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Letters to the Editor - Brief Communications

Amniotic bands and associated polymicrogyria: In favor of a unique ischemic cause

Dear Editor,

We present a case of multiple amniotic bands with spontaneous release of the fetus during pregnancy and, at birth, foot malposition and skin scars. Additional occurrence of neuro-developmental disorders associated with polymicrogyria led to the hypothesis of common vascular etiology.

The patient was referred to our center for fetal immobility and suspicion of amniotic band syndrome. Ultrasound at 14 weeks of gestation (WG) confirmed immobility of the fetus, with permament hyperflexion of the limbs, and permanent rotation of the fetal head in an unusual axis. Multiple linear bands surrounding the fetus were visualized vaginally. Anatomical study proved to be very difficult. A trophoblast biopsy was performed, resulting in normal karyotype. The parents were informed of possible ischemic lesions. They requested termination of the pregnancy, which was deemed admissible by our center. Secondarily, they wished to continue with the pregnancy.

The following ultrasound examinations made it possible to objectify the presence of movements of the lower limbs, and then movements of the trunk and of the upper limbs. Cutaneous furrows facing the amniotic bands were noted. Malpositioned fetal foot was suspected with no associated abnormalities, especially at the cerebral level.

At 34 + 4 WG, the patient gave birth by spontaneous vaginal delivery to a boy weighing 2460 g and 44 cm; cranial perimeter to 30 cm (>−2 SD); Apgar 10/10.

Examination of the newborn showed malpositioned feet, a band on the left ankle remaining amniotic band circling the left ankle and, at the left shoulder level, striations that might correspond to scar lesions secondary to the amniotic bands (Fig. 1).

Macroscopic examination of the placenta was in favor of an extra-amniotic pregnancy. Indeed, extra-amniotic pregnancy occurs following a rupture of the amnion and its retraction around the placental insertion of the cord, as was observed in our case with the fetus located in the extra-embryonic coelom, between the two membranes.

At the age of 8 months, the child developed drug-resistant epilepsy. Brain magnetic resonance imaging showed diffuse polymicrogyria. Follow-up for developmental delay was put in place. Genetic assessment has been negative up to now (CGH Array, search for Congenital Disorder of Glycosylation (CDG) syndrome and fragile X syndrome).

This case allows us to discuss the mechanism of amniotic bands, which is still a matter of debate in the literature between the exogenous and endogenous theories [1]. According to the exogenous theory [2], the triggering event would be an early rupture of the amnion, with leakage of amniotic fluid between the amnion and the chorion. The outer surface of the amnion would produce amniotic bands. According to the endogenous theory, abnormal development of the embryonic disk would cause malformations and amniotic bands. In a study on 25 fetuses with limb-body wall complex, Van Allen found persistent extra-embryonic coelom in 85% [3]. For him, early vascular disturbances are at the origin of ischemic fetal lesions.

Polymicrogyria is a malformation of cortical development, defined by gyri too numerous and too small. Early ischemic events can cause polymicrogyria [4]. In the literature, one case has been described associating amniotic band syndrome and polymicrogyria [5].

To conclude, we presented a case of multiple amniotic bands leading to fetal immobility in the first trimester, with spontaneous release of the fetus. Polymicrogyria might have ischemic causes. The endogenous theory would explain the association of amniotic band syndrome with polymicrogyria. In case of continued pregnancy and in the absence of major malformation, systematic MRI of the fetal brain should be proposed in case of amniotic band syndrome, in search for ischemic lesions.

