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Early diagnosis and prenatal care of a foetus with sacrococcygeal teratoma – a case study

Wczesne rozpoznanie i opieka prenatalna nad płodem z teratomą krzyżowo-guziczną – opis przypadku

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Abstract

Objective: Assessment of cases of sacrococcygeal teratoma (SCT) in terms of early prenatal diagnosis, intrauterine foetal monitoring, common complications, and available treatment methods. **Materials and methods:** Retrospective review of literature data regarding cases of SCT diagnosed in 2009–2023. Description of the course of pregnancy in a 29-year-old patient diagnosed with SCT in the foetus in the first trimester of pregnancy. Ultrasound examinations were performed by one of the authors using the Voluson E8 and E10 systems. **Results:** A patient with SCT was successfully diagnosed during the first trimester of pregnancy. This allowed the implementation of proper supervision over the development and well-being of the foetus and the condition of the mother. These activities enabled the pregnancy to be completed successfully and the newborn to be delivered in good condition for further surgical treatment. **Conclusion:** Diagnosis of SCT is possible using ultrasound examination even in the first trimester of pregnancy. Monitoring tumour progression and assessing cardiovascular function are essential elements of surveillance of a foetus with SCT. Evaluation of prognostic factors such as tumour size, morphology, and Altman type helps provide parents with a prognosis for the foetus and newborn. A multidisciplinary approach plays an important role in achieving the best possible treatment results.

Keywords: teratoma, sacrococcygeal region, prenatal diagnosis, prenatal ultrasound (prenatal US), echocardiography, first trimester

Streszczenie

Cel: Ocena przypadków potworniaków krzyżowo-guzicznych (*sacrococcygeal teratoma*, SCT) z uwzględnieniem wczesnej diagnostyki prenatalnej, wewnątrzmacicznego monitorowania płodu, występujących powikłań oraz dostępnych metod leczenia. **Materiał i metody:** Retrospektywny przegląd danych z piśmiennictwa dotyczący przypadków SCT diagnozowanych w latach 2009–2023 oraz opis przebiegu ciąży u 29-letniej pacjentki z rozpoznaniem SCT u płodu w I trymestrze. Badania ultrasonograficzne zostały wykonane przez jednego z autorów z wykorzystaniem aparatów Voluson E8 i E10. **Wyniki:** W I trymestrze ciąży trafnie rozpoznano u płodu SCT. Umożliwiło to wdrożenie prawidłowego nadzoru nad rozwojem i dobrostanem płodu oraz stanem matki, a także pomyślne ukończenie ciąży i przekazanie noworodka w stanie dobrym do dalszego leczenia chirurgicznego. **Podsumowanie:** Rozpoznanie SCT jest możliwe przy zastosowaniu badania ultrasonograficznego już w I trymestrze ciąży. Monitorowanie progresji guza i ocena wydolności układu krążenia

to podstawowe elementy nadzoru nad płodem z SCT. Ocena czynników prognostycznych, takich jak wielkość guza, morfologia czy typ wg Altmana, umożliwia przedstawienie rodzicom rokowania dla płodu i noworodka. Podejście wielodyscyplinarne odgrywa istotną rolę w uzyskaniu jak najlepszych wyników leczenia.

Słowa kluczowe: potworniak, okolica krzyżowo-guziczna, diagnostyka prenatalna, ultrasonografia prenatalna (USG prenatalne), badanie echokardiograficzne, I trymestr

INTRODUCTION

Teratoma belongs to the group of tumours of embryonic origin and, like all such tumours, it originates from immature (primordial) germ cells located in Hensen's node. The germ cell non-physiologically differentiates into all germ layers (ectoderm, mesoderm, and endoderm). The most common location of teratomas in the neonatal period is the sacrococcygeal region. Other possible tumour locations include the brain, middle pharynx, mediastinum, gonad, abdominal cavity, and retroperitoneal space or pericardium[1,2].

