

case report

Lhermitte-Duclos disease and pregnancy

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Background. Lhermitte-Duclos disease or dysplastic gangliocytoma of the cerebellum is a rare disorder that can cause progressive mass effects to the structures occupying posterior fossa. Magnetic resonance imaging is a diagnostic modality of choice demonstrating characteristic non-enhancing gyriform pattern with the enlargement of cerebellar folia, hypointense on T1 and hyperintense on T2 weighted magnetic resonance images.

Case report. The authors present a case of 37-year old woman with previously unknown Lhermitte-Duclos disease in the third trimester of pregnancy from the first signs of the disease to the first six months after delivery.

Conclusions. More experience will be needed with this disease in pregnancy and post delivery period to recommend pregnancy for women with such condition. However, this case shows that a pregnant woman with Lhermitte-Duclos disease could reach full-term pregnancy and deliver a healthy child, without life-threatening risk.

Key words: cerebellar neoplasms; ganglioneuroma

Introduction

Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease) is a rare disorder, characterized by a slowly progressive unilateral neoplastic mass of the cerebellar cortex. The histopathological findings of Lhermitte-Duclos disease (LDD) include the widening of

the molecular layer, which is occupied by abnormal ganglion cells, absence of the Purkinje cell layer and hypertrophy of the granular cell layer. Magnetic resonance imaging (MRI) is a diagnostic modality of choice and reveals a characteristic non-enhancing gyriform pattern with the enlargement of cerebellar folia. The lesion is hypointense on T1- and hyperintense on T2-weighted magnetic resonance images.¹ In patients with a posterior fossa tumour suggestive of a dysplastic gangliocytoma on neuroimaging studies, a pathologic confirmation is necessary.²

Dysplastic cerebellar gangliocytoma is commonly associated with the progressive mass effect in the posterior fossa and is typi-

Received 5 January 2006

Accepted 15 February 2006

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cally presented with headaches, cerebellar dysfunction, occlusive hydrocephalus and cranial nerve palsies. The disease usually manifests in young adults, but the age at presentation ranges from birth to the sixth decade. There is no sex predilection. The therapy consists of decompression of the posterior fossa by a total surgical removal of the tumour mass.³ A problem of surgical removal of these tumours is to miss the borderline between tumour and healthy cerebellum tissue so that the incomplete removal of the tumour is not rare.⁴

To our knowledge there were no reports of LDD in pregnancy. In our case report we intend to present the potential influence of LDD on pregnancy and delivery.

Case report

A 37-year-old woman was admitted at the Clinic of Gynaecology and Obstetrics in the 27th week of her first pregnancy for monitoring and programming the childbirth. Hospitalisation and programmed delivery by caesarean section has been recommended from her neurosurgeon before the control MRI was performed, because she had a history of partial cerebellar tumour resection. Namely, 19 years ago, CT was performed due to the cerebellar dysfunction and the increased intracranial pressure (intensive headaches, nausea, dizziness and optical nerve oedema). CT had shown a large mass of the right cerebellar hemisphere suspicious of gliomal tumour and she underwent the neurosurgical extirpation. Only a partial resection was performed with the implantation of ventriculoatrial shunt. The histopathological findings included a widening of cerebellar cortex due to hypercellular granular layer without a clear border with thin molecular layer and presence of large Purkinje cells. The presence of true neoplastic tissue was not found and regular CT controls were recom-



Figure 1. Axial FLAIR-weighted image (IR, 6741/110/1700) reveals ill-defined area of the increased signal intensity occupying the right cerebellar hemisphere with perifocal white matter oedema (arrow).

mended. She constantly suffered from headache and dizziness in exertion when she had the opportunity to do MRI eight years ago. After this first MRI which described tumourous mass in the pontocerebellar angle and the right cerebellar hemisphere, a new resection with drainage was recommended when she was 29-years old, but our patient did not accept surgery. The symptoms were stable during pregnancy when in the third trimester became more frequent and aggravated. The neurosurgeon recommended MRI before making decision for the route of child delivery. MRI was performed in the 29th week of pregnancy showing the expansive lesion of the right cerebellar hemisphere with characteristic features (Figures 1 and 2).

Nineteen years from her first symptoms our radiologist, based on typical MRI findings and history, concluded that it must have been Lhermitte-Duclos disease. The neurosurgeon recommended delivery by caesarean section and our patient delivered a healthy male child after 40 weeks of pregnancy. After delivery she reported the aggravation of symptoms: headache, dizziness, disturbance of balance



Figure 2. Coronal T2-weighted image (SE 4350/88) shows a large mass in the right cerebellar hemisphere with the incomplete distortion of the cerebellar folia (arrowhead).



