

## Multicentric CD34 negative epithelioid angiosarcoma of the digestive system

Fabio Pagni<sup>1</sup>, Biagio Eugenio Leone<sup>1</sup>, Susanna Ronchi<sup>1</sup>, Paola Sartori<sup>2</sup>, Luigi Corti<sup>2</sup>, Dario Maggioni<sup>2</sup>

(1) Department of Pathology ; (2) Department of Surgery and Endoscopy, Desio Hospital, Università Milano Bicocca, Desio, Italy.

### To the Editor,

We would like to report a particular case of a gastrointestinal angiosarcoma that recently occurred in our hospital. A 74-year-old male was referred to our surgical department because of severe anemia due to repeated episodes of melena and right cervical swelling associated with an oral lump of the upper dental arcade which was bleeding profusely (Fig. 1). A gastroscopy showed some haemorrhagic gastric and duodenal polyps, some of which were removed. A colonoscopy revealed the presence of multiple rectal, sigmoid and colonic polyps grossly similar to the gastric ones (Fig. 1). A total body CT scan showed multiple nodules, localized both in peritoneum, next to duodenum, spleen and mesenteric artery and in retro-peritoneum. Histological examination of gastric and rectal polyps showed epithelioid malignant cells with large clear cytoplasm, pleomorphic nuclei and often an eosinophilic nucleolus. The immunohistochemical panel excluded carcinoma (CK pool, CK7, CK20, EMA-), melanoma (S100, HMB45, MART1-) and hematologic malignancy (CD45, CD20, CD79a, CD68pgm1, CD3-). Among mesenchymal tumors, also GIST (CD117, CD34-) and leiomyosarcoma / rhabdomyosarcoma (SMA, myogenin-) were eliminated. These cells were immunoreactive for CD31 (Fig. 1) and focally for factor VIII. A final diagnosis of CD34 negative epithelioid angiosarcoma was proposed. A cervical lymph node FNAB demonstrated a metastatic lesion with morphological and immunocytochemical features similar to gastrointestinal findings. The patient was scheduled for chemotherapy with gemcitabine but his clinical status quickly got worse due to ongoing bleeding requiring daily red blood cell transfusions. Thus chemotherapy could not be started and he received only palliative care. This case is in accordance to literature reviews (1) with regard to the aggressive behavior of these rare multi-centric lesions (2). While literature supports that CD34 staining is the most sensitive marker for angiosarcoma (3), our paper shows that performing only this marker of vascular differentiation in an immunohistochemical panel is not enough and could lead to diagnostic delays or mistakes. Indeed, there are vascular tumors completely negative for CD34, but positive for other specific markers (CD31, Fly-1, D2-40) (4).

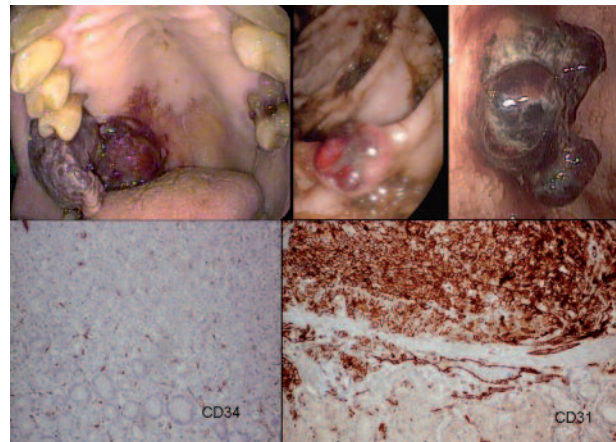


Fig. 1. — Macroscopic appearance of epithelioid angiosarcoma in oral cavity, stomach and large bowel. Immunohistochemical negativity of the neoplastic cells for vascular marker CD34 (note internal control in normal endothelium) ; strong reactivity for more specific CD31.

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Correspondence to : Dr. Fabio Pagni, M.D., Department of Pathology, Desio Hospital, Università Milano Bicocca, Desio, Italy. E-mail : petala.83@tiscali.it

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