Despite their rarity, angiosarcomas of the sacrococcygeal region are the most common group of neoplasms found in newborns. They occur with a frequency of 1:35,000 to 1:40,000 live births, more frequently in girls than in boys (3–4:1)[1,3]. They occur sporadically, and in most cases chromosomal abnormalities have not been described[4].

According to Altman's classification (AC), which includes intra- and extramedullary spread of the tumour mass, there are four types of SCT[3] (Tab. 1).

Tumours with an external component, i.e. types I, II, and III, are easier to diagnose prenatally as well as in the neonatal period, and have a low malignancy potential[5]. Type IV tumours, due to their intraparenchymal growth, are diagnosed later, sometimes not until the postnatal period, and have a higher malignancy potential[6,7].

Ultrasonographically, tumours can be divided into solid, cystic, and mixed forms[1]. This classification is an important prognostic factor. Tumours with a predominantly cystic structure account for 15% of cases diagnosed prenatally, are poorly vascularised, and pose less of a threat to intrauterine foetal well-being. Tumours with a cystic-solid structure are the most common, accounting for 71–96% of cases. In contrast, solid tumours are the least common but tend to be richly vascularised and are associated with the highest rate of complications[4]. Abundant vasculogenesis and multiple arteriovenous leaks in the tumour mass cause circulatory volume overload, which initially triggers foetal defence mechanisms such as tachycardia and cardiomegaly, but ultimately leads to foetal circulatory failure, non-immune oedema, and intrauterine death.

Type of teratomas	Description
Type I	Predominantly external with small internal components
Type II	Dumbbell-shaped tumour with similar internal and external components
Type III	Predominantly internal and smaller external components
Type IV	Exclusively internal (intrapelvic) tumour

Tab. 1. Altman classification of sacrococcygeal teratoma according to the anatomical configuration[1]

Other foetal complications of SCT include hydronephrosis due to urinary tract obstruction, and infiltration of the sacrum and other adjacent tissues. Long-term complications may range from anal sphincter dysfunction, bowel dysfunction, and lower extremity muscle weakness or paralysis, to tumour recurrence with potential malignancy[8].

There is also a histopathological distinction of teratomas into mature and immature forms. Mature teratomas are characterised by fully differentiated tissues, such as bones, teeth, and hair. Immature angiosarcomas contain embryonic components or incompletely differentiated tissue structures, which increases the risk of malignancy. Newborns typically have benign lesions. After complete removal of mature haemangioma, the prognosis is favourable, regardless of the location of the tumour. The immature form metastasises and also leads to persistent recurrence after incomplete excision[1].

In addition to ultrasound, magnetic resonance imaging (MRI) is useful in the diagnosis of SCT, as it is a more precise technique for evaluating the intrauterine portion of the tumour. MRI allows assessment of tumour penetration and ingrowth into the spinal canal, and it is useful in differentiating other pathologies of the area, especially when diagnosing SCT during the second half of pregnancy or when the patient is obese[9]. In 15% of foetuses with SCT, sacral anomalies, spina bifida, meningo-spinal hernia, overgrown anus, and uterine and vaginal defects may be present[10]. The differential diagnosis of SCT should also include lipoma, hamartoma-like tumours, haemangioma, lymphangioma, and lymphangioendothelioma[6].

Intrauterine treatment of sacrococcygeal teratoma is considered when the prognosis for the foetus is poor and when the benefits of the procedure outweigh the risk of preterm delivery. Treatment can be symptomatic: amnioreduction – in the case of polycystic delivery, or transplacental pharmacotherapy – administration of digoxin for symptoms of foetal circulatory failure. Surgical treatment involves opening the uterine cavity to facilitate the resection of the tumour. An alternative to this treatment is minimally invasive procedures, performed under ultrasound guidance. These include percutaneous laser coagulation, radioablation, vascular thermocoagulation or embolisation with coils or alcohol[4]. In the literature, there is no consensus on the optimal treatment method for large, rapidly growing SCTs with full-thickness circulatory failure. Decisions on the type

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of treatment depend on the experience of the facility and are made on a case-by-case basis.

