Duodenal Ewing’s sarcoma: Unusual location and atypical EWRS-1 translocation

Ernesto Jesús Barzola-Navarro1, José Ángel Flores-García1, Diego López-Guerra1, Cristina Tejera-Pérez2, Nerea Rodríguez-Díez3, Aurea Gómez-Durán4, Alejandro Rubio-Fernández4 and Gerardo Blanco-Fernández1

1Hepatobiliary and Pancreatic Surgery Unit, 2Endocrine Unit, 3Digestive Unit, and 4Pathology Unit. Hospital Universitario de Badajoz. Badajoz, Spain

CASE REPORT

A 20-year-old woman was admitted, because 5 months before admission, she started having epigastric abdominal pain and lost weight. On examination the patient was pale, had abdominal pain when deep palpation was applied.

In laboratory tests, a microcytic hypochromic anemia was found. Results of computed tomography (CT) of the abdomen showed a rounded heterogeneous mass adjacent to the pancreas (Fig. 1).

An oral endoscopy revealed in the fourth duodenum portion a neoplastic appearance of an ulcer, where a biopsy was taken (Fig. 2). The pathological and immunohistochemistry studies showed neoplastic cells positive for CD99, FLI-1 (friend leukemia integration 1 transcription factor), and CD 117. With a suspected sarcoma, a molecular study of traslocation t (11; 22), (q24; q12) in the locus 22q12 of EWSR1 gene (Ewing sarcoma breakpoint region 1) was done (Fig. 3). These findings were compatible with Ewing’s sarcoma (ES).

The surgery performed was cephalic pancreaticoduodenectomy (Fig. 4).

Fig. 1. Abdominal scan: Solid mass of 40 mm in diameter situated in the third and fourth duodenum portion.

Fig. 2. Endoscopy: Ulcer of neoplastic appearance with thickened edges and fibrin at the bottom.
After that, the adjuvant chemotherapy was administered. After 9 months of follow up treatment the patient has recovered.

DISCUSSION

Only a few cases of gastrointestinal location of ES have been reported, being mostly located in the small intestine and more frequently in young people (1). The symptoms are aggravated by duodenal compression and ulcers. Total surgical removal is the best solution for a full recovery (2).

Systematic use of chemotherapy in ES in the last two decades had increased the survival level (3). ES is a radiotherapy sensitive tumor and this treatment is applied in cases of affected margins.

REFERENCES


Fig. 3. A. HE x 40 tumors cells with rounded nucleus and eosinophilic cytoplasm. B. Nuclear expression of FLI-1 gene in tumor cells. C. FISH (fluorescence in situ hybridization) region 22q12 shows a translocation of EWSR1.

Fig. 4. Surgical piece from cephalic pancreaticoduodenectomy.