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Case report

A rare case of vulvar superficial myofibroblastoma associated with ambigous and unusual differential diagnosis

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1. Backgroud

Superficial myofibroblastoma (SMF) is a rare benign mesenchymal tumour affecting the female genital tract. To date, only 51 cases, (Laskin et al., 2001; Ganesan et al., 2005; Stewart et al., 2005; Olinici et al., 2007; Wang et al., 2010; Magro et al., 2012; Gonzàlez Martìnez et al., 2005; Abdelaziz et al., 2017; Peng et al., 2019; Adams et al., 2009) aged between 23 and 80 years old, (Ganesan et al., 2005) have been reported in the English language literature, one of which occurred in a pregnant woman. (Adams et al., 2009) For many years, SMF has been known as "superficial cervico-vaginal myofibroblastoma", as it was believed to occur exclusively in the cervix and vagina. In 2005, Ganesan *et al.*, 2005 discovered that some tumours with the same histological and immunohistochemical features could also occur in the vulva; for this reason, they proposed to rename this kind of neoplasm with the term "superficial myofibroblastoma of the lower genital tract". Hitherto, only 5 cases of SMF with vulvar localization have been reported; (Ganesan et al., 2005; Magro et al., 2012; Peng et al., 2019) therefore, vulvar SMF can be considered an extremely rare condition, that may not be easy to recognize. Histopathologically, it is characterized by a discrete, even though unencapsulated, myofibroblast proliferation located in the subepidermal tissue, with a thick Grenz zone separating the lesion from the epidermis. It is constituted by oval to spindle-shaped cells, with wavy nuclei and scant cytoplasm, within a loosely collagenous stroma. The cellularity is always moderate to low, while dilated thin-walled blood vessels may be abundant, sometimes with a stag-horn pattern. Cytological atypia as well as mitoses and necrosis are extremely rare. (Laskin et al., 2001; Ganesan et al., 2005) For its clinical and histological features, especially whether an extensive stromal oedema is associated, the main condition with which SMF is usually placed in differential diagnosis is aggressive angiomyxoma, whose clinical and prognostic characteristics are radically different from SMF. (Peng et al., 2019) Other potential mimics of SMF mentioned in the international scientific literature include: mammary type myofibroblastoma, angiomyofibroblastoma and cellular angiofibroma, with which SMF shares similar histological features, as well as a myxoid type of dermatofibrosarcoma, which can be difficult to differentiate whenever SMF shows prominent myxoid changes. (Peng et al., 2019)

2. Case presentation

A 77-year-old Caucasian woman referred to our Hospital due to a bulky swelling of the right Labia Majora (Fig. 1A), which developed about one year earlier, suddenly and without an apparent trigger, and had gradually increased in size. She did not report any kind of symptoms related to the swelling, except for a sense of gravity and an annoying encumbrance. The patient's medical history revealed she was on therapy for blood hypertension and took antiplatelet agents for primary cardiovascular prevention. She had had two vaginal deliveries and was in iatrogenic menopause since the age of 46, when she underwent total hysterectomy with bilateral salpingo-oophorectomy for benign disease, following which she never took hormonal replacement therapy. The only other surgical procedures reported in her medical history were a saphenectomy and appendenctomy. Her Pap-smear was up-to-date and negative. The gynecological examination revealed the presence of a painless, elongated, soft and rubbery in texture swelling, measuring about 10 cm in length, placed at the front portion of the right Labia Majora and extended to the ipsilateral portion of the Mons pubis. It seemed to be superficial and movable with respect to the surrounding anatomical planes (Fig. 1A). The inner margin of the right Labia Majora, as well as the left Labia Majora and both Labia Minora, both vaginal entry and vagina were regular. The differential diagnosis was posed between a cyst/hydrocele of the Nuck duct or an inguinal/crural hernia (even if it did not increase in size under Valsalva maneuver and was not manually reducible). However, a neoplasm could not be clinically excluded. On ultrasound examination, the swelling showed a mixed content (liquid and probably mucous) and did not appear vascularized at power Doppler; moreover, there was no evidence of intestinal loops inside. The surgical removal of the above described formation was decided. A median longitudinal incision was made on the most inferior portion of the bulky swelling, which was firstly carefully