The primary treatment is surgical management after delivery, which includes complete resection of the tumour with subsequent shell reconstruction. Due to the risk of presence of a malignant lesion, the sacrum is removed in its entirety during surgery[1].

CASE REPORT

A primiparous woman, 29 years old, Caucasian, was referred to the Prenatal Screening Programme at 13 weeks' gestation because of a cystic lesion in the distal part of the spine. The patient is a non-smoker with Hashimoto's disease, and a carrier of Group B *Streptococcus* (GBS), with no other significant medical history. The previous course of pregnancy was uncomplicated. The examination, conducted on 5 May 2023 at the "Volumed" Specialised Clinic in Kraków, confirmed the presence of a cystic lesion measuring 3.4 × 3.6 mm, located superficially between the hyoid bone and the genital region of the foetus (Fig. 1 A, B). The crown-rump length (CRL) was 69 mm (13 + 0 weeks' gestation) and nuchal translucency (NT) – 1.6 mm. No other pathological changes in the foetal structure were found. The adjusted risk for trisomies T21, T18 and T13 was within the low-risk group.

At 15 + 3 weeks' gestation, a follow-up transabdominal and intra-abdominal ultrasound was performed along with a perinatology consultation. A solid, poorly vascularised

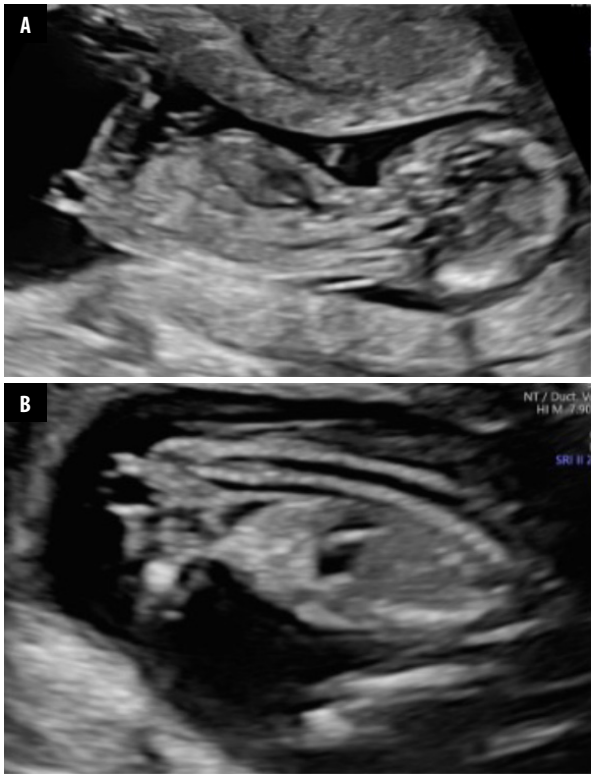

Fig. 1 A, B. SCT image at 13 + 0 weeks'

Pregnancy period [weeks' gestation]	Size of the internal component [mm]	Size of the external component [mm]
13 + 0 (ultrasound I trimester)	3.4 × 3.6	–
15 + 3	10 × 8	14 × 11
17 + 4	10 × 9	14 × 12
20 + 3 (ultrasound II trimester)	15 × 10	19 × 17
23 + 4	20 × 16	33 × 31
26 + 4	33 × 18	38 × 37
32 + 4	70 × 32	37 × 37
34 + 2	42 × 33	49 × 38
36 + 4	85 × 30	54 × 42

