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Letter to the Editor

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Reply to Commentary on "Clinical Characteristics and Adequate Treatment of Familial Adenomatous Polyposis Combined with Desmoid Tumors"

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Virgilio et al. [1] reported a case with familial adenomatosis coli (FAP)-related giant desmoid tumors (DTs) of the mesentery following proctocolectomy, and I mostly agree with the authors' inoperable mortality. They reported a 28-year-old female with FAP and DTs abruptly aggravated to death at 6 months postoperatively which was probably related with extensive DTs. Similarly, mesenteric DTs were the main cause of death in FAP patients with DTs, whereas recurrent cancers were the main cause of death in those without DTs, in our previous study [2]. The patient seems to carry three potential risk factors for DTs including female, *APC* mutation site 3′ to 1440, and previous history of surgery [3]. Herein, the current case gives us an important lesson for careful attention about possible DTs during operation and immediately after proctocolectomy, particularly in FAP patients with risk factors. Although the response might be unpredictable for complicated DTs, non-surgical treatment options would be immediately provided if any symptoms or signs were identified during early postoperative period. For the extensive DTs, nonsurgical treatment options including radiotherapy, systemic chemotherapy including doxorubicin plus dacarbazine, biologics such as imatinib, non-steroidal anti-inflammatory agents, and anti-hormonal agents, have been provided to yield variable responses. Among these options, anti-estrogen agents, alone or in combination with nonsteroidal anti-inflammatory drugs, were identified with an overall response rate of 51% according to a systematic analysis using a total of 168 DTs [4]. We have experienced estrogen receptor antagonist combined with or without luteinizing hormone releasing hormone agonist to stabilize extensive mesenteric DTs. In a total of 10 FAP patients accompanying extensive mesenteric DTs which were not completely resected, seven patients have been alive well leading ordinary lifestyles for 17-133 months (median, 101 months) after diagnosis, showing partial response or stable disease to tamoxifen with or without goserelin acetate. On the other hand, several groups presently reported the graft preservation rate after multi-visceral transplantation as approximately 50% in patients with extensive abdominal DTs [5]. Although bowel transplantation is considered as a life-saving procedure, knotty subjects must be preferentially verified to be an established practice, i.e., severe morbidity, high graft failure and mortality, recurrent DTs, and accessibility. Conclusively, further studies including surgical and non-surgical modalities treating FAP patients with extensive DTs are needed to consolidate the evidence for the efficiency and safety of respective regimen.

Conflicts of Interest

Conflict of interest relevant to this article was not reported.

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