LETTER TO THE EDITOR

Intra-abdominal leiomyosarcomas in two sisters. Müllerian or somatic type tumors?

Dear Editor,

We read with interest the reports of intra-abdominal leiomyosarcomas by Koczkowska *et al*. [1]. The tumors occurred in two sisters in the mesentery, and both were leiomyosarcomas with almost identical histological and genetic features, indicating a possible autosomal recessive trait in the tumor development.

As both patients were female, it would be interesting to know whether these leiomyosarcomas were of Mullerian or somatic type. Although current data are still limited, several recent published findings indicate that extra-uterine leiomyomas and leiomyosarcomas occurring in the region of secondary Müllerian system [2] (peritoneum, retroperitoneum, extragenital pelvic tissue, inguinal region) can be subdivided into Müllerian (uterine type) and non-Müllerian (somatic) types, and that these types should represent distinctive groups of smooth muscle tumors with different morphological, clinical and genetic features [3-6]. Immunohistochemically, the Müllerian type leiomyomas and leiomyosarcomas express estrogen receptor, progesterone receptor, and WT1, in contrast with non-Müllerian somatic type tumors, which are negative for these antigens.

In case the tumors reported by Koczkowska *et al*. [1] are Müllerian type leiomyosarcomas, the significance of such a finding for carcinogenesis and an estimation of the risk of such leiomyosarcoma for male members of the family would be our questions to our colleagues in oncologic genetics.

References

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LETTER TO THE EDITOR REPLY

We appreciate the interest in our paper [1] and raising the issues referring to the potential histogenesis of leiomyosarcoma (LMS). Indeed, LMS developing in the "secondary Müllerian system" area may be split into Müllerian and non Müllerian somatic types. Expression of the estrogen (ER) and progesterone receptor (PR), and Wilms tumor 1 protein (WT-1) may be helpful in this regard, since only the tumors from the former group present these markers.

At diagnosis, both our tumors did not express ER and PR. Similarly, there was no nuclear expression of Wilms tumor 1 protein (WT-1), suggesting somatic

derivation of these tumors. As pointed out in the mentioned papers [1, 2], significance of the histogenetic derivation of LMS is not established. Expression of ER may have some diagnostic value in distinguishing uterine from extrauterine LMSs. Of course, as shown earlier [1, 3], the absence of these markers does not totally exclude Müllerian derivation of ER-/WT-1-negative LMSs. In addition, LMS showing ER expression may have more favorable clinical behaviour [1, 2], although this data is preliminary. Reassuming, at the present state of knowledge, significance of somatic vs. Müllerian phenotype in LMS is not finally proven. Likewise, estimation of the LMS risk in male members of the family reported in our case study would be highly speculative and not based on the scientific grounds.

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