Tab. 2. Dimensions of SCT according to the period of pregnancy

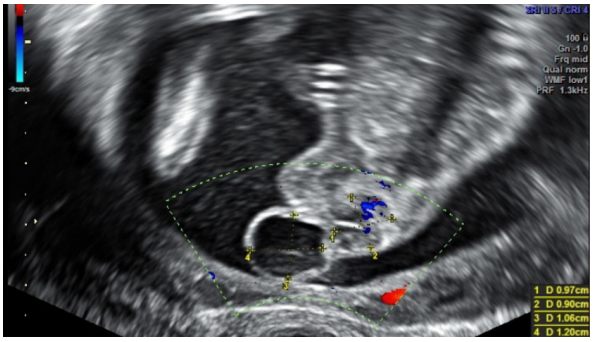

Fig. 2. Vascularisation of SCT at 17 weeks' (ultrasound TV head imaging)

tumour, measuring 10 × 7 × 8 mm, was visualised in the sacrococcygeal region inside the foetal pelvis, connecting with an external cystic lesion in the gluteal region measuring 14 × 10 × 11 mm. A diagnosis of teratoma of the sacrococcygeal region was made. The circulatory system was functional. Subsequent follow-up examinations and tumour dimensions are shown in Tab. 2 and Figs. 2–5.

A foetal echocardiogram at 22 weeks' gestation confirmed normal cardiac anatomy and cardiovascular capacity. Compared with the previous examination, there was a predominance of cystic structures in the intraparenchymal part of the tumour and a slightly larger cystic external part. The lesion was still poorly vascularised. The patient was consulted online with the foetal therapy centre at the Children's Clinical Hospital in Warsaw. Due to the cystic type of the lesion and the absence of signs of foetal circulatory failure, further observation without intrauterine intervention was recommended.

A follow-up foetal echocardiogram was performed at 30 + 1 weeks' gestation, showing mild cardiomegaly (Ha/Ca – 0.38), but the foetal circulatory system was efficient. The cystic type of SCT and slight progression of the lesion were confirmed.

Due to the increased dynamics of tumour growth after 30 weeks and the reduced amount of amniotic fluid, which made it difficult to assess the intra-abdominal part of the tumour, an MRI scan was performed at 33 + 3 weeks.

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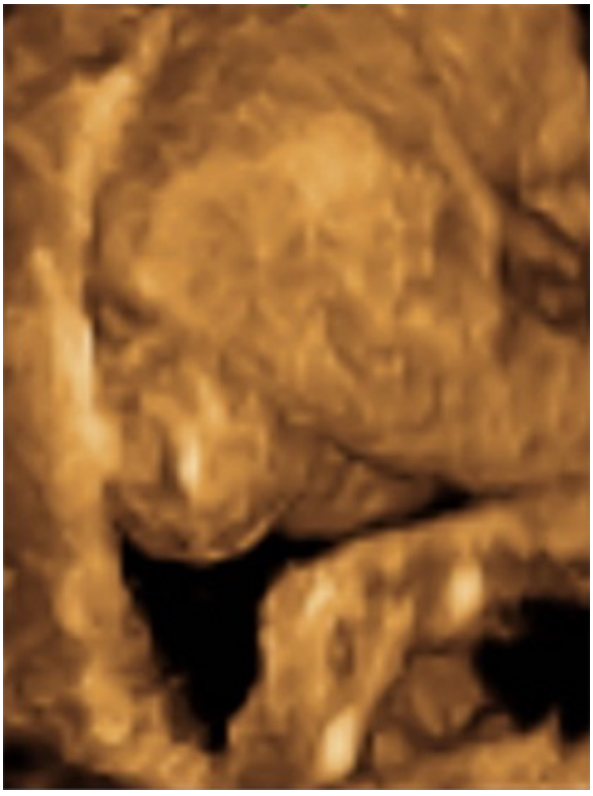

Fig. 3. 3D imaging – external component of SCT (17 + 4 weeks')

The examination confirmed the cystic type of SCT. A well-demarcated polycystic lesion measuring 71 × 34 mm was described inside the foetal pelvis. The connectivity of the tumour with the spinal canal was not visualised. In the sagittal axis, a cystic lesion with a thickened wall up to 7 mm and the largest dimension of 5 cm was identified in the interscapular region. The bowel loops and urinary bladder were displaced and deformed by the tumour.

