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Extra-abdominal desmoid tumors. Medical treatment or surgery?

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Background: Extra-abdominal desmoid tumor is a rare benign neoplasm. The etiology is unknown although some studies associate mutations of the APC (adenomatous polyposis coli) gene and the regulation of β -catenin. Its treatment has varied from simple clinical and imaging surveillance to surgery and adjuvant therapies. Enlarged excision has been the treatment of choice but, as it is benign, mutilating interventions are not justified to achieve tumor-free margins. Presently there have been outlined various therapies as an alternative to surgical treatment.

Methods: 4 women and 1 man, aged between 21 and 69 years old, operated in the following anatomical locations: cervical, distal posterior thigh, paravertebral lumbar, dorsal and periescapular. Diagnoses were made by percutaneous biopsy. Margins were considered R1 only in periescapular tumor and so was the only one proposed for adjuvant radiotherapy.

Results: After an average follow-up of 23.6 months (4-40) all are free of disease. The tumor of the thigh recurred one year later and was treated with surgery and radiation, without new recurrence. Functional outcomes were good, except for periescapular case with a poor outcome (16/30 MSTS).

Discussion/Conclusions: Characteristically this tumor spans between muscle fibers, invades nerve structures and doesn't form a pseudocapsule, and therefore is a challenge for the enlarged excision. Most recent literature claims good results in the control and monitoring of these tumors with radiotherapy alone. Some cytotoxic molecules are also able to control the disease: vincristine, methotrexate, vinorelbine, anthracyclines, dacarbazine and pegylated liposomal doxorubicin. Imatinib and sorafenib also have promising results. Cryoablation is also an option, particularly if they are small and distant from important anatomical structures. Although wide tumor excision is still the safest treatment, there are recurrences. Besides, if there is involvement of important neurologic structures, the expected poor functional outcome should make us think in medical treatment