

Title:
**Desmoid fibromatosis of the
esophagogastric junction**

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Desmoid fibromatosis of the esophagogastric junction

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Key words: Desmoid fibromatosis. B-catenin. FAP.

Dear Editor,

Desmoid fibromatosis is a mesenchymal clonal proliferation which lacks metastatic potential. Nevertheless, it has an infiltrative growth and thus implies a high morbidity (1). Although the etiology remains unclear, mutations in the B-catenin or APC genes are involved. Some risk factors include pregnancy, hormonal exposure or surgery. Desmoid fibromatosis can be sporadic (80%) or FAP-associated (2). In sporadic cases, it is caused by mutations in the B-catenin (CTNNB1) gene. Whether it is FAP-associated or not should be determined, as the treatment for each condition is different. A radiologic test is essential for diagnosis, although a biopsy is necessary for confirmation. With regard to treatment, there is a wide range of different alternatives such as observation only, medical treatment or even surgery (3). However, a recurrence rate that ranges from 30% to 40% has been reported in the major published series (4) and thus, conservative treatment is more common nowadays.

Case report

We present the case of an 82-year-old male with constitutional syndrome. A computed tomography was performed, which identified a 69 x 52 mm mass in the esophagogastric union (Fig. 1). A computed tomography guided biopsy was performed and the histological analysis identified a fusocellular tumor compatible with desmoid fibromatosis. Treatment was started with indomethacin. However, a control computed

tomography three months later showed that the mass had grown. Thus, indomethacin treatment was stopped and tamoxifen treatment was started. The patient has had an excellent performance status since symptom presentation.

Conclusion

In conclusion, desmoid tumors are rare and most are sporadic. However, they may also be associated with familial adenomatous polyposis syndrome. It must be emphasized that our patient did not have any risk factors and the anatomical location in the esophagogastric union is not a common location. Desmoid fibromatosis represents a clinical challenge for diagnosis and treatment and thus, management should be individualized.

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Fig. 1. ACT scan showing a 69 x 52 mm mass in the esophagogastric union (red circle).

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