The pregnant woman was admitted at 37 + 2 weeks to the Ujastek Obstetrics and Gynaecology Hospital in Kraków on an elective basis for observation and preparation for the termination of pregnancy. Delivery by elective caesarean section took place at 37 + 3 weeks. A live term daughter was born with a birth weight of 2,980 g and a length of 55 cm, scoring 10 on the Apgar scale at 1, 3, 5, and 10 minutes post-birth. Physical examination of the newborn showed a convex tumour, measuring approximately 70 × 60 mm, covered with skin with visible bruising in the dorsal part, and with epidermal loss. In the abdominal part of the tumour, the skin was thicker, undulating, with two fistulas. The child was transferred to the Intensive Care and Neonatal Pathology Unit, and then moved in good general condition to the Department of Surgery at the University Children's Hospital in Krakow, where tumour resection was performed via abdominal and perineal

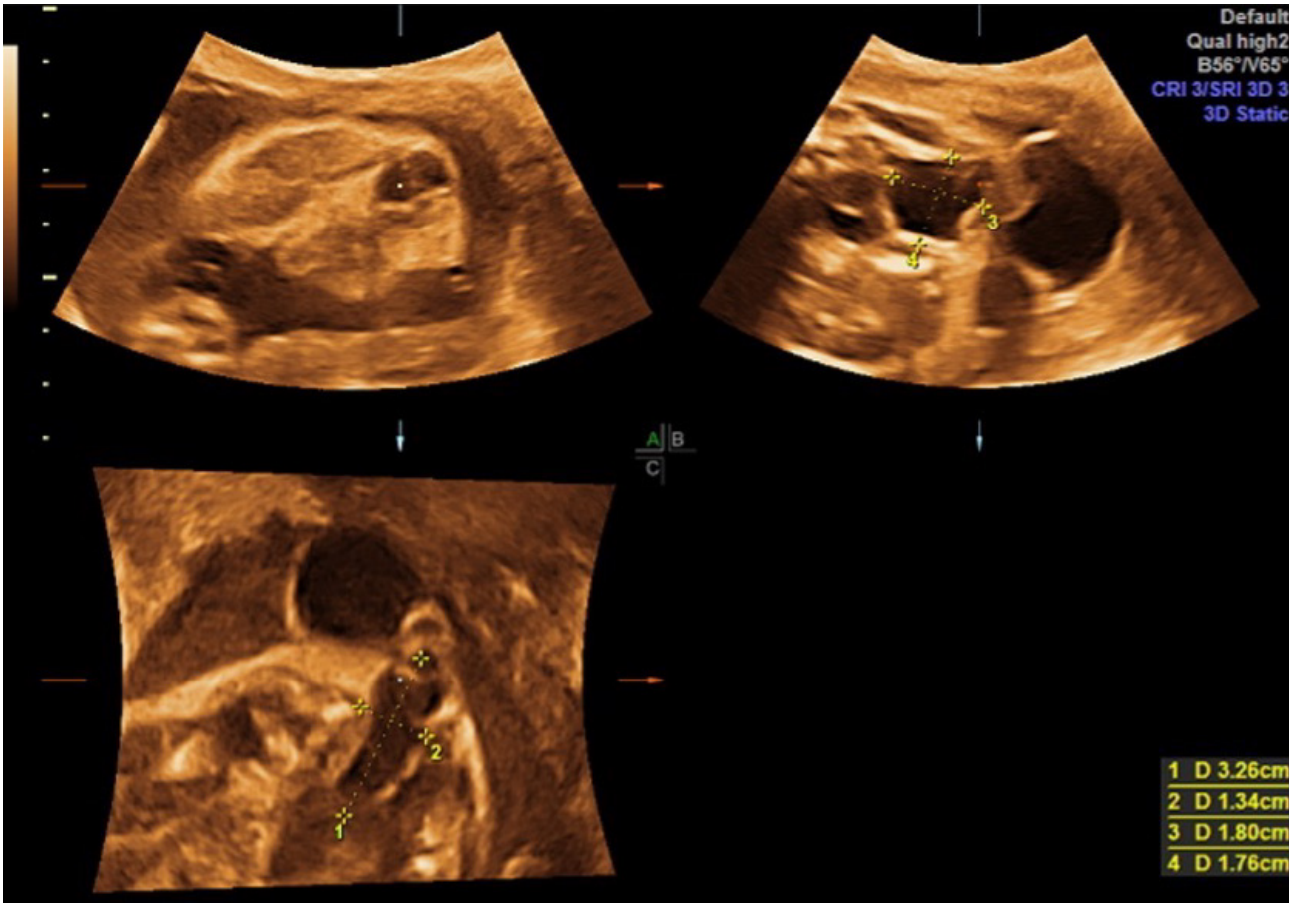

Fig. 4. 3D imaging – image of the tumour in relation to adjacent structures (26 + 4 weeks')

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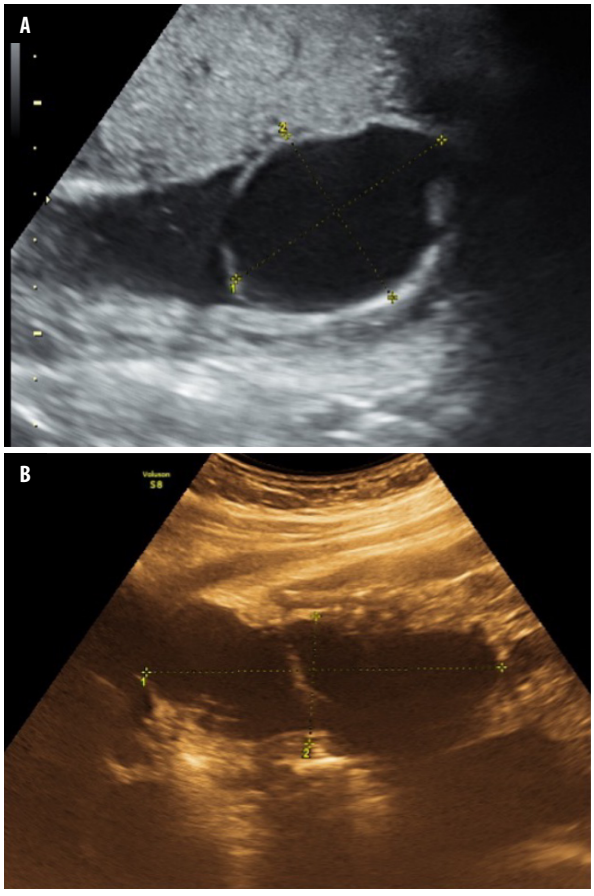

Fig. 5. Tumour image at 36 + 4 weeks'. **A**. *Outer component of SCT.* **B**. *Intra-abdominal component of the SCT*

access. Histopathological examination diagnosed mature teratoma (teratoma maturum). The condition of the newborn after the procedure was good. In further outpatient care the child did not show any urinary tract or lower bowel complications.

DISSCUSION

The paper presents a case of sacrococcygeal teratoma correctly diagnosed in the first trimester of pregnancy and monitored through serial ultrasound examinations. In the available literature, reports on such an early diagnosis of SCT in the first trimester of pregnancy are scarce. It is likely that the early diagnosis was possible because of the external component of the tumour. Most often, SCT is diagnosed in the second trimester.

Correct ultrasound imaging of sacrococcygeal teratoma is crucial for determining the prognosis for both the foetus and newborn. Depending on the morphology of the tumour, its size and type according to AC, the prognosis varies.

Factors indicating a poor prognosis include the solid nature of the lesion, rich vascularisation, and Altman type IV[6]. A good prognosis is suggested by the cystic form and the absence of signs of circulatory failure. The prognosis worsens when the ratio of tumour volume to estimated foetal weight increases. The tumour volume to foetal weight ratio (TFR) helps identify foetuses with a poor prognosis if its value is greater than 0.12 for foetuses before 24 weeks' gestation[11]. In the described case, the TFR ratio was 0.08 at 22 weeks' gestation, and the teratoma presented as a predominantly cystic, poorly vascularised tumour with moderate progression of the lesion during pregnancy. Based on our own experience and data from the literature, with such teratoma features, foetal circulatory failure was unlikely to occur. The prognosis for the foetus and newborn was presented to the parents as favourable, but they were also informed about the potential complications associated with postnatal treatment.

Ultrasound is an excellent diagnostic tool for SCT. The 3D ultrasound technique allows for accurate localisation of the lesion in relation to neighbouring structures and measurements of the tumour. Colour Doppler imaging enables assessment of the intensity of tumour vascularisation, including the size of the vessels supplying the lesion. The use of various ultrasound techniques optimises the monitoring of tumour growth and also allows for the assessment of foetal circulatory capacity. If features of circulatory insufficiency are found, or in cases of richly vascularised tumours with a poor prognosis, intrauterine treatment can be used. In our case, the morphological characteristics and tumor type and size did not result in significant circulatory overload in the foetus. Only in the third trimester of pregnancy was mild cardiomegaly observed, which at this stage of pregnancy did not pose any threat to foetal well-being.

Due to oligohydramnios in the third trimester of pregnancy and the increasingly difficult evaluation of the structures of the small pelvis, along with the steadily increasing size of the tumour, it was decided to perform an MRI scan. It confirmed the diagnosis and determined the clear boundary of the tumour and its relationship to surrounding structures. These results enabled us to rule out infiltration of the spinal canal.

It is recommended that termination of pregnancy in critical SCT cases should be by caesarean section as soon as possible after confirmation of foetal lung maturity (>32 weeks')[3]. In contrast, delivery after 37 weeks' gestation is the most favourable option. In the case of cystic lesions, less than 5 cm, natural childbirth can be considered[12]. In the case described here, the pregnancy was terminated by caesarean section after foetal maturity at 37 + 3 weeks' gestation. The decision was made jointly by obstetric and neonatology teams, taking into account the baby's small size and the extent of the external portion of the tumour.

The case report demonstrates the importance of a multidisciplinary approach to ensure the best therapeutic outcome. A pregnant woman diagnosed with a sacrococcygeal angiosarcoma should be transferred to a level III referral centre for the necessary diagnosis. Proper perinatal care allows for early detection of foetal circulatory insufficiency and informed decision-making about possible intrauterine

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treatment. Safely guiding the patient through the prenatal period and delivery creates optimal conditions for postnatal treatment.

CONCLUSIONS

Diagnosis of teratoma of the sacrococcygeal region is possible using ultrasound as early as the first trimester of pregnancy. However, the sensitivity of the method depends on the type of teratoma. Regular monitoring of lesion growth and vascularisation using modern ultrasound techniques is a crucial aspect of pregnancy surveillance. Due to the risk of developing circulatory failure, regular foetal echocardiography is recommended. Tumour size and type correlate well with foetal and neonatal mortality. However, there is no correlation between the ultrasound image of the tumour and the histopathological result. It is important to remember the differential diagnosis of lesions that may occur in the sacrococcygeal region, and in difficult cases, foetal MRI is indicated. While prenatal diagnosis and intrauterine treatment are very important for reducing the risk of intrauterine foetal death, the essential life-saving treatment is the postnatal surgical resection of the tumour.

Conflict of interest

The authors report no financial or personal relationships with other individuals or organisations that could adversely affect the content of the publication and claim rights to this publication.

Author contributions

All authors contribute equally to the idea and construction of the article.

Ethics

The content presented in the article is in accordance with the principles of the Declaration of Helsinki, European Union directives and uniform requirements for biomedical journals.

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