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Multisurgical approach for recurrent fetus-in-fetu of the skull

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ABSTRACT

Foetus in fetu is a rare tumor of the child. CT and/or MRI suggest this diagnosis. Differential diagnosis with mature multicellular teratoma can be difficult. Here we reported the case of a girl born at 37 weeks, that presented a voluminous mastoïd mass on the left side treatment by initially by surgery. Until age of 9 years, she developed a tumor recurrence localized at the skull base near the left cerebello ponting angle requiring a neurosurgical approach with a good following. The pathology was finally a teratoma. Total removal must be a goal of the initial surgical treatment but a long term follow up is mandatory to avoid recurrence.

1. Introduction

Foetus in fetu (FIF) is a rare abnormality characterized by the incorporation of a foetus within another foetus. The incorporated foetus develops inside the other one but its growth is abnormal, resulting in a "pseudo-tumoral" mass. The incidence is estimated of around 1/500,000 births [1–3]. The diagnosis can be considered before or confirmed after birth. Imaging (Sonography, CT and MRI) allows the differentiation between FIF and teratoma [4]. The foetus in fetu is characterized by an axial skeleton with metameric segmentation. This mass develops mostly in the abdominal cavity (70%) [5]. There is frequently a main feeder artery allowing cellular multiplication of the foetus in fetu. The foetus in fetu is generally surrounded by an amniotic pouch [6]. Its removal has to be macroscopically complete and a long-term follow-up is necessary. This tumor can relapse under the form of a mature multicellular teratoma as in our case.

2. Case report

The child, a girl born at 37 weeks, presented a voluminous mastoïd mass on the left side measuring 12 × 6.5 cm, pushing back and deforming the left ear and going down into the left cervical region (Fig. 1A). Projecting from its surface were growths consistent in appearance with foetal limbs. Neurological examination revealed a left facial palsy. The child was referred in the pediatric chirurgical department and was managed by pediatric surgeons, plastic surgeons and neurosurgeons.

Standard blood analyses (blood count, biochemistry) were normal. Blood tumor markers were normal for this age, alfa foeto-protein was at 34,644 UI/ml (normal rate lower than 12,000 UI/ml) [7] and BHCG were at 0.1 ng/ml (raised compared to the adult).

X-rays of the skull showed the extrinsic development of the mass and ossification forming an hypoplastic diaphysis. Sonography showed its solid density with a well-organized vascularization via a main pedicle. CT and MRI confirmed the presence of a fatty and bony mass developed inside the left petrous bone (Fig. 1B, C, D, E, F & G). The left internal jugular vein seemed flattened.

Surgical removal of the mass was performed at 15th of life, without skin graft (Fig. 1H). Pathological examination revealed a FIF, including bony structures organized (follicular appendixes, sweat glands and pilosebaceous glands, glandular formation, various types of epithelium of bronchial and digestive aspect, immature nerve tissue, choroid plexi, areas of hepatocytes, streked pancreatic, muscular tissue, cartilaginous nodules).

Dissection of the mass was possible using a plan of cleavage but not without difficulties. A microscopic analysis of the limits of resection showed that they were not in healthy margins. At this time, two diagnosis were still possible: mature multi-tissular cervical teratoma or FIF. After a multidisciplinary discussion, the diagnosis of FIF was essentially privileged because of the relatively well-organized aspect with foetal limbs, the presence of a main vascular pedicle as well as the existence of a plan of cleavage.

The first CT post-operation showed a remodeling abnormality of the left petrous bone (Fig. 1I and J). The outer ear had a normal aspect but aeration of the middle ear was poor, with an absence of external auditory canal. There was also a bony deformity with fatty tissue. The

