of antiviral and immunosuppressive agents to prevent post herpetic autoimmune disorders.

AUTHORS' CONTRIBUTIONS

I. Soto treated the patient and wrote the manuscript. A. Bernardo treated the patient and contributed substantially to the manuscript. T. Arias treated the patient, performed bibliographical research and read the manuscript critically. C. Ramón treated the patient and read the manuscript critically. I. Noval performed image analysis procedures to monitor the patient. C. Palomo contributed as an internist to the knowledge of the autoimmune disease underlying the pathological process, contributed substantially to the manuscript and read the manuscript critically.

DISCLOSURES

The authors stated that they had no interests which might be perceived as posing a conflict or bias.

REFERENCES

1. Ichinose A. Autoimmune acquired factor XIII deficiency due to anti-factor XIII/13 antibodies: a summary of 93 patients. Blood Rev 2017;31:37–45.
2. Tone KJ, James TE, Fergusson DA, et al. Acquired factor XIII inhibitor in hospitalized and perioperative patients: a systematic review of case reports and case series. Transfus Med Rev 2016;30:123–131.
3. Ichinose A, Kohler HP, Philippou H; on behalf of the Factor XIII and Fibrinogen SSC Subcommittee of the ISTH. Recommendation for ISTH/SSC Criterion 2015 for autoimmune acquired factor XIII/13 deficiency. Thromb Haemost 2016;116:772–774.
4. Peyvandi F, Palla R, Menegatti M, et al. Coagulation factor activity and clinical bleeding severity in rare bleeding disorders: results from the European Network of Rare Bleeding Disorders. J Thromb Haemost 2012;10:615–621.
5. Tha M, Tien S. Acquired factor XIII deficiency: still a clinical challenge in the era of novel therapy. Haemophilia 2014;20:79–112.
6. Miesbach W. Case report—rituximab in the treatment of factor XIII inhibitor possibly caused by ciprofloxacin. Thromb Haemost 2005;93:1001–1003.
7. Kotake T, Souri M, Takada K, Kosugi S, Nakata S, Ichinose A. Report of a patient with chronic intractable autoimmune hemorhaphia due to anti-factor XIII/13 antibodies who died of hemorrhage after sustained clinical remission for 3 years. Int J Hematol 2015;101:598–602.
8. Leypoldt F, Titulaer MJ, Aguilar E, et al. Herpes simplex virus-1 encephalitis can trigger anti-NMDA receptor encephalitis: case report. Neurology. 2013;81:1637–1639.
9. Gilbert GJ. Herpes simplex virus-1 encephalitis can trigger anti-NMDA receptor encephalitis: case report. Neurology. 2014;82:2041.
10. Meyding-Lamadé UK, Oberlinner C, Rau PR, et al. Experimental herpes simplex virus encephalitis: a combination therapy of acyclovir and glucocorticoids reduces long-term magnetic resonance imaging abnormalities. J Neurovirol 2003;9:118–125.

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Progression of an untreated pseudotumor

The patient was a fifty-two-year-old male who was diagnosed with factor IX deficiency at the age of two. He developed an abdominal pseudotumour in this twenties. Surgery was considered but not performed for fear of operative complications. Over a five-year period beginning at age 47 years, the pseudotumour rapidly increased in size. The pseudotumour reached a final volume of 21 litres, roughly equivalent in volume and dimensions to a 5 gallon (U.S.) carboy of water. No antibodies to factor IX were detected upon repeated testing. By the end of the fifth year, the patient was suffering from a multitude of compressive complications secondary to the pseudotumour. Figure 1 shows the three-dimensional image reconstruction of the mature pseudotumour. Figure 2 shows the tumour’s growth over the five-year preoperative period. To produce these images, semi-automated segmentation and rendering were performed on two systems: General Electric Advantage Workstation v 4.3 (Waukesha, WI, USA).
LETTERS TO THE EDITORS

FIGURE 1 Three-dimensional image reconstruction of the mature pseudotumour in relationship to adjacent bony structures. The pseudotumour is shown in dark gray: (A) posterior view; (B) view from right side; (C) view from left side. The superior portion of the pseudotumour is within the costal margins. The insert in the lower right is used for image reconstruction purposes.

USA) and Osirix Lite 7.0.2 (Geneva, Switzerland). Images were semi-automated segmented using region growing and with manual segmentation on the Advantage Workstation. Pseudotumour volumes were computed and adjusted for slice thicknesses for each of the cases. Volume rendering for display was performed in Osirix. Surgery was eventually attempted due to severe satiety, but proved unsuccessful. The patient eventually succumbed to sepsis.

