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CASE-BASED UPDATE

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Prenatal diagnosis of a craniopharyngioma: a new case with radical surgery and review

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Abstract *Case report:* A case of the antenatal diagnosis of a craniopharyngioma with radical surgery in the neonatal period is reported. *Review of the literature:* We have reviewed the literature of such cases in an attempt to isolate specific features in this age group and to determine the appropriate management. Only six cases of the truly antenatal diagnosis of craniopharyngiomas have been reported. Diagnosis has resulted from routine ultrasound during pregnancy or from polyhydramnios. Clinically, there is often macrocephaly due to hydrocephalus or a significant-sized tumor. *Conclusions:* Management of these rare cases is controversial with high postoperative mortality and significant morbidity, including panhypopituitarism, visual disturbance, and neuropsychological disorders. From the available literature, no conclusions concerning the management can be drawn at present, due to the rarity of early surgical intervention. Our case, despite the lack of important follow-up, seems to confirm the possibility of attempting radical surgery in the neonatal period as a result of advances both in surgical techniques and in neonatal intensive care.

Keywords Intracranial tumors · Antenatal diagnosis · Craniopharyngioma

Introduction

The prenatal diagnosis of intracranial tumors is extremely rare. Teratomas, carrying a very poor prognosis, are the most frequent lesion diagnosed in this setting. Although craniopharyngiomas account for 9% of all intracranial tumors in children, only six cases of antenatal craniopharyngiomas have been reported [1, 3, 12, 14, 16, 18]. Management of these lesions has been controversial in children [7, 8] and is especially so with an antenatal diagnosis. Adding one case of surgical treatment in a neonate with prenatal diagnosis, we have reviewed the literature since the first neonatal case published, which was in 1952 by Iyer [10]. We aim to discuss the management and results of the treatment of craniopharyngiomas diagnosed during pregnancy, excluding those where diagnosis was during labor or at birth.

Case report

A 24-year-old gravida was referred for routine prenatal ultrasound at 29 weeks' gestation. Examination revealed a large echogenic suprasellar mass measuring 32×23×22 mm^3. A prenatal MRI, performed at 31 weeks, confirmed the suprasellar lesion containing a solid component and no hydrocephalus (Fig. 1). The diagnosis of craniopharyngioma was suspected. The parents decided to continue with the pregnancy. The prenatal course was uncomplicated until the premature delivery at 34 weeks' gestation. A 2,600-g boy was delivered. The neonate had normal reactivity with a flat and soft fontanel and a head circumference of 31.5 cm. Visual examination revealed pupils equal and symmetrically reactive to light. Endocrinologic evaluation showed hypothyroidism. A new MRI was performed, which showed an intrasellar and suprasellar tumor, extending into the third ventricle. The lesion was isointense on the T1-weighted image and hyperintense on T2 with gadolinium enhancement (Figs. 2, 3). Due to the lack of intracranial hypertension we waited for 40 days before performing surgery.

A right pterional craniotomy was performed with a subfrontal approach and an extensive opening of the sylvian fissure. The tumor was progressively removed with conservation of the basal

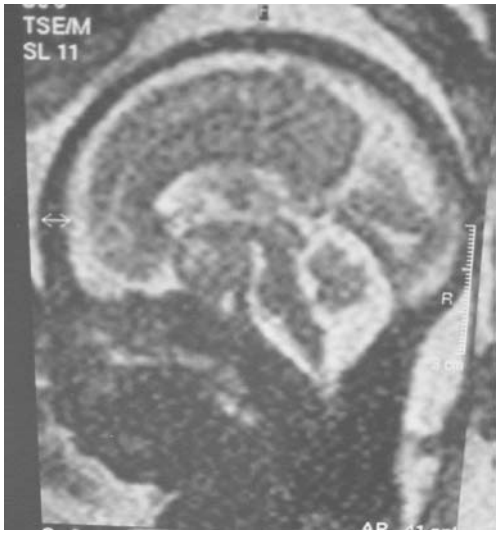

Fig. 1 Prenatal MRI at 31 weeks: sagittal T1-weighted image demonstrating an homogenous suprasellar mass

