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CLINICAL CASE

Intra-abdominal desmoplastic small round cell tumour☆

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KEYWORDS
Peritoneal tumour;
Peritoneum;
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Abstract
Background: The desmoplastic small round cell tumour is a rare and aggressive intra-abdominal neoplasia, with only 200 cases reported, and a higher incidence in men and predilection for the second decade of life. Histologically it is characterised by the presence of small nests of undifferentiated tumour cells, wrapped in fibrous desmoplastic stroma.
Clinical case: A 24-year-old male started with abdominal pain of 4 weeks onset in the right upper quadrant, colic type, sporadic, self-limiting and accompanied by early satiety, decreased appetite, and involuntary weight loss of 10 kg in 3 months. At the time of admission the abdomen was globular, with decreased peristalsis, soft, and depressible. Computed tomography of the abdomen showed multiple enlarged lymph nodes in the abdominal-pelvic cavity. A laparotomy was performed, with a subsequent omentum resection due to the presence of multiple tumours, which microscopically were characterised by groups of small, round, blue cells, separated by a desmoplastic stroma. The immunohistochemistry was positive for desmin (>75%), epithelial membrane antigen (>75%), CD99 (>50%), and S100 (25%), concluding with an abdominal tumour of small, round, blue cells as a diagnosis. Chemotherapy treatment was initiated based on IMAP plus GM-CSF.

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Conclusions: The desmoplastic small round cell tumour is a rare neoplasia, with diagnostic complexity and a lethal course. Its clinical presentation is unspecific. Histologically, it is classified as an aggressive soft tissue sarcoma that shares similar characteristics with the family of the small and blue cells tumours.

PALABRAS CLAVE
Tumor peritoneal;
Peritoneo;
Tumor de células pequeñas y redondas;
Células mesoteliales

Tumor intraabdominal desmoplásico de células pequeñas y redondas

Resumen
Antecedentes: El tumor intraabdominal desmoplásico de células pequeñas y redondas es una entidad rara y agresiva con solo 200 casos reportados, con una incidencia mayor en varones y predilección por la segunda década de la vida. Histológicamente se caracteriza por la presencia de nidos de células tumorales pequeñas e indiferenciadas, envueltas en estroma fibroso y desmoplásico.
Caso clínico: Varón de 24 años, que en las 4 semanas previas comienza con dolor abdominal en hipocondrio derecho, tipo cólico, esporádico, autolimitado, acompañado de saciedad temprana, hiporexia y pérdida involuntaria de 10 kg de peso en 3 meses. A su ingreso se encuentra abdomen globoso, con peristalsis disminuida, blando, depresible. La tomografía computada de abdomen evidencia múltiples adenomegalias en la cavidad abdominopélvica. Se realiza laparotomía exploradora con la consecuente resección del omento por la presencia de múltiples tumoraciones, las cuales microscópicamente se caracterizaban por grupos de células redondas, pequeñas y azules separadas por un estroma desmoplásico. La inmunohistoquímica reveló positividad para desmina (>75%), antígeno de membrana epitelial (>75%), CD99 (>50%) y S100 (25%), por lo que se diagnostica tumor desmoplásico abdominal de células redondas, pequeñas y azules. Se inició tratamiento quimioterapéutico con base en esquema IMAP más GM-CSF.
Conclusiones: El tumor intraabdominal desmoplásico de células pequeñas y redondas es un tumor poco frecuente, de complejidad diagnóstica y de curso letal. Clínicamente presenta manifestaciones inespecíficas. Histológicamente se clasifica como un sarcoma agresivo de tejidos blandos, que comparte características similares con la familia de tumores de células pequeñas y azules.

Background

The intra-abdominal desmoplastic small round cell tumour is a very rare aggressive neoplasm, with approximately 200 cases reported since it was first described in 1989 by Gerald and Rosai,[1] who described that it has a higher incidence in the male gender, with a 2–10:1 ratio and it was more frequent during the twenties in 80% of the cases (range 4–52 years).[1–9]

It mainly affects serous membranes, particularly those of the peritoneum, and of these lesions, 62% are located in the abdomen and the remaining 36% in the pelvis; there are cases of primary tumours located in the testicles, scrotum, pleura, posterior fossa, petrous portion of the temporal bone, eye orbit and abdominal organs. Most of the cases are advanced stage, large tumours without apparent organic origin, which may be accompanied by large tumour implants located in the peritoneum.[2–6,8]

Histologically, it is characterised by the presence of small and undifferentiated tumour cell nests, embedded in a fibrous and desmoplastic stroma. It is a sarcoma (of round cells) which is genetically characterised by the expression of a reciprocal translocation of the gene t(11;22)(p13;q11 or q12), resulting from a fusion of the Ewing sarcoma gene and chromosome 22 of the Wilms tumour (WT1), mainly located in the intra-abdominal region.[2,3,5,6,8–11]

The *objective* of this report is to present a typical case of a desmoplastic small round cell tumour.

Clinical case

A 24-year-old male patient, with no history of surgeries, trauma, autoimmune diseases or medication intake. He presented symptoms 4 weeks before his admission, including sudden colic, sporadic, 10-minute long abdominal pain in the right hypochondrium, accompanied by early satiety, hyporexia and involuntary loss of 10 kg of weight in the last 3 months. The patient denied having fever, asthenia, adynamia, as well as other relevant history. On admission, he was found to be haemodynamically stable, without fever and without relevant pathological data. He presented a soft,