This patient was at the extreme end of the pseudotumour spectrum. Hopefully earlier intervention in the course of pseudotumour development is now generally being considered. Three-dimensionally reconstructed images and corresponding tumour volume calculations may provide a longitudinal perspective of pseudotumour development. These techniques may prove useful in planning the extent and timing of surgical intervention for pseudotumours.

AUTHOR CONTRIBUTIONS

MI reviewed the patient’s clinical data, wrote the manuscript and analysed radiographic data; PC originated the study, reviewed the manuscript and radiographic data; DW analysed the radiographic data.

FIGURE 2 Development of the pseudotumour over the five-year period as viewed from the left side. At the first time point only the inferior portion of the abdominal cavity is filled by the pseudotumour (shown in lighter red). As time passes, the pseudotumour expands to fill most of the abdominal cavity. The pseudotumour is depicted (from the left) at (A) 60 months, (B) 33 months, (C) 10 months and (D) 1 month prior to surgery.
Prenatal genetic testing by late amniocentesis to guide delivery management in haemophilia carriers

Infants with bleeding disorders have increased risk of haemorrhage during delivery and in the neonatal period. Since 2011, the Department of Clinical Genetics at Copenhagen University Hospital Rigshospitalet, Denmark, has offered late—near term—prenatal diagnosis by amniocentesis (AC) as an alternative to first trimester chorion villus sampling (CVS) to couples not considering termination of pregnancy in case of an affected foetus. Prenatal diagnosis enables the expectant parents and the health care professionals to prepare an optimal birth plan and to monitor and take care of the newborn with a known bleeding disorder.

Although recommended in international guidelines, there are few publications concerning late amniocentesis in haemophilia carriers. The option of late AC is desirable as early CVS/AC is associated with a risk of miscarriage of 0.5%-1.0%. While the mode of birth for infants with known risk of haemophilia remains controversial, there is consensus in guidelines and among obstetricians worldwide that invasive procedures such as foetal blood sampling and electrodes, mid-cavity forceps and vacuum extraction should be avoided in affected foetuses to minimize the risk of bleeding, especially intracranial haemorrhage. In cases with known risk of haemophilia in the offspring, the standard procedure is to offer parents counselling on the choice between vaginal birth and planned caesarean section (CS). Determination of gender is performed by ultrasound. Without prenatal diagnosis, this approach leads to unnecessary planned CS, or in case of vaginal birth excessive restrictions in the birth plan and even the risk of unnecessary emergency CS, as a male foetus has a 50% chance of being unaffected.

We here report on the results regarding management of delivery in 14 pregnancies during a 5-year period where late amniocentesis was performed. A similar approach has been published from Guy’s and St. Thomas’ Foundation Trust and in London. Our results underline the usefulness of foetal genotype determination in delivery management.

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The Hemophilia Centre of Copenhagen refers all expectant parents from eastern Denmark known to be affected by or carrier of haemophilia A or B for genetic counselling at Copenhagen University Hospital Rigshospitalet. The Hemophilia Centre has 244 registered patients with haemophilia A or B. All pregnancies from January 2011 to March 2016 in which late AC was performed to determine foetal genetic status regarding haemophilia A or B were identified through laboratory records. The patients gave written consent for inclusion in this study, and no patients declined counselling. Maternal carrier status, foetal genotype, gestational age by late AC, mode of birth and delivery management were recorded and are summarized in Table 1. Patient C and F both had had another pregnancy with a girl foetus seen by ultrasound and they declined late AC. No other patients declined late AC.

In her third pregnancy, she was referred to genetic counselling and late AC was performed. In all the pregnancies, the AC was performed around gestational week 36 because the risk of adverse outcomes at this gestational age is extremely low, and most likely the result is obtained before spontaneous delivery. No prohaemostatic treatment was given, and no adverse events related to the procedure occurred.

Fourteen pregnancies in ten women were eligible for inclusion in the study. Two girls were tested and found not to be carriers, five boys were found to be affected with haemophilia and seven boys were not affected. In all cases, the results from the amniocentesis were known prior to delivery, providing time to plan the optimal mode of delivery for each couple. Of the seven non-affected boys, two of them would in all probability have been delivered by an unnecessary emergency CS, had it not been for the performed late amniocentesis.

One of nine foetuses in all probability would have been delivered by emergency CS, had it not been for the performed late amniocentesis and the hereby known genetic status of the foetus. Patient E and patient F in our series both needed continuous monitoring of the foetus with scalp electrode and ST waveform analysis (STAN) together with foetal blood sampling as regular cardiotocography (CTG) was not sufficient. These procedures would not have been advisable, had the genotype of the foetus been unknown prior to delivery. Therefore, delivery would most likely have been by emergency CS. Three of five foetuses with predicted haemophilia were delivered by CS (60%). One of nine foetuses...