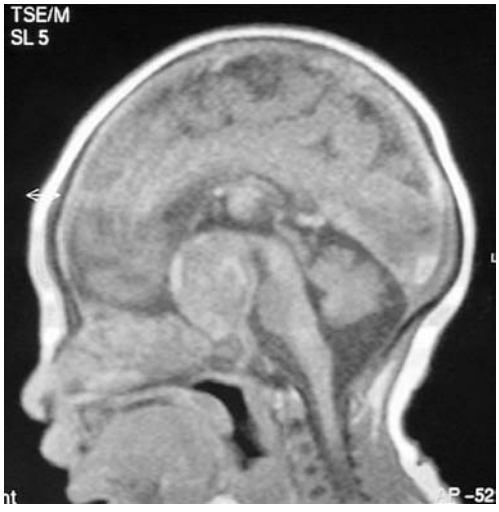

Fig. 2 Postnatal MRI at 3 days: sagittal T1-weighted image revealing an intrasellar portion of the craniopharyngioma

vessels. At the posterior part, the pituitary stalk was not preserved. The removal of the tumor was considered to be total.

The tumor specimen proved to be a craniopharyngioma. The postoperative course was uneventful. Polyuria was controlled by administration of a vasopressin analogue and hormonal deficits were corrected by replacement. The visual acuity could not be measured, but the boy seemed to have a slight visual disturbance. Postoperative MRI confirmed the total removal of the tumor. The neonate was discharged 15 days after surgery with the following treatment: hydrocortisone 2 mg/day, desmopressin 0.4 μg/day and L-thyroxin 25 μg/day. One year later, the boy was in a good condition, with a substitutive treatment and no recurrence on the last MRI.

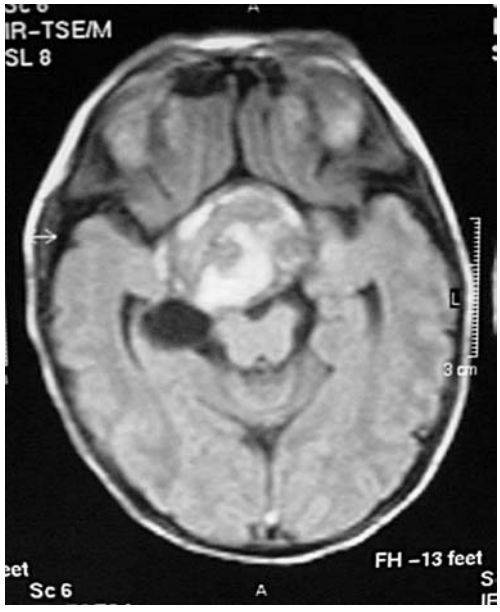

Fig. 3 Postnatal MRI at 40 days: axial T2-weighted image showing slight progression of the lesion compared with the previous MRI, with hypersignal and hyposignal and a posterior cystic component

Discussion

Intracranial tumors in neonates are rare, representing between 0.5 and 1.9 of all brain tumors in children [11]. The prenatal diagnosis of intracranial tumors remains exceptional and is usually of teratomas and of neuroepithelial and mesenchymal tumors.

Craniopharyngiomas are thought to arise from embryonic squamous-cell remains of an incompletely involuted ectodermic hypophyseal pharyngeal duct or Rathke's pouch. This structure, extending from the sella to the pharynx, is at the origin of the adenohypophysis.

The lower part of Rathke's pouch is gradually obliterated and its epithelial lining is converted into a cord of cells or several islands of squamous epithelium, along with the rest of the epithelium of the buccal cavity.

We describe the management of six previously reported cases (Table 1) in which a diagnosis of craniopharyngioma was made during pregnancy, by a routine ultrasound in 4 cases and in 2 cases because of polyhydramnios. One of them resulted in a preterm labor [16]. In more recent cases, as in the present case, a prenatal MRI was performed [3, 12], allowing better definition of the tumor.

Delivery was made either spontaneously [1, 14] or by cesarean section [3, 12, 16]. Increasing hydrocephalus throughout the pregnancy was present in only one case [3]. In the presence of hydrocephalus, cesarean section is indicated and in cases of progressive enlargement of the ventricles an early delivery, at the time of pulmonary maturity, may be considered. Some cases have been di-