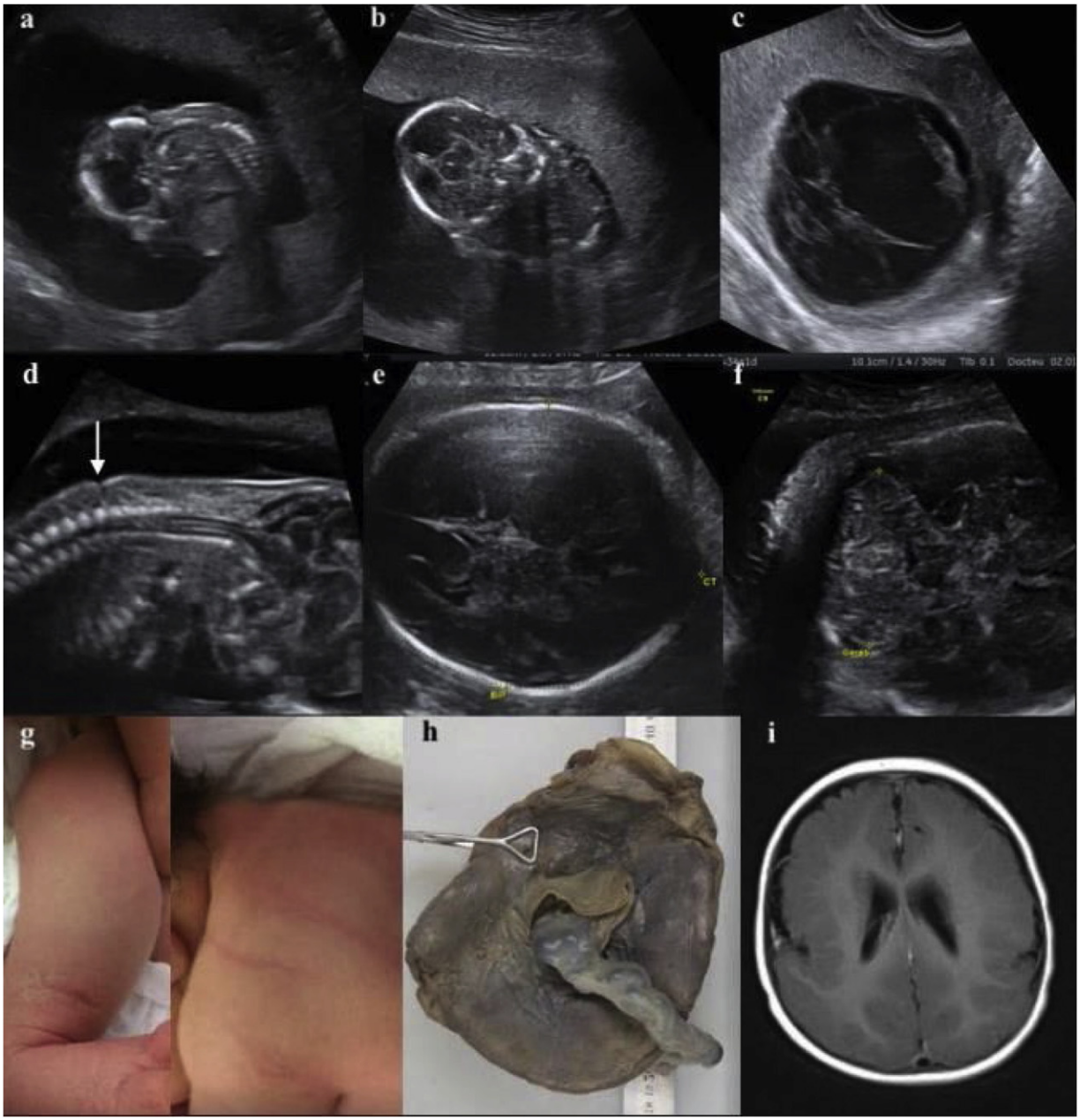

Fig. 1. a–c : 14 Weeks of Gestation (WG) : limbs in a state of hyperflexion, permanent rotation of the fetal head in an unusual axis, multiple linear bands surrounding the fetus visualized vaginally; Figure d : 18 WG, suspect dorsal cutaneous furrow; Figures e–f : 29 WG: fetal brain imaging seems normal; Figure g: striations at the left ankle and left shoulder; Figure h: macroscopic examination of the placenta; Figure i: Magnetic Resonance Imaging at the age of 8 months: polymicrogyria.

References

[1] Moerman P, Fryns JP, Vandenberghe K, Lauweryns JM. Constrictive amniotic bands, amniotic adhesions, and limb-body wall complex: discrete disruption sequences with pathogenetic overlap. Am J Med Genet 1992;42:470–9.
[2] Torpin R. Amniochorionic mesoblastic fibrous strings and amnionic bands: associated constricting fetal malformations or fetal death. Am J Obstet Gynecol 1965;1(91)65–75 Jan.
[3] Van Allen MI, Curry C, Walden CE, Gallagher L, Patten RM. Limb-body wall complex: II. Limb and spine defects. Am J Med Genet 1987;28:549–65.
[4] Squier W, Jansen A. Polymicrogyria: pathology, fetal origins and mechanisms. Acta Neuropathol Commun 2014;2:80.
[5] Yamanouchi H, Ota T, Imataka G, Hagiwara Y, Nakagawa E, Eguchi M. Congenital bilateral perisylvian syndrome associated with congenital constriction band syndrome. J Child Neurol 2002;17:448–50.

