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Primary Ewing's sarcoma of the small intestine

José J. Paricio[1*], Juan Ruiz Martín[2] , Esther Sánchez Díaz[1]

[1]Pathology Department. Hospital Nuestra Señora del Prado. Talavera de la Reina. Spain

[2]Pathology Department. Hospital Virgen de la Salud. Toledo. Spain

*Correspondence: jparicio@alumni.unav.es

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Dear Editor,

We present the case of a 17 year old woman, who presented to the emergency deparment for abdominal pain. In the abdominal CT scan a pelvic mass of 12 x 10 cm is observed, which seems to depend on the ovary. Surgery is programmed for extirpation of the tumor, during which it is observed that the lesion originates in the ileum, which is removed.

The histological examination shows a neoplasm of small round cells (Fig1), of vesicular nuclei with nucleolus and scant cytoplasm, with atypical mitoses and necrosis. The immunohistochemical study showed CD99 and ERG positivity, being negative for cytokeratins, FLI1, WT1, DOG1 and lymphoid markers. By means of FISH it was demonstrated rearrangement of the EWSR1 gene.

Ewing's sarcoma is an aggressive neoplasm with cells of neuroectodermal origin that usually occurs in long bones in pediatric and young adult patients[1]. In the gastrointestinal tract, the small intestine is the location with the highest incidence, mainly in ileum[2].

The most frequent clinical presentation is abdominal pain accompanied by fatigue, although it may debut as obstructive conditions, perforation or even be asymptomatic[3]. Due to its clinical inespecificity and low incidence, it can be confused with other lesions, such as gastrointestinal stromal tumors[4].

Management in these patients is surgery followed by polychemotherapy. Evolution is usually unfavorable, with poor prognostic factors being advanced age, poor response to chemotherapy and the presence of metastases at diagnosis[3].

Microscopically, it is a proliferation of atypical small round cells, whose differential diagnosis includes lymphoma, desmoplastic small round cell tumor, rhabdomyosarcoma, microcytic carcinoma or undifferentiated adenocarcinomas with rhabdoid characteristics[5].

This entity presents a characteristic molecular alteration[1], the translocation of the EWSR1 gene, located on chromosome 22. The most frequent translocation (85 %) occurs with FLI1, of chromosome 11, with the next most frequent being the rearrangement with ERG, of chromosome 21.

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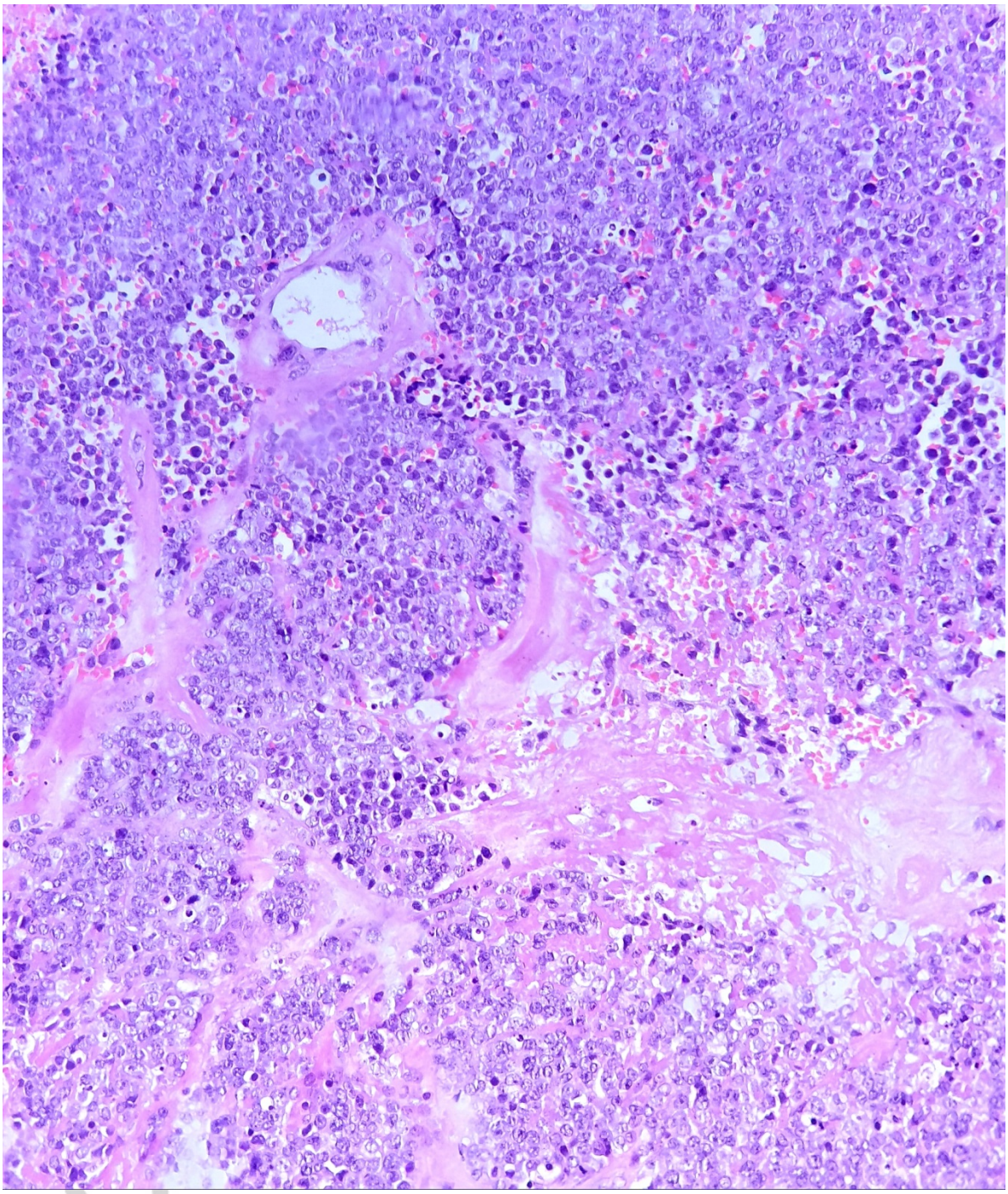

Figure 1. Solid pattern malignant proliferation, consisting of small round atypical cells, with vesicular nuclei that frequently present prominent nucleoli. Areas of necrosis are recognized.