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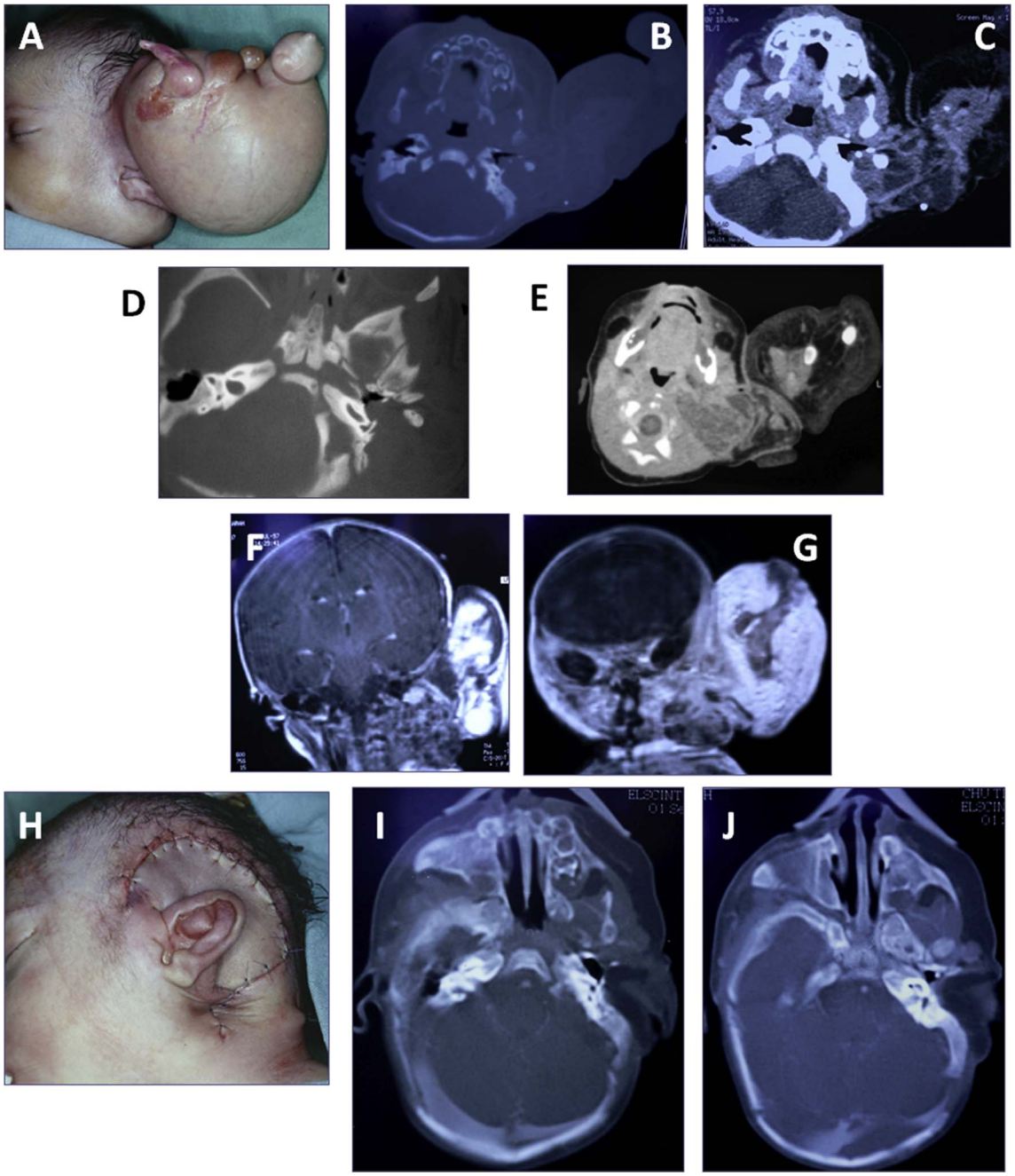

Fig. 1. (A) Picture of the FIF before surgery. Fatty and bony mass developed inside the left petrous bone. CT axial view, bony window **(B & D)**, and parenchymal view **(C & E)**. Coronal views **(F & G)** on MRI, T1 with gadolinium. **(H)** Picture after surgery. Control CT, axial views, bony window **(I & J)**.

child kept the left facial palsy.

The patient was followed-up until age of 9 years. She presented an advanced puberty with metrorragia and mammary development. Endocrinological evaluation showed an increasing of growth. Blood tumor markers were still normals. Hence, she received a breaking treatment. MR ruled out the pituitary origin of the hormonal disturbance and showed a tumor recurrence at the skull base near the left cerebello-pontine angle (Fig. 2A and B). The child was referred to pediatric neurosurgery department for a complete resection of the recurrent tumor (Fig. 2C and D). The mass did not invade the dura.

Pathological examination revealed a mature teratoma containing largely cystic structures lined with bronchial epithelium (brown fat, mature cartilage, smooth muscular fibers, and collagen tissue). MR at 4 years post-resection showed no recurrence.

3. Discussion

Foetus in fetu is an exceptional abnormality which arises in 89% of cases in infants less than 18 months [8]. The FIF is an inclusion of a parasite foetus inside a host foetus. According to the “twin theory”, this abnormality could result from a disorder during embryogenesis of a monozygot monochorial biamniotic pregnancy. One or several abnormal foeti developed in a foetus which pursued its normal development. This pathological process could be a consequence of an abnormal anastomosis of the vitellin circulation [9]. In contrast, according to the “teratoma theory”, FIF could be a particular form of highly organized and well differentiated teratoma [10]. FIF could also behave as a mass containing a vertebral axis around of which develop other tissues or organs [11]. In contrast, according to her (it) “teratoma theory”, the FIF would be the particular shape of teratome highly organized and well