Undifferentiated Pleomorphic Sarcoma of the Sigmoid Colon: A case report

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Key clinical message: Elderly patients with abdominal pain are easy to be misdiagnosed as gastrointestinal cancer. The possibility of UPS should be recognized in clinical work.

Abstract: UPS is the most common soft tissue malignant tumor in the elderly. It usually occurs in the proximal limbs and retroperitoneum, and is usually manifested as a progressively enlarged painless mass. Due to its histological aggressiveness, it has a poor prognosis, with a five-year survival rate of less than 50% and frequent metastasis or recurrence. UPS in the colon is rare. Due to its special location, atypical clinical manifestations, and lack of characteristic pathological signs, the diagnosis is usually a diagnosis of exclusion. Early and complete resection of the tumor is the main treatment for UPS. Although some studies

have shown that postoperative adjuvant radiotherapy and chemotherapy have a certain local control rate, the effect of radiotherapy and chemotherapy is not completely certain, and more cases are needed to further study its efficacy.

Key words: Undifferentiated pleomorphic sarcoma, colon, treatment.

Introduction

Undifferentiated pleomorphic sarcoma UPS is the most common soft tissue malignant tumor in the elderly. It usually occurs in the proximal limbs and retroperitoneum, and is usually manifested as a progressively enlarged painless mass[1]. Due to its histological aggressiveness, it has a poor prognosis, with a five-year survival rate of less than 50% and frequent metastasis or recurrence. UPS in the colon is rare. Due to its special location, atypical clinical manifestations, and lack of characteristic pathological signs, the diagnosis is usually a diagnosis of exclusion. Early and complete resection of the tumor is the main treatment for UPS. Although some studies have shown that postoperative adjuvant radiotherapy and chemotherapy have a certain local control rate[2], the effect of radiotherapy and chemotherapy is not completely certain, and more cases are needed to further study its efficacy.

Case report

A 54-year-old man was admitted to the hospital due to left abdominal pain for 1 week. The pain was persistent and tolerable, without other accompanying symptoms. Abdominal Computed tomography (CT) scan showed a huge mass of mixed density shadow in the lower abdomen and pelvis, the size was about 16.3*6.8cm. Enhanced scan showed uneven enhancement of the lesion, and the mass was closely related to the intestine, and the boundary was not clear (Figure 1). The tumor was suspected as gastrointestinal stromal tumor. During the operation, a new tumor was found on the serosal surface of the sigmoid colon, with the size of 19cm*14cm*10cm, and no metastatic lesions were found around it (Figure 2). Gross examination showed that the tumor was encapsulated on the surface, with rich blood vessels. The cut surface was gray white and gray red, which was cystic and solid, soft in texture, containing a large amount of mucus, accompanied by necrosis and hemorrhage in some areas. Microscopically, the tumor cells were arranged disorderly, irregularly shaped, varying in size, with obvious atypia, numerous mitotic figures, prominent nucleoli, and a large number of tumor giant cells and multinucleated tumor giant cells (Figure 3). The tumor stroma was rich in blood vessels with myxoid degeneration. Immunohistochemically, the tumor cells showed a high Ki-67 proliferation index, focally positive for CD34 and S-100 (Figure 4), and a mutant expression pattern of P53. The pathological diagnosis was high-grade undifferentiated pleomorphic sarcoma with negative margins (no tumor cells in the margins). The patient did not receive any adjuvant therapy after surgery. Three months later, CT scan showed multiple mixed density shadows in the lower abdomen and pelvis, considering tumor metastasis or recurrence.

Discussions

High-grade undifferentiated pleomorphic sarcoma (UPS), formerly known as malignant fibrous histiocytoma (MFH), is characterized by a progressively enlarged, painless mass[1]. It mainly occurs in the soft tissues of the extremities and the retroperitoneum, and rarely occurs in the digestive tract. Grossly, the tumor was a solitary nodule with a fish-like, offwhite to brown cut surface. Hemorrhage and necrosis were common[1]. UPS of the colon may originate from mesenchymal stem cells in the mesentery[3][4]. Histologically, UPS of the colon is characterized by large atypical cells with more mitoses and prominent nucleoli. The atypical cells can show different morphologies, including pleomorphic, myxoid, xantomatous, giant cell and angiomatoid types. They are broadly positive for CD68 and vimentin, and focally positive for CK and EMA. Due to its high degree of malignancy and strong invasion, it is easy to relapse and distant metastasis after surgery. Distant metastasis is most common in lung, followed by bone and liver[5][6]. Wide resection with negative resection margins (RO resection) is the main treatment for UPS, and negative resection margins help to reduce the risk of tumor recurrence. However, the recurrence rate of UPS is still high, and the 5-year survival rate is less than 50%. Tumor size, location, degree of differentiation and completeness of surgery are closely related to the prognosis of UPS[5]. The effects of postoperative adjuvant radiotherapy and chemotherapy have not been

reported, but some studies have shown that compared with surgery alone, although adjuvant radiotherapy and chemotherapy can't improve the overall survival rate, they can significantly improve the local control rate of high-grade soft tissue sarcoma[2].

Conclusions

Due to the low incidence of UPS, there are few cases available for clinical study, and the effective treatment of this disease still needs further research. UPS arising from the colon is rare. For patients with abdominal pain and obvious abdominal mass, the possibility of UPS should be considered. Early and complete resection of the mass is the main treatment.

Author contributions:

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Writing – review & editing: Yuchang Hu, Yuxia Yang

Abbreviations:

UPS= Undifferentiated pleomorphic sarcoma

CT= Computed tomography

MFH= Malignant fibrous histiocytoma

EMA= Epithelial membrane antigen

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