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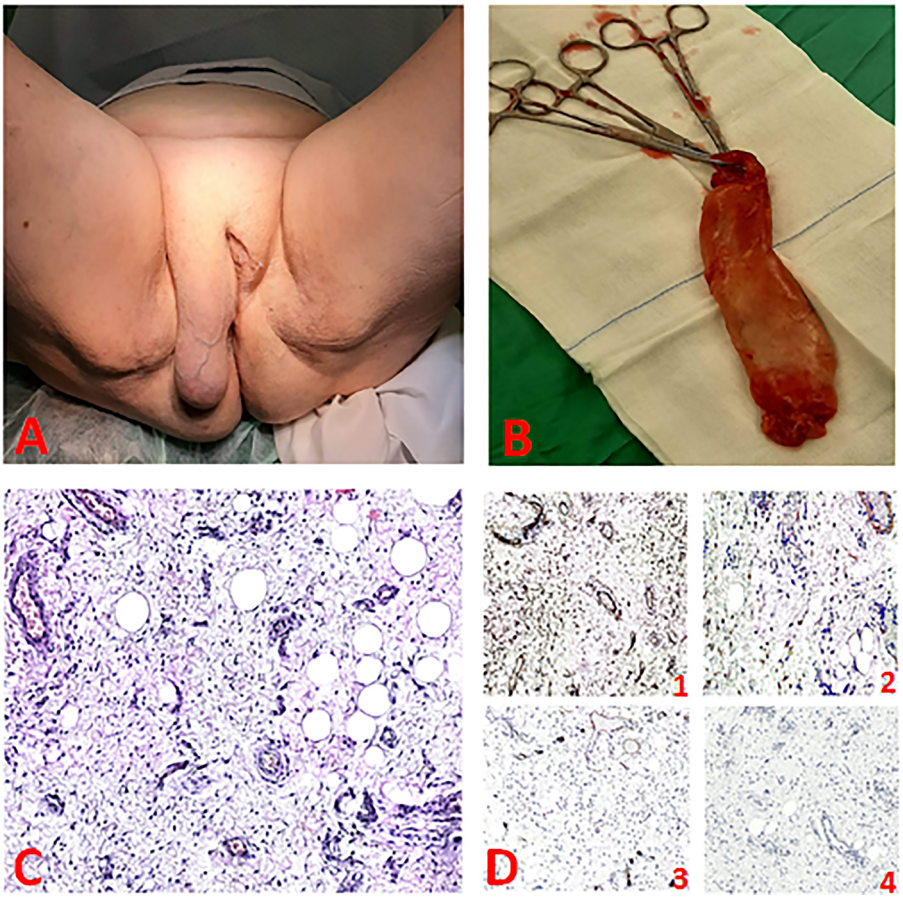

Fig. 1. A: Clinical appearance; B: Macroscopic appearance of the removed lesion; C: Histopathological findings: Finely collagenous stroma with dilated thin-walled vessels contains oval or spindled neoplastic cells with bland and wavy nuclei and scant, if present, cytoplasm. Mitoses, necrosis and cytological atypia are absent (Haematoxylin-eosin staining. Original magnification: 10x). D: Immunohistochemical findings: D1: CD34 +; D2: desmin +; D3: smooth muscle actin -; D4: S100 protein - (Original magnification: 20×).

isolated from the surrounding anatomical planes and then completely removed. Grossly, the tumour mass was located subcutaneously and measured 12x3x3cm (Fig. 1B); it did not have a fibrous capsule but was well circumscribed. The cut surface showed a yellowish-white mass with mucoid appearance and dilated cysts. The tumour mass appeared completely removed and showed focal dilatation of Bartholin glands; haemorrhagic extravasation was detected in the overlying skin. On histopathological examination, it was constituted by bland spindle-shaped, more than oval, cells, with wavy nuclei and scarce amount of cytoplasm, when present. Cellular elements were immersed in a finely collagenous stroma, with focal mucoid changes, in the context of which slightly dilated, thin-walled vessels were also observed. No necrosis, mitoses or cytological atypia were detected (Fig. 1C). On immunohistochemical analysis, tumour cells showed positivity for CD34 (Fig. 1,D1), desmin (Fig. 1, D2), and ER, while they were uniformly negative for smooth muscle actin (Fig. 1, D3), S100 protein (Fig. 1, D4), and Melan-A (data not shown). The proliferation index, assessed by Ki-67, was lower than 1%. The complex of histopathological and immunohistochemical findings were compatible with a SMF, which appeared completely excised.

