Multifocal Sarcomatoid Carcinoma of the Small Intestine

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Introduction
Sarcomatoid carcinomas (SCs) or carcinosarcomas are rare, biphasic tumors, with poor prognosis, only rarely located in the small and large intestine. The first treatment option of these tumors is surgical approach, adjuvant therapy showing no important role in the treatment protocol. From immunohistochemical point of view, tumor cells were strongly positive for cytokeratin AE1/AE3 as well as for vimentin. They were negative for epithelial membrane antigen (EMA), CD117, CD34, S100, chromogranin-A and synaptophyisin. Based on the macroscopic and microscopic appearance, respectively the immunohistochemical feature of the tumor, the patient was diagnosed with multifocal sarcomatoid carcinoma of the small intestine.

Keywords: sarcomatoid carcinoma, small intestine, immunohistochemistry

Case presentation
We present the case of a 63 years old male patient, hospitalized in the Surgical Department II of the County Emergency Clinical Hospital Tîrgu Mureș with the diagnosis of small intestine tumor with jejunal localization. Microscopically, in hematoxylin-eosin (HE) staining, the tumor was consisting of two components, an epithelial and a mesenchymal one. From immunohistochemical point of view, tumor cells were strongly positive for cytokeratin AE1/AE3 as well as for vimentin. They were negative for epithelial membrane antigen (EMA), CD117, CD34, S100, chromogranin-A and synaptophyisin. Based on the macroscopic and microscopic appearance, respectively the immunohistochemical feature of the tumor, the patient was diagnosed with multifocal sarcomatoid carcinoma of the small intestine.

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Received: 6 July 2012 / Accepted: 31 July 2012
nodes were free of metastasis. (TNM stage pT4N0, Mx). From immunohistochemical point of view, the tumor cells were strongly positive for cytokeratin AE1/AE3 as well as for vimentin (Figure 2, Figure 3). They were negative for epithelial membrane antigen (EMA), CD117, CD34, S100, chromogranin-A and synaptophisin (Figure 4).

Discussions

Small intestine carcinoma is a rare entity in current practice, as represented in general by adenocarcinoma and only exceptionally by the sarcomatoid carcinoma [3,10–12]. Only 23 cases of small intestine SC are described in the literature. The patients related general data showed that SC is more frequent in the male gender (M/F=2/1) while the average age of onset is 55 years. Regarding the localization it was more frequent described in the ileum and jejunum (12 and 11 cases respectively), and only one case was found in the duodenum. Macroscopically it appears as a single tumor mass, with size of about 8 cm. Most of these tumors recorded in the literature were polypoid, pedunculated, or exophytic, the infiltrative appearance being rarely described [3,10,13].

The multifocal nature [2] of the primary tumor was described only in two cases, a situation encountered also in our patient, the two tumor masses being located from each other at a distance of 30 mm. Even though both tumors were small, because of their endophytic nature they were protruding on the surface of the serosa.

Histogenesis of malignant tumors in literature represents an uncertain and very controversial topic. Some of the authors are of opinion that carcinomatous proliferation can also induce an excessive stromal proliferation – theory of stromal induction/metaplasia/fusion -, while other researchers support the theory that malignant epithelial and mesenchymal proliferation/ hyperplasia occurs simultaneously but separately – collision tumors theory – the two components originating from different stem cells [14,15]. The third group of researchers are sustaining the concept of the existence of a “totipotent stem cells”, capable of epithelial and mesenchymal differentiation. These issues are also reflected
by the current histopathologic examination which describes SC of monophasic or biphasic nature [1]. Microscopically the typical biphasic SC includes the presence of the epithelial and mesenchymal components, cells of both components expressing epithelial markers such as cytokeratin and also markers of mesenchymal differentiation such as vimentin, from immunohistochemical point of view [8,13].

In terms of nomenclature this is also very diverse, due to the opinion of different researcher groups. Some authors in describing the SC considered the architecture, organization and the laying out of the two tumor components. In this respect the two components were arranged in distinct areas inside the same tumor, these being also called „true carcinosarcomas”, while in other cases the two components were overlapping thus being described as "so-called carcinosarcomas” [13,14].

There is a certain classification of these tumors in three categories. In type 1 of the „collision tumors”, there is a distinct boundary between the two tumor components. In type 2 or „combined tumors” the two components are coexisting, being confluent and present in all areas of the tumor, while the mesenchymal component shows no distinctive features. And in type 3 or “composite tumors” the two components of the tumor are present, the stromal component showing a different feature [3,8,15].

Other researchers in the nomenclature of these tumors gave importance to the immunohistochemical profile. If the epithelial and stromal cells express the epithelial marker cytokeratin, the tumor is termed „sarcomatoid carcinoma”, but if stromal cells do not express epithelial markers, the term „carcinosarcoma” is preferred [1,3,8,12,15]. In these cases it is important to assess some staining technical problems that may lead to misdiagnosis, respectively to carry out immunohistochemical reactions on several sections from different areas of the tumor.

The epithelial component is usually present in the form of glandular structures or organized in nests or cords, and rarely as isolated tumor cells. Cells of the epithelial component are large, polygonal cells with abundant, eosinophilic cytoplasm, and rounded or oval, vesicular nuclei, with an obvious nucleolus. Arrangement of epithelial tumor cells in the form of trabeculae may raise problems of differential diagnosis with neuroendocrine tumors. But the epithelial tumor cells do not express neuroendocrine markers, while in cases of immunopositivity this is observed focally and isolated in some cells [12].

The mesenchymal component is composed of spindle cells arranged in bundles more or less organized, with a small amount of eosinophilic cytoplasm, and elongated, oval, vesicular nuclei, with obvious nucleoli, showing a marked mitotic activity. SC in that the mesenchymal component predominates can raise problems of differential diagnosis with gastrointestinal stromal tumors (GISTs), leiomyomas, and leiomyosarcomas.

The immunohistochemical profile is important, SC being CD 117 and CD 34 negative tumors, what differentiates them from GISTs and do not express myoid differentiation markers, such as desmin or smooth muscle actin, markers that are positive in all benign and malignant tumors of muscular origin. This component may contain occasionally anaplastic cells of multinucleated gigantic cell type, and also various tissue components of differentiated mesenchymal origin such as the osteoid, chordoid, neural, adipose or rhabdoid type [7,10,15,16].

Regarding the management of these tumors, it is described that the only effective therapy method proved to be surgical resection due to early local recurrence and development of hepatic and lymphonodular metastasis, patients presenting an ineffective response to chemo- and radiotherapy [3,6,7,17].

Usually all small intestine carcinomas have very bad prognosis. Thus prognosis of SC is also poor, most of the cases described in the literature being large tumors, with local and distant metastasis, death in patients occurring within a range of 2–39 months after diagnosis.

Conclusions

The sarcomatoid carcinoma is a rare tumor, with a mixed, epithelial and mesenchymal, malignant component, whose microscopic appearance represents a real challenge for pathologists. Immunohistochemistry is a modern and very useful tool for positive and differential diagnosis, with a major importance in the subsequent management of patients.

The histopathological feature, tumor stage and the evolution indicates the fact that usually they are very aggressive tumors, with bad evolution characterized by rapid growth, local invasion, and early metastasis inducing death in patients shortly after diagnosis.

References


