Newly diagnosed sporadic desmoid-type fibromatosis in pregnancy: is watchful waiting the option?

Franka Menge¹, Bernd Kasper², Ulrich Ronellenfitsch ³, Harald Abele ³, Peter Hohenberger¹

¹Division of Surgical Oncology and Thoracic Surgery, Mannheim University Medical Center, University of Heidelberg, Theodor-Kutzer Ufer 1-3, 68167 Mannheim; ²ITM - Interdisciplinary Tumor Center Mannheim, Theodor-Kutzer-Ufer 1-3, 68167 Mannheim ³Division of Gynaecology, University of Tübingen, Calwerstr. 7, 72076 Tübingen, Germany

Introduction: Desmoid-type fibromatosis (DTF) is an uncommon non-metastatic fibrous neoplasm with a poorly understood pathogenesis. A significant proportion of female patients with a DTF shows a recent pregnancy in their medical history. Nevertheless a modulation of DTF through hormonal signalling remains obscure. We describe the systematically assessed clinical presentation of 224 patients with histologically proven DTF with a special interest to assess the treatment needs if DTF and pregnancy develop synchronously. The course of a 35 yr. old patient is very intriguing and illustrative.

Patients: Of 224 patients with DTF treated from 2007-2012, 86 showed an abdominal manifestation of the DTF (43 females in the rectus abdominis muscle and another 43 patients with intraabdominal manifestations, see table 1). Of these, seven patients had a familial adenomatous polyposis syndrome (FAP). Of the 51 female pts. with documented pregnancies six got pregnant with an already known DTF or DTF was diagnosed first during pregnancy. All of them delivered a healthy child, see table 2.

Special case: An otherwise healthy female was referred during week 10 of her second pregnancy (first delivery by caesarean section two years ago). She had a history of repetitive in-vitro fertilisations for both pregnancies and of an overstimulation syndrome of the ovaries before the first pregnancy. Now she presented with a bifocal manifestation of a histologically proven DTF (β-catenin mutation T41A, estrogen and progesterone receptors negative) detected incidentally. The tumor was located in the paraduodenal retroperitoneum close to the mesenteric root and showed a size of 8.5 x 10.2cm. The patient was in excellent condition with no clinical complaints and a medically indicated abortion was no subject of debate for the patient. A watch and wait strategy was applied with frequent ultrasonographic monitoring. MRI controls showed a slowly growing DTF with a maximum size of 8.8 x 13.7cm over the time of pregnancy. Uncomplicated caesarean section followed in week 36 of pregnancy. With now 18 months of follow-up, a spontaneous downsizing of the DTF to 3.1 x 2.5cm was observed. No further treatment than continued surveillance is required.

To our knowledge only very few patients with a DTF occurring during pregnancy stayed without any treatment at all.

Conclusion: Even during pregnancy and in desmoids located at critical abdominal sites, a watch and wait strategy is the preferred option for patients with slowly growing tumors. There is no reason to stop pregnancy if a DTF is diagnosed in parallel and there is also no reason for an impetuous aggressive treatment after delivery.