International Conference "ADVANCES IN PNEUMOLOGY"
Poznań, 6 – 7 June, 2008

RECURRENCE OF ARTERIO-VENOUS MALFORMATIONS WITH LIFE-THREATENING COMPLICATIONS IN A PREGNANT WOMAN WITH HEREDITARY HEMORRHAGIC TELEANGIECTASIA

E. Swietlik[1], A. Doboszynska[1, 2]

[1]Second Department of Internal and Pulmonary Medicine, Miedzyleski Hospital, Warsaw, Poland; [2]Clinical Nursing Department, Medical University of Warsaw, Warsaw, Poland;
em_swietlik@wp.pl

Hereditary hemorrhagic teleangiectasia is an autosomal dominant vascular disorder with incidence of 1 to 2,300 and high penetration, characterized by teleangiectasia, arterio-venous malformations and aneurysms. Arterio-venous malformations may occur in any organ but with the lungs, the brain and the liver as the most common ones. It has been noted that there is a significant morbidity and mortality risk associated with pregnancy in women in this otherwise relatively benign condition. To support this suggestion we present the case of 43-year-old woman, diagnosed with hereditary hemorrhagic teleangiectasia and treated with lobectomy at the age of 5 and with transcatheter coil closure of pulmonary feeding artery at the age of 30, who developed recurrence of arterio-venous fistulas during pregnancy. The woman was admitted to hospital with fatigue, dyspnea and severe right-sided headache in 15 HBD. 3-dimensional chest computed tomography revealed multiple arterio-venous fistulas. A head computed tomography scan revealed enhancing right parietal mass with surrounding edema and she underwent craniotomy. The diagnosis was brain abscess, suggesting an odontogenic etiology after dental treatment procedure. On the second day after craniotomy she developed right-sided ischemic stroke with left-sided hemipharesis and ischemia induced epilepsy. She went into premature labor at 35 weeks and the child was delivered by elective cesarean section. Seven days after delivery she developed severe pulmonary insufficiency, was diagnosed with hemothorax and treated with trascatheter embolotherapy. During the last 7 years of surveillance she developed progressing intrapulmonary shunt deterioration and hypertrophic pulmonary osteoartopathy. The presented patient developed the whole variety of pregnancy induced complications. The possible reasons for deterioration during pregnancy are: increased plasma volume, progesterone induced venous distensibility and a decrease in smooth muscle contractility. It is a matter of future research to establish genotype- phenotype correlations and assess disease severity using genetic tests. This would influence early diagnosis, prevention and treatment. For the time being regardless of the treatment modality,

long term follow up with spiral CT should be mandatory to monitor the development of new lesions and recurrence of those previously treated.