Figure 3. Axial T2-weighted image (SE 4350/88) demonstrates a typical "tiger-striped" pattern of the lesion with compression of the fourth ventricle.

and paresthesia in both arms. She suffered from this discomforts for six months when she went on control MRI. Control MRI was unchanged, but she accepted the operation at that time. Pathohistological findings confirmed our radiological diagnosis.

Discussion

Tumours of ganglion cells are very rare. They include: gangliocytoma, ganglioneurinoma, Lhermitte-Duclos disease and dysembryoplastic neuroepithelial tumour. Some considered them to be dysplasias rather than true neoplasm; others refer to them as malformations.⁵ Lhermitte-Duclos disease is a rare cerebellar lesion with features of both malformation and benign neoplasm. MR imaging usually distinguishes the LDD by its characteristic "tiger-striped" appearance (Figure 3).⁶

In recent years several cases involving the association between LDD and Cowden's syndrome (CS), an autosomal dominant condition characterized by multiple hamartomas and neoplastic lesions in the skin and inter-

nal organs were reported. These included mucocutaneous lesions, acral keratosis, thyroid adenoma, fibrocystic disease ovarian cyst, intestinal polyposis, and arteriovenous malformation. Patients with LDD should receive a complete dermatological and systemic screening, because some of the lesions can develop into malignant tumors.⁷ The association between Lhermitte-Duclos disease and Cowden disease has been under-recognized and under-reported. The recognition of this association has a direct clinical relevance because a diligent long-term follow up monitoring of individuals with Lhermitte-Duclos disease and Cowden disease may lead to the early detection of malignancy.⁸ In approximately 40% of documented cases of LDD, CS can be diagnosed, and in 60% of cases LDD appears to occur sporadically.⁹ Patients diagnosed with Lhermitte-Duclos disease must be adequately evaluated for Cowden's syndrome.¹⁰

We presented a 37-year-old pregnant woman who had an isolated form of LDD beginning in her teenage period (16 years). During pregnancy she was under the permanent supervision of her obstetrician and no

obstetric complications were obtained. Pregnancy is an aggravating factor for brain tumours acting by three mechanisms: acceleration of tumour growth, increase of peritumoral oedema and development of immunotolerance to foreign tissue agents. There may be a relation between pregnancy hormones. According to Depret-Mosser *et al.*¹¹ induced therapeutic abortion and caesarean section are no longer routinely performed, and now being replaced by vaginal delivery with a systematic instrumental extraction. The presence of an intracranial neoplasm during pregnancy has a serious implication for the anaesthetic management of labour and delivery. The physiological changes of pregnancy and labour are potentially hazardous to women with intracranial neoplasm, but the provision of adequate pain relief during labour reduces the risk for the mother.¹² The other group of authors recommended caesarean delivery with the patient under general anaesthesia, followed by the immediate neurosurgical decompression in neurologically unstable patients to minimize temporal lobe or cerebellar herniation.¹³ The delivery should be advocated in the early third trimester after documentation of foetal pulmonary maturity. In our case we had a neurologically stable patient who reached the full term pregnancy. The obstetrician took into consideration her age (37 years), reported deterioration of symptoms in exertion and recommended neurosurgical examination before making his decision for elective caesarean delivery.

The way of delivery is still a question and should be solved between the obstetrician and the neurosurgeon for each patient individually. More experience with LDD in pregnancy is necessary for making a solid attitude about a way of delivery in neurologically stable patients. The management of brain tumours should be tailored to the individual patient. There may be a relation between pregnancy hormones and the rate of brain tumour growth mediated through specific intracellular receptors.¹⁴

More experience will be needed with this disease in pregnancy and post delivery period to recommend pregnancy for women with this condition. However, this case shows that a pregnant woman with LDD could reach full-term pregnancy and deliver a healthy child, without serious risk for her life.

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Lhermitte-Duclosova bolezen in nosečnost

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Izhodišča. Lhermitte-Duclosova bolezen ali displastični gangliocitom malih možganov je redka bolezen, ki lahko povzroča napredujočo tumorsko rast. Ugotovimo jo z magnetno resonanco, ki pokaže značilne neobarvane girusne spremembe, različne intenzitete na T1 In T2 magnetnih slikah.

Prikaz primera. Opisujemo 37-letno bolnico, ki smo ji dokazali Lhermitte-Duclosovo bolezen v tretjem tromesečju nosečnosti.

Zaključki. Prikazan primer kaže, da lahko nosečnica z Lhermitte-Duclosovo boleznijo brez življenjskega tveganja donosi in rodi zdravega otroka. Potrebni pa je več izkušenj, da bi to opažanje lahko posplošili.