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CLINICAL CASE

Case report: Rapidly growing abdominal wall giant desmoid tumour during pregnancy☆

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KEYWORDS
Desmoid tumour;
Pregnancy;
Rapid growth

Abstract
Background: Desmoid tumours are one of the rarest tumours worldwide, with an estimated yearly incidence of 2–4 new cases per million people. They are soft tissue monoclonal neoplasms that originate from mesenchymal stem cells. It seems that the hormonal and immunological changes occurring during pregnancy may play a role in the severity and course of the disease.
Clinical case: The case is presented on 28-year-old female in her fifth week of gestation, in whom an abdominal wall tumour was found attached to left adnexa and uterus while performing a prenatal ultrasound. The patient was followed up under clinical and ultrasonographic surveillance. When she presented with abnormal uterine activity at 38.2 weeks of gestation, she was admitted and obstetrics decided to perform a caesarean section. Tumour biopsy was taken during the procedure. Histopathology reported a desmoid fibromatosis. A contrast enhanced abdominal computed tomography scan was performed, showing a tumour of 26 cm × 20.5 cm × 18 cm, with well-defined borders in contact with the uterus, left adnexa, bladder and abdominal wall, with no evidence of infiltration to adjacent structures. A laparotomy, with tumour resection, hysterectomy and left salpingo-oophorectomy, components separation techniques, polypropylene mesh insertion, and drainage was performed. The final histopathology report was desmoid fibromatosis. There is no evidence of recurrence after 6 months follow-up.

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Conclusions: Desmoid tumours are locally aggressive and surgical resection with clear margins is the basis for the treatment of this disease, using radiotherapy, chemotherapy and hormone therapy as an adjunct in the treatment.

PALABRAS CLAVE
Tumor desmoide;
Embarazo;
Rápido crecimiento

Reporte de caso: tumor desmoides gigante de pared abdominal con rápido crecimiento durante el embarazo

Resumen
Antecedentes: Los tumores desmoides son neoplasias raras monoclonales de tejido blando, que surgen a partir de células madre mesenquimales. Son uno de los tumores más raros en todo el mundo, con una incidencia anual estimada de 2-4 nuevos casos por millón de personas. Los cambios hormonales e inmunológicos que ocurren durante el embarazo pueden desempeñar un papel en la severidad y curso de la enfermedad.
Caso clínico: Mujer de 28 años de edad, en su quinta semana de gestación, a quien, al realizar ultrasonido de control prenatal, se le encontró tumoración de pared abdominal adherida a anexos izquierdos y útero. Se dejó a la paciente en vigilancia clínica y ecográfica. Acudió con actividad uterina anormal, se ingresó a Obstetricia con 38.2 semanas de gestación, se realizó cesárea y se tomó biopsia de la tumoración. Anatomía patológica reporta fibromatosis desmoide. Se realizó tomografía axial computada contrastada, que reportó tumoración con bordes bien definidos, en contacto con el útero, anexo izquierdo, vejiga y pared abdominal; sin datos de infiltración a estructuras adyacentes de 26 × 20.5 × 18 cm. Se operó de forma electiva, se realizó laparotomía exploradora, con resección tumoral, histerectomía y salpingo-ooforectomía izquierda, técnica de separación de componentes, colocación de malla de polipropileno y drenajes. El reporte definitivo de enfermedad fue fibromatosis desmoide. Seis meses después de su intervención quirúrgica no ha presentado recurrencia.
Conclusión: Los tumores desmoides son localmente agresivos y la resección quirúrgica con márgenes libres representa la base en el tratamiento de esta dolencia; la radioterapia, quimioterapia y la hormonoterapia se utilizan como complemento en el tratamiento de estas pacientes.

Background

Desmoid tumours (or desmoid fibromatosis) are rare soft tissue monoclonal neoplasms arising from the mesenchymal stem cells. No association with metastatic disease has been reported. However, they often show a strong tendency to infiltrate local adjacent tissue, which causes significant morbidity and mortality.[1] Desmoid tumours are amongst the rarest in the entire world, with an estimated annual incidence of 2–4 new cases per million people.[2,3]

Desmoid tumours associated with pregnancy are rarer still, with few available publications in the literature. They have no known aetiological factor, however they can be associated with some familial syndromes such as familial adenomatous polyposis.[2] They have also been associated with hyperoestrogenic states and trauma, but the evidence for this is largely based on retrospective cases and anecdotal. Hormonal and immunological changes that occur during pregnancy can play a role in the severity and course of the disease.[1]

Clinical case

A 28-year-old women in the fifth week of gestation who, during a routine prenatal ultrasound, was found to have mass of approximately 11 cm × 15 cm × 18 cm, involving the left adnexa and uterus. The patient was placed under clinical and ultrasound surveillance. She consulted with abnormal uterine activity and was admitted to the obstetrics department at 38.2 weeks gestation. She underwent a midline caesarean section and a mass was found adhering to the abdominal wall, uterus and left adnexa. The general surgery department were consulted during the operation, a biopsy of the mass was taken and it was decided not to intervene until definitive diagnosis. The anatomopathological result was desmoid fibromatosis.

The patient was referred to the general surgery clinic. Physical examination found a distended, soft abdomen, peristalsis was present and there was no pain on palpation. The laboratory tests reported haemoglobin 11.5 g/dl. Contrast computed axial tomography (CAT) was performed,