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A case of gastric-type mucinous endocervical adenocarcinoma in presence of nabothian cysts

Dear Editor,

A group of benign and malignant endocervical glandular lesions unrelated to human papillomavirus (HPV) and showing gastric differentiation, has been described in the last years [1]. Minimal deviation adenocarcinoma (MDA) and lobular endocervical glandular hyperplasia (LEGH) exhibit a gastric phenotype and immunophenotype [2]. Gastric-type adenocarcinoma (GAS) has been recently described as a subtype of cervical adenocarcinoma [3].These neoplasms have aggressive clinical behavior. There is a likely LEGH-GAS sequence, and an absence of HPV in cases of LEGH, and MDA suggested that this sequence represents an HPV-independent pathway of carcinogenesis [1]. Morphologically defined GAS exhibits immunoreactivity for MUC6 and/or HIK1083, representative markers for pyloric gland differentiation. Notably, GAS is negative for p16INK4a, a marker for high-risk HPV-driven neoplasm [4]. Clinical manifestations include watery vaginal discharge, although women can be asymptomatic [5]. Surgery remains the main treatment for patients with early-stage lesions. Radiotherapy and/or chemotherapy are recommended for women with advanced disease.We described a case of gastric-type mucinous endocervical adenocarcinoma in a woman with positive Pap smear tests for atypical glandular cells of undetermined significance (AGUS) but negative HPV-DNA test. A 46 years old nulliparous woman gave a medical examination for hydrorrhea. The gynecological examination showed plentiful mucinous secretions from vagina with vaginal fornix preserved. The specular examination showed an eroded and enlarged cervix. The transvaginal ultrasound (US) scan showed multiple Naboth cysts, as well as an uneven appearance and a plentiful vascularization of the uterus, but regular ovaries. The last two Pap smear tests, performed six months and one year before respectively, showed the presence of atypical glandular cells of undetermined significance (AGUS), so patient underwent colposcopy that confirmed the eroded and enlarged aspect of cervix but turned out to be negative for HPV-related lesions. After 3 months, the patient redid Pap smear test which showed the persistence of AGUS, whereas HPV-DNA test was negative. For this reason, the woman underwent a cervical conization: the histologic examination of cervical sample, measuring $4 \times 4 \times 2.5$ cm, showed a mucinous adenocarcinoma (NOS) moderately differentiated (G2) infiltrating endocervical margin and part of squamous cervical epithelium tissue, with vascular infiltration. Her cancer markers (α-FP, CEA, CA 19-9, CA 15-3, CA 125) turned out to be all negative. Abdomen and Pelvic MRI showed a tumor that invaded the cervix for all its thickness without involvement of nearby lymph nodes. The patient was referred to our institution and underwent a laparotomy procedure. Uterus and both annexes appeared macroscopically regular. She underwent a total hysterectomy, with bilateral salpingo-oophorectomy and pelvic and obturator lymphadenectomy. Definitive pathologic examination showed a mucinous adenocarcinoma gastric-type, moderately differentiated (G2), infiltrating cervical canal for all its thickness, with a microscopic metastasis in a lymph node isolated in the right parametrium and in two of pelvic and obturator lymph nodes (pT1B1N1) (Fig. 1A–B). Immunohistochemical analysis turned out to be negative for estrogen receptor (ER) and progesterone receptor (PR) (Fig. 1C) and positive for MUC6 (Fig. 1D). The patient is currently being treated with pelvic EBRT (external beam radiation therapy) and concurrent chemotherapy with cisplatin. The diagnosis of GAS is established primarily based on morphology: tumor cells with abundant eosinophilic cytoplasm, distinct cell borders, and a greater degree of cytological atypia characterize it [2,4]. However, immunohistochemistry may contribute to the diagnosis. GAS shows the gastric phenotype, as demonstrated by HIK1083, MUC6, or carbonic anhydrase type IX staining, negativity for p16, as well as a frequent mutant pattern of p53 staining [2]. Unlike MDA and LEGH, the MRI findings of GAS have not been clearly described yet, although image analysis based on T2-WI have recently showed a tumor shape classified as type II, infiltrative [5]. GAS is associated with aggressive behavior and a poor prognosis, including a possible propensity for peritoneal and adnexal dissemination [5]. Indeed, since the first description of GAS in 2007, it has been reported its poorer outcomes than those of usual-type adenocarcinomas (UEA), tipically associated to high-risk HPV in over 90% of cases. GAS is rather common in Japan, accounting for up to 20%–25% of all endocervical adenocarcinomas while it is considered rare in Western countries [2]. Kojima et al. showed that GAS had a 5-year disease-specific survival of 30% compared to 77% for UEA [5]. The mean age of presentation ranges between 45–48 years. It's related to high rate of lymphovascular invasion (LVI) and regional lymph node metastases [2].Here, we discussed the diagnostic difficulties for endocervical glandular lesions with gastric differentiation and the possible relationship with the