

Prenatal Diagnosis and Perinatal Management of a Cervical Mass : About a Case of Sialoblastoma

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Original Research Article

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Article History

Received: 02.10.2018

Accepted: 14.10.2018

Published: 30.10.2018

DOI:

10.36347/sjams.2018.v06i10.050



Abstract: Neonatal or congenital tumors concern mainly the fetal soft tissues. Lymphangiomas and sacro-coccygeal teratomas are the most common. Teratomas concern the cervicofacial area in 5 to 15% of cases. We report the case of a cervical mass diagnosed at 22 AS. In prenatal, a left facial teratoma is evoked. An adapted follow-up in coordination with the concerned specialists (pediatricians, ENT surgeons, obstetricians gynecologists). A perinatal surgical excision of the mass rectifies the diagnosis, objectifying a sialoblastoma of the left parotum, at anatomopathological examination.

Keywords: sialoblastoma, malignant salivary gland tumor, antenatal diagnosis, ultrasound.

INTRODUCTION

Sialoblastoma is a tumor that originates from the salivary gland, ductal cells or secretory epithelial cells. It is an aggressive and potentially malignant tumor occurring in the neonatal period. A total of about 40 cases have been reported in the literature.

Malignant tumors of the child's salivary glands are very rare: they represent 8-10% of pediatric tumors of the head and neck. Epithelial tumors are the most common (mucoepidermoid carcinomas of the parotid in particular). Surgery is the treatment of choice for these tumors. Any indication of adjuvant radiotherapy should be discussed. The place of chemotherapy in case of these tumors remains limited and must be discussed in case of an inoperable or metastatic tumor.

Its antenatal diagnosis may be suspected on the imaging data, but its confirmation remains histological.

OBSERVATION

Mrs. Y. B. is 31 years old, G2P2. Her first pregnancy was normal, with full-term delivery by caesarean section due to transverse presentation. There were no known malformations in family history. The current pregnancy had been well followed, no concept of alcoholism, smoking nor taking medication during pregnancy.

The ultrasound of the first trimester was without particularity with a risk of trisomy 21 calculated with 1/12932.

The second trimester ultrasound at 22 weeks of amenorrhoea (WA) showed a slightly vascularized, echogenic and homogeneous mass, well limited to the

level of the left cheek and under the ear measuring 45x44mm, not seeming to invade or deform the maxillofacial structures, nor the thyroid (image1). No other morphological anomaly was identified. Weekly ultrasound examination showed the progressive increase in mass size, with heterogeneous content measuring 68 x 40 x 47 mm and containing vascularized septa and fluid ranges. The growth was well preserved, and the amount of amniotic fluid was normal at 31WA ultrasound (images 2 and 3).

A fetal MRI performed at 28 WA showed an appearance suggestive of a teratoma of the hemiface and left neck without repercussions on the aerial sector. Amniocentesis was performed at 32 WA, revealing a normal 46 XY fetal karyotype.

The last obstetrical ultrasound at 37 WA showing a mass of 82x31x70 mm, little vascularized

with numerous fluid formations and a free aerial branch.

Delivery occurred at 38 WA and two days by caesarean section because of breech presentation and uterus scar, allowing the birth of a newborn male, weighing 3290g (49th percentile), Apgar score 10 /10/10.

Examination of the newborn at birth found permeate choanas, esophagus and anus, no respiratory distress. The ENT examination found a heterogeneous left facial mass of about 9cm in diameter with limited intraoral extension without obstacle on the airways with a neurological examination related to the term. A left facial teratoma without facial palsy is evoked. Breastfeeding was easy.

A preoperative assessment is carried out with a facial MRI which found a clearly defined left temporal

and cervical mass with sharp contours located in front of the ear measuring 67 x 66 x 42 mm, with heterogeneous tissue and fluid signal with a multi-septate appearance, walls and partitions are enhanced after injection. The appearance suggested a cervical teratoma.

Biological assessment was correct

The neonate benefited from an excision of the mass by cervicotomy on day 4 with a good postoperative evolution and the anatomo-pathological examination of the operative specimen was in favor of a sialoblastoma with a complete resection of the tumor.

The couple was informed of the risk of local recurrence and lung metastases. An initial assessment did not find other locations and regular consultations at 2 months and 6 months of life were reassuring.



Image-1 : 2D ultrasound of orbit, 22nd WA, well limited mass with heterogeneous content of 33x20 mm



Image-2 : Stable mass with heterogeneous content 69x50mm, with liquid gaps during ultrasound control of 31st WA



Image-3: 2D ultrasound, Doppler mode, mass still little vascularized at 31st week of amenorrhea

DISCUSSION

Sialoblastoma is a rare entity individualized in 1988 by Taylor. There is about 40 cases described [1, 2]. It's currently classified in malignant epithelial tumors because of the existence of cases with recurrences and metastasis to lymphatics and lungs [3, 4].