Table 1 Recorded cases of the prenatal diagnosis of craniopharyngiomas

Reference	Time and reason of diagnosis	Perinatal management	Clinical course	Postnatal exploration	Treatment	Evolution
[16]	Antenatal 34 weeks, polyhydramnios, ultrasonography	Cesarean section	Macrocephaly	CT scan		Died at 3 days
[3]	Antenatal 27 weeks, polyhydramnios, ultrasonography, antenatal MRI	Cesarean section 36 weeks	Macrocephaly, hydrocephalus	CT scan	CT-guided needle biopsy, ventriculoperitoneal shunt, refused surgery	Died at 8 weeks
[12]	Antenatal 29 weeks, routine ultrasound, antenatal MRI	Cesarean section	Macrocephaly	CT scan, MRI	Surgery at 4 weeks	Died during surgery
[14]	Antenatal 28 weeks, routine ultrasound	Delivered spontaneously at 38 weeks	Head circumference, normal, no endocrine disturbance	CT scan	Surgery at 17 days	Recurrence, reoperation at 14 months, follow-up 8 years, hemiparetic visual acuity 2/10
[18]	Antenatal 20 weeks, routine ultrasound	Pregnancy termination (21 weeks)				
[1]	Antenatal 33 weeks, routine ultrasound	Delivered spontaneously at 40 weeks	Head circumference normal, normal basal values of all the pituitary hormones	CT scan, MRI	Surgery at 9 months	Follow-up 5 years, diabetus insipidus treated subduroperitoneal shunt, substitutive hormonal therapy
Our case 4report (2003)	Antenatal 29 weeks, routine ultrasound, antenatal MRI	Delivered spontaneously at 34 weeks	Head circumference normal	MRI	Surgery at 40 days	Follow-up 1 year, slight visual disturbance, substitutive hormonal therapy

agnosed just before birth [6], as in the first reported case [10] in which the diagnosis was made by X-rays revealing a very large head without calcifications.

A postnatal MRI must be performed to evaluate the extent of the lesion and its relation to adjacent structures. These tumors are always large in size with a possibility of further growth causing hydrocephalus [3]. They may have a suprasellar or an intrasellar and suprasellar location. Radiologically, there are no specific features of these antenatal lesions; a cystic component of the lesion has been noted in three cases [3, 12, 16] and calcifications in all. Contrast enhancement is precise in only two cases [1, 14].

Preoperative investigations must include visual evaluation, such as is feasible at this age, and assays of electrolyte and hormone levels. Electrolyte levels have only been described in two cases [1, 14], being normal with no signs of diabetes insipidus.

Of the 6 previous cases, the parents decided on pregnancy termination in 1 case [18] and refused surgery in another [3], apart from the placement of a ventriculoperitoneal shunt, and the child died at 8 weeks of age. Another child succumbed on the 3rd day of life with a huge, partially cystic holocerebral mass [16], and the other 3 were operated on. One child [12] died of cardiac arrest during surgical resection at 4 weeks of age. The second one [14] was operated on at 17 days, with no preservation of the pituitary stalk. In this second case, post-operative MRI at 5 months revealed recurrence of the tumor and infarction of the right medial cerebral artery with a clinical hemiparesis. This recurrence was treated by further surgery with total removal of the tumor. At 8 years of age, the boy was hemiparetic, but able to walk without support, with a 2/10 visual acuity and ongoing hormone replacement. The third one [1], which is the most recent case, was operated on at 9 months, despite a huge tumor, with total removal. No recurrence has been detected. At 5 years of age, the girl was in good condition and almost normal mentally and physically, receiving replacement therapy of pituitary hormones.

Including those cases of craniopharyngioma diagnosed neonatally [2, 4–6, 9, 10, 15, 17, 19, 20], we observed that the results of surgery for craniopharyngiomas in the neonate are extremely poor, with significant mortality, even during surgery. Morbidity is also significant, including panhypopituitarism, visual disturbance with decreased visual acuity, and psychological disorders. Nevertheless, surgery remains the treatment of choice, especially in children to prevent a recurrence, since radiation therapy is extremely deleterious. The recent neonatal cases [1, 14], including our own, despite only a very short follow-up, demonstrate that in good conditions it is possible to cure the tumor with early surgery. Electrolyte disorders in the postoperative course at this age seem easier to compensate than in the older age group, in our experience. Substitutive hormonal therapy is now well

established [13]. The main problem remains visual acuity due to the considerable size of these lesions, causing a compression of the visual pathways whether the location of the tumor is prechiasmatic or retrochiasmatic.

Conclusions

The antenatal diagnosis of craniopharyngioma is exceptional. Reviewing the literature of antenatal and neonatal cases demonstrates that the results of surgery are poor, considering the mortality and morbidity rates. Indeed, these lesions are usually huge, causing a compression of the optic pathways and making preservation of the pituitary stalk difficult.

Termination of the pregnancy may be suggested in cases of early diagnosis, especially with associated progressive hydrocephalus. Nevertheless, surgery on such lesions in neonates can be feasible in good conditions and may be associated with only minor sequelae.

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