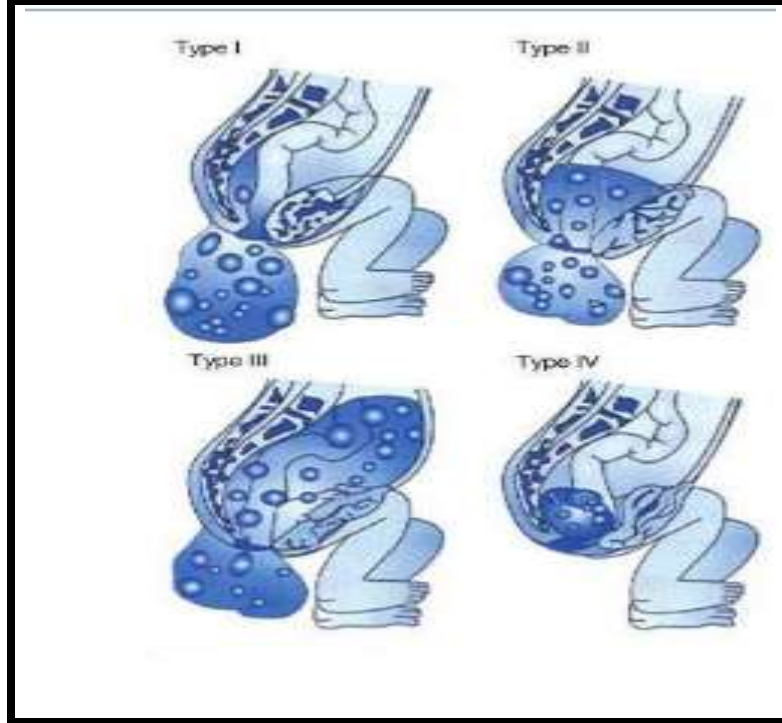


Fig 4 : ALTMAN classification of sacrococcygeal tumors

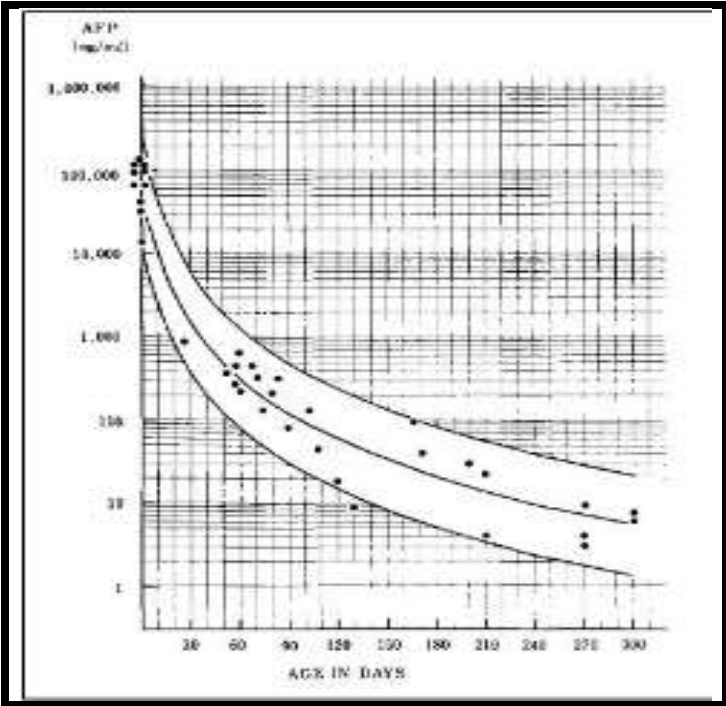


Fig 5 :Tsushida's graph showing the range of normal AFP values as a function of age

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