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Disseminated ovarian alveolar rhabdomyosarcoma as a rare cause of peritoneal carcinomatosis

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ABSTRACT

Peritoneal carcinomatosis due to the presence of primary ovarian rhabdomyosarcoma of alveolar subtype (OARMS) is an extremely rare malignant tumour, with very few cases reported in the international literature. Herein, we present an interesting case of diffuse peritoneal dissemination of OARMS, in a previously asymptomatic patient, who was investigated for progressive abdominal distension.

KEY WORDS: *Ovary; rhabdomyosarcoma; carcinomatosis*

CASE PRESENTATION

A 56-year-old Caucasian female patient was assessed in the outpatient surgical clinic with symptoms of abdominal distension and alteration of bowel habits with predominance of constipation. Clinical examination revealed the presence of a palpable pelvic mass extending to the periumbilical level. An urgent computed tomography (CT) scan of the abdomen and pelvis detected a 19x25cm multi-lobulated mass of probable gynaecological origin, with encasement of the greater omentum and transverse colon. Colonoscopy was unremarkable and no obvious metastatic deposits were evident on a subsequent staging chest CT, while pelvic ultrasound confirmed the gynaecological origin of the mass (Figure 1).

After informed consent, the patient was taken to the operating theatre for exploratory laparotomy, with the intraoperative findings revealing the presence of disseminated peritoneal carcinomatosis, with the described complex pelvic mass infiltrating the right and transverse colon, as well as a significant portion of the small bowel. Hence, debulking surgery was performed and the patient returned to the ward for routine postoperative care; she was discharged in a stable condition on the 6th postoperative day. The patient succumbed approximately 40 days from the operation due to pulmonary embolism, prior to discussions regarding palliative adjuvant therapy. Histopathological assessment of the extracted specimen revealed the presence of disseminated ovarian rhabdomyosarcoma of alveolar subtype, with diffuse positive staining for vimentin, 60-70% positive staining for desmin and 30-40% positive nuclear staining for myogenin (Figure 2).

Being an uncommonly encountered tumour, OARMS has an unpredictable biological behaviour and diagnosis is made after meticulous histopathological assessment [1,2]. Therefore, to the best of our knowledge, no solid consensus exists regarding its optimal treatment. In our case, upfront exploratory was performed due to the absence of non-peritoneal disease, however, due to the extensive small bowel encasement in multiple

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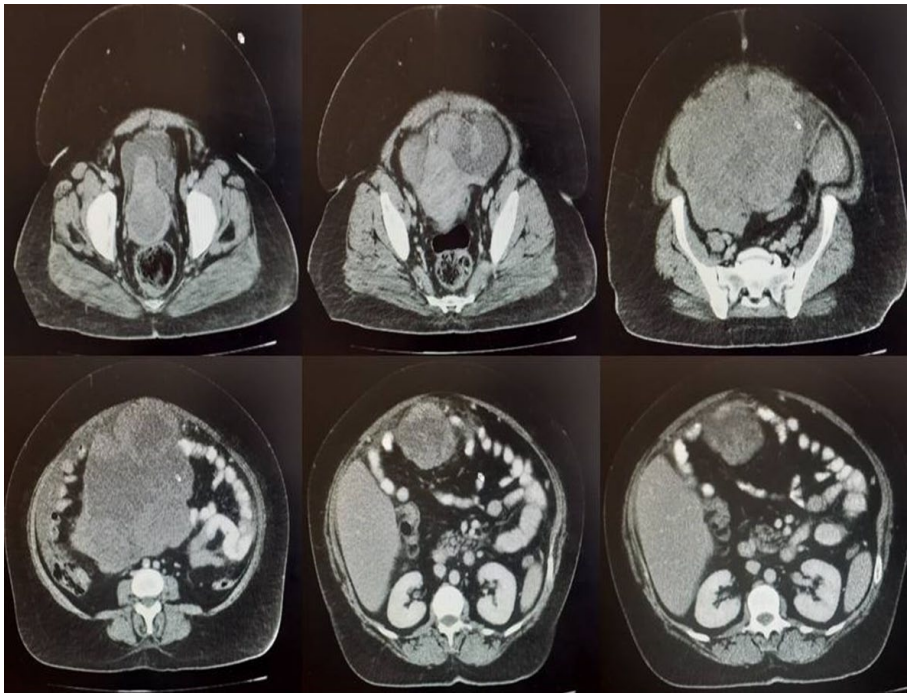

FIGURE 1. Compilation of CT images demonstrating a complex pelvic mass extending to the upper abdomen.

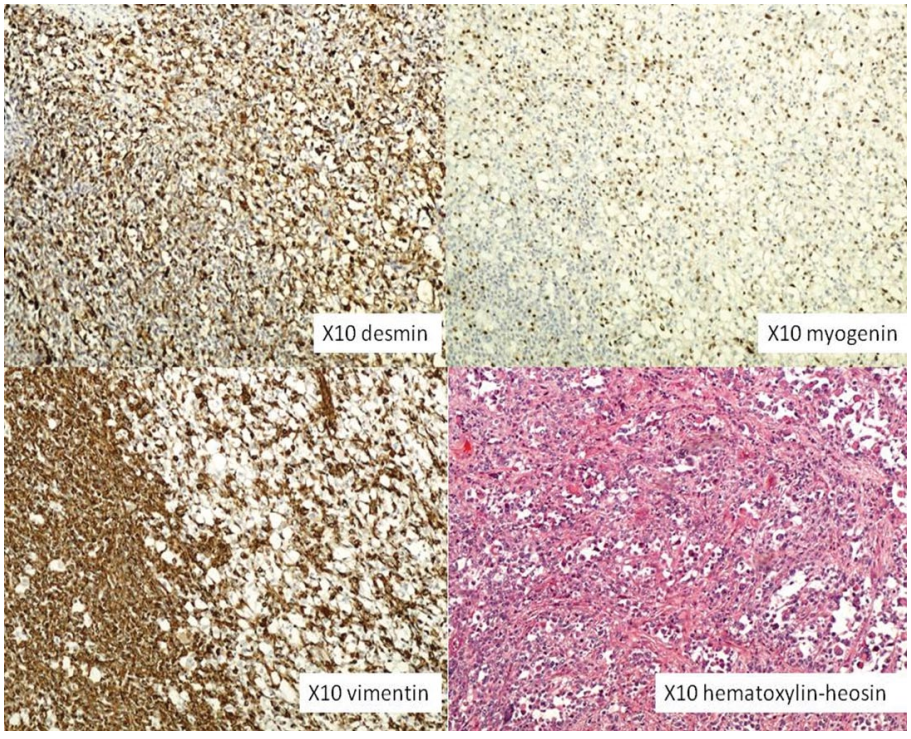

FIGURE 2. Compilation of ovarian alveolar rhabdomyosarcoma key histopathological images demonstrating -including others- positive staining for desmin, myogenin and vimentin.

levels, complete cytoreduction was not achieved and we opted for maximal tumour debulking.

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GZ: contributed to the design of the case report presentation and performed the final revision of the manuscript.

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