It's a congenital tumor, found most often at birth, it appears benign or of low grade of malignancy while remaining potentially aggressive [5]. Parotid localization is three times more common than in the sub-mandibular gland [1]. The other salivary glands do not seem affected. This tumor affects children of both sexes [1]. The newborn and the young child (before 2 years old) only exceptionally present sialoblastoma.

From a clinical point of view, sialoblastoma manifests itself as a more or less large swelling, solid and mobile. The asymptomatic mass has a nonhomogeneous consistency, its size can vary from 1 to 15cm. Multilobulated, the tumor is well circumscribed, unencapsulated, but may have irregular boundaries [6]. The tumor does not show infiltration. The mass can displace the facial nerve, but the paralysis remains exceptional. The growth of this type of tumor is, in most cases, quite fast. It may be accompanied by lymphadenopathy [6].

The ultrasound remains the reference examination, allows to realize in most cases the diagnosis of the cervical masses [7]. Sialoblastoma appears as a solid mass, which has an echogenicity similar to that of the placenta. Doppler shows a vascular morphology of the tumor similar to a lymphangioma. The MRI and CT scan can locate the sialoblastoma and define its extensions; the mass appears iso-intense in 1st trimester [8].

Antenatal diagnosis is essentially based on ultrasound, which may be supplemented with an antenatal MRI. Most often, the diagnosis is made in the

second trimester [9] and sometimes later in the third trimester of pregnancy. The diagnosis of teratoma is often wrongly mentioned in imaging given the similarity of the ultrasound images and is subsequently reversed in histology.

The differential diagnosis arises in front of a mass of the parotid region with the other parotid gland tumors including cystic gland such as cystic lymphangioma, branchial cyst, lipoma, teratoma and arteriovenous malformation. Surgical excision is the treatment of choice [10]. It must be complete to prevent recurrence. In parotid localization, exo-facial parotidectomy is performed if the tumor occupies the superficial lobe, it will be total if the tumor sits at the level of the deep lobe. Radio chemotherapy may be necessary and clinical monitoring is essential looking for recurrence or metastases [11]. The prognosis is closely related to the histological nature.

Histologically it is presented in the form of solid islands of basaloid cells with round nucleus and eosinophilic cytoplasm poorly limited and tubular structures, separated by a fibrous stroma or fibro myxoid [2]. The histological appearance of sialoblastoma is similar to the early stages of salivary gland development [5].

The diagnosis of these tumors is based on ultrasound [7]. It is essentially the size of the tumor, its evolutionary potential, its proximity to the aerodigestive junction, the amount of amniotic fluid and any associated malformations that condition the prognosis. A discussion in a multidisciplinary center of antenatal diagnosis will make it possible to program the various investigations necessary to the diagnosis. Establishing the surgical therapeutic possibilities at the birth is difficult to evaluate in antenatal. The decision to, whether or not, continue the pregnancy is up to the parents, who must be well informed about the future of the child who will be born. The care should take place in a reference center, able to perform all the necessary

examinations. The birth must be programmed in a level 3 maternity, in the presence of a multidisciplinary team including pediatrician and surgeon resuscitation [9].

There is no consensus about the delivery route. It will depend on the tumor size, which can lead to dystocia. The vaginal delivery is possible for small tumors. In our case, the volume of the tumor and its location were not compatible with a vaginal delivery.

At birth, care is multidisciplinary, provided by pediatric resuscitation teams and specialized surgeons whose presence in the delivery room increases the chances of these newborns in terms of neonatal mortality and morbidity [7]. The treatment is based on excision surgery which must be performed as soon as possible [9]. Imaging, MRI and computed tomography (CT) examinations, which prepare the surgical procedure, improve the prognosis. The University of Hamburg has described only three cases out of a total of 6,646 tumors of the salivary gland since the years 1965 to 1994 [12].

Differential diagnoses are often evoked, particularly teratomas, classically of very variable size (a few centimeters to 20-25 cm). At the cervical level, they are located laterally, or in axial position at the level of the oropharynx. Their echo structure is polymorphic and variable in time by an increase in volume and by the appearance of intra-tumoral hemorrhages. They have a mixed echogenicity associating a quite large heterogeneous echogenic mass and sparse fluid ranges. Pure cystic or solid forms are possible. The proliferation of immature tissue or embryonic tissue characterizes malignant forms [9]. This malignancy is manifested by local aggression and by the occurrence of distant metastases [13]. Given the tissue heterogeneity of these tumors, it is impossible to assert the absence of immature tissue before postnatal exeresis and anatomopathological examination. It is essentially the size of the lesion, its depth extension and the proximity of the aerodigestive junction that condition the pejorative character of the tumor. A bulky tumor mass, evolutive from one examination to another, the presence of a hydramnios (by paralysis of the pharyngeal muscles or by mechanical obstruction), the absence of visualization of the stomach, the demonstration of deep extensions are elements of poor prognosis [14].