3. Discussion and conclusions

The diagnosis of genital mesenchymal tumours is usually challenging as they are rare and some of them share many similar clinic-pathological features. Differently from the cases of SMF reported in literature, in which the problem of differential diagnosis was mainly histological, in the case we have presented the problem of differential diagnosis was related to both location and clinical aspect of the vulvar swelling. As a matter of fact, if the diagnosis of SMF appeared unequivocal from a pathology point of view (considering the absence of myxoid degeneration or stromal oedema, the superficial localization, the hypocellularity, the thin-walled vessels, the expansive growth pattern with clear margin detected, as well as the immunohistochemical findings), the localization at the front portion of the Labia Majora and its extension to the ipsilateral portion of the Mons pubis, as well as the elongated, soft and rubbery in texture appearance, could generate confusion with a cyst/hydrocele of the Nuck duct or with an inguinal/crural hernia, or an otherwise undetermined neoplasm. As evidenced by the clinical case we have presented, since vulvar SMF is extremely rare, it can undergo a differential diagnosis with conditions other than those commonly reported in literature, some of which even not of gynaecological relevance. In the diagnostic-therapeutic framework of vulvar swellings, it is therefore important to take into account the wide spectrum of clinic-pathological conditions that can be faced with, among which SMF represents a rare but anyway possible occurrence. Owing to the variety of the possible differential diagnoses, it is important to have the most comprehensive knowledge of the conditions underlying vulvar swellings, in order to be able to direct the surgeon towards the most suitable and appropriate surgical approach.

Informed consent

Written informed consent was obtained from the patient for anonymized publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

CRediT authorship contribution statement

Lodovico Patrizi: Conceptualization, Methodology, Resources, Project administration. **Barbara Borelli:** Investigation, Writing - original draft, Visualization. **Monia Di Prete:** Resources. **Valentina Bruno:** Investigation. **Alessandro Mauriello:** Supervision. **Emilio**

Piccione: Supervision. **Carlo Ticconi:** Validation, Writing - review & editing.

Declaration of Competing Interest

The authors declared that there is no conflict of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.gore.2020.100637.

References

Laskin, W.B., Fetsch, J.F., Tavassoli, F.A., 2001. Superficial cervicovaginal myofibroblastoma: fourteen cases of a distinctive mesenchymal tumor arising from the specialized subepithelial stroma of the lower female genital tract. Hum. Pathol. 32 (7), 715–725. https://doi.org/10.1053/hupa.2001.25588.

Ganesan, R., McCluggage, W.G., Hirschowitz, L., Rollason, T.P., 2005. Superficial myofibroblastoma of the lower female genital tract: report of a series including tumours with a vulval location. Histopathol 46 (2), 137–143. https://doi.org/10.1111/j.1365-2559.2005.02063.x.

Stewart, C.J.R., Amanuel, B., Brennan, B.A., Jain, S., Rajakaruna, R., Wallace, S., 2005. Superficial cervico-vaginal myofibroblastoma: a report of five cases. Pathol 37 (2), 144–148. https://doi.org/10.1080/00313020500058284.

Olinici, C.D., Crisan, D., Zolog, A., Puscas, M., 2007. Vaginal superficial myofibroblastoma. Case report and review of the literature. Roman J Morph Embriol 48, 165–170 PMID: 17641804.

Wang, Q.F., Wu, Y.Y., Wang, J., 2010. Superficial cervicovaginal myofibroblastoma: report of four cases and literature review. Chin. Med. J. 123, 1093–1096 PMID: 20497721.

Magro, G., Caltabiano, R., Kacerovskà, D., Vecchio, G.M., Kazakov, D., Michal, M., 2012. Vulvovaginal myofibroblastoma: expanding the morphological and immunohistochemical spectrum. A clinicopathologic study of 10 cases. Hum. Pathol. 43 (2), 243–253. https://doi.org/10.1016/j.humpath.2011.04.027.

Gonzàlez Martìnez, M.B., Dìaz Dìaz, C., Martìnez Alonso, M.C., Lòpez-Anglada Pérez, J.E., 2005. Miofibroblastoma cérvico-vaginal superficial. Presentaciòn de un caso y revisiòn de la literatura. Rev Esp Patol 38 (3), 183–187.

Abdelaziz, M., Eziba, N., Sharma, S., Kleven, D., Al-Hendy, A., 2017. Cervical superficial myofibroblastoma: case report and review of the literature. SAGE Op Med Case Rep 5, 1–5. https://doi.org/10.1177/2050313X17726936.

Peng, W.X., Wada, R., Kure, S., Fukunaga, M., Naito, Z., 2019. Superficial myofibroblastoma in the vulva mimicking aggressive angiomyxoma: a case report and review of the literature. Case Rep Pathol 1–5. https://doi.org/10.1155/2019/1582714.

Adams, B., Fogarty, P., McKenna, M., McManus, D., 2009. Superficial myofibroblastoma of the lower femal genital tract: first case report of a pregnant patient. Obst Case Rep 28 (6), 657–658. https://doi.org/10.1080/01443610802421668.