All these elements explain the difficulty of the diagnosis, the similarity in the prenatal diagnosis procedures and the indication of a complete resection if possible of the mass. Anatomopathological examination, in our case, finally established the diagnosis of sialoblastoma of the left parotid without invasion of nearby tissue.

CONCLUSION

The ultrasound discovery of a cervico-facial mass in a fetus poses a diagnostic and prognostic

problem. Indeed, if the detection of the tumor mass seems easy during an ultrasound examination, the etiological diagnosis, meanwhile, is often not univocal. The histological type of these tumors remains hypothetical. Their evolutionary potential during pregnancy is random. Possible surgical options at birth and their sequelae are difficult to assess. The parotid localization of the cervical masses is exceptional. The positive diagnosis is suspected on imaging and confirmed by histological examination.

Delivery should be at a Level 3 maternity, with the presence of a multidisciplinary team, pediatric resuscitation teams and specialized surgeons. The treatment is based on surgery. The excision should be complete to prevent recurrence. Rigorous and prolonged surveillance is recommended to detect early recurrence.

The prognosis is quite favorable, and the death remains exceptional.

REFERENCES

1. Shan XF, Cai ZG, Zhang JG, Zhang J, Gao Y, Yu GY. Management of sialoblastoma with surgery and brachytherapy. *Pediatric blood & cancer*. 2010 Dec 15;55(7):1427-30.
2. Aggarwal R, Bhalla S, Chopra P, Kulshreshtha R. Congenital sialoblastoma: A case report and review of literature. *Journal of Orofacial Sciences*. 2013 Jul 1;5(2):135.
3. Huang R, Jaffer S. Imprint cytology of metastatic sialoblastoma. *Acta cytologica*. 2003;47(6):1123-6.
4. Williams SB, Ellis GL, Warnock GR. Sialoblastoma: a clinicopathologic and immunohistochemical study of 7 cases. *Annals of diagnostic pathology*. 2006 Dec 1;10(6):320-6.
5. Ellis GL. What's New in the AFIP Fascicle on Salivary Gland Tumors: A Few Highlights from the 4 th Series Atlas. *Head and neck pathology*. 2009 Sep 1;3(3):225-30.
6. Biraud Guillaume, Les pathologie salivaires : donnés actuelles .Thèse pour le diplôme d'état en chirurgie dentaire. 2009.
7. Azizkhan RG, Caty MG. Teratomas in childhood. *Curr Opin Pediatr*.1996;8(3):287-92.
8. A. Aubina S. Pondavena. Oropharyngeal teratomas in newborns: Management and outcome
9. Benouaiche L, Couly G, Michel B, Devauchelle B. Diagnosis and management of cervicofacial congenital teratomas: about 4 cases, literature review and restatement. In *Annales de chirurgie plastique et esthetique* 2007 Apr (Vol. 52, No. 2, pp. 114-123).
1. Yekeler E, Dursun M, Gun F, Kilincaslan H, Ucar A, Genchellac H, Acunas G. Sialoblastoma: MRI findings. *Pediatric radiology*. 2004 Dec 1;34(12):1005-7.
10. Brandwein M, Al-Naeif NS, Manwani D, Som P, Goldfeder L, Rothschild M, Granowetter L.

Sialoblastoma:
clinicopathological/immunohistochemical study.
The American journal of surgical pathology. 1999
Mar 1;23(3):342-8.

11. Seifert G, Donath K. Juvenile pleomorphic parotid adenoma of embryonal structure. *Der Pathologe*. 1998 Jul;19(4):286-91.
12. Grisey A, Roth P, Martin A, Czorny A, Riehl-Duvinage C, Maillet R, Schaal JP. Prenatal

diagnosis and management of a case of rapidly involuting congenital hemangioma. Review of literature. *Journal de gynécologie, obstétrique et biologie de la reproduction*. 2006 Jun;35(4):405-10.

13. Kdous M, Hachicha R, Risk E, Sauvanet E. Foetal rhinopharyngeal teratoma: antenatal diagnosis and prognosis. Case report. *Gynécologie, obstétrique & fertilité*. 2006